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CASE REPORT

Takayasu Arteritis Presenting as Orbital Apex Syndrome With Irreversible Monocular Blindness and Long-Segment Right Carotid Stenosis: A Case Report With 58 Months of Follow-Up

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ABSTRACT

Background: Takayasu arteritis is a granulomatous large-vessel vasculitis that usually becomes apparent only after stenotic damage has occurred and can be difficult to recognise when classical peripheral vascular signs are absent.

Objective: To describe the clinical course, diagnostic challenges and long-term outcome of Takayasu arteritis presenting as orbital apex syndrome with irreversible monocular blindness and long-segment right carotid stenosis.

Case Presentation: A 36-year-old man presented in August 2020 with a chronic progressive right hemicranial headache that acutely worsened and was followed by complete loss of vision in the right eye. Examination showed right optic nerve dysfunction, ptosis, proptosis, absent pupillary reflexes and complete ophthalmoplegia involving the third, fourth and sixth cranial nerves, a constellation corresponding to orbital apex syndrome. There were no constitutional symptoms, no limb claudication and no significant inter-arm blood pressure difference. Contrast-enhanced computed tomography of the brain and orbits was unrevealing. Computed tomography angiography seven days later showed long-segment luminal irregularity and stenosis of the right common carotid artery extending continuously into the cervical, petrous, cavernous and supraclinoid internal carotid segments, with a preserved contralateral carotid and posterior circulation; the erythrocyte sedimentation rate was elevated. Takayasu arteritis was diagnosed and treated with high-dose glucocorticoid followed by azathioprine and antiplatelet therapy. The headache resolved but the visual loss did not. Applied retrospectively, the 2022 American College of Rheumatology/EULAR criteria yielded one point against a threshold of five, because three examination items were never recorded and aortic imaging was never done. At 58 months the patient was clinically stable but had grade 3 lymphopenia and grade 1 thrombocytopenia.

Conclusion: Takayasu arteritis should be considered in young adults with progressive headache and focal neuro-ophthalmic deficits even when pulses and blood pressures are symmetrical.

1. Introduction

Takayasu arteritis is a chronic granulomatous vasculitis of the aorta and its major branches. Its clinical expression is determined less by the intensity of systemic inflammation than by which arterial beds are narrowed, and the disease therefore presents as a collection of loosely related ischaemic syndromes rather than as a single recognisable illness. In a data-driven cluster analysis of 852 patients drawn from a large international cohort, in whom 81.5% were women and the median age was 30.6 years, the anatomical distribution of disease separated patients into phenotypes with materially different prognoses: compared with a supra-aortic pattern, an abdominal pattern carried the highest risk of vascular complications (hazard ratio 1.79, 95% confidence interval 1.12–2.87) and a diffuse pattern the greatest risk of relapse (hazard ratio 2.31, 95% confidence interval 1.81–2.95).[1] A 13-year single-centre series from a Chinese national referral centre similarly recruited 852 patients, of whom 670 were female and 182 male, a female-to-male ratio of 3.7:1, and found that male patients more often had systemic hypertension (62.1% versus 42.4%) and renal artery involvement (62.7% versus 53.9%).[2] A Japanese nationwide cohort of 129 patients with newly diagnosed disease reported that 84% were female and that the mean age at onset was 35 years.[3] The disease is therefore not rare in men, and male sex has repeatedly been associated with the more damaging end of the phenotypic spectrum: among patients with coronary involvement, severe stenosis or occlusion was more frequent among the coronary lesions of men than of women (70.5% versus 46.0%, p=0.004), and death from any cause over up to seven years of follow-up was substantially higher at the patient level (21.4% versus 1.5%, p=0.003).[4]

There is no disease-specific laboratory biomarker, and acute-phase reactants track arterial inflammation imperfectly, so diagnosis rests on the integration of clinical assessment with vascular imaging. The 2022 American College of Rheumatology/EULAR classification criteria formalised this dependence by making both an age of 60 years or younger at diagnosis and imaging evidence of vasculitis absolute requirements, then summing ten weighted clinical and imaging items to a threshold of five points; in the validation set this produced a sensitivity of 93.8% (95% confidence interval 88.6% to 97.1%) and a specificity of 99.2% (95% confidence interval 96.7% to 100.0%).[5] Independent validation has been favourable but not uniform. In 504 patients with Takayasu arteritis and 222 controls the new criteria were more sensitive than the 1990 American College of Rheumatology criteria (95.83% versus 82.94%) but considerably less specific (63.51% versus 90.54%),[6] whereas in a Chinese cohort of 778 patients and 378 comparators both sensitivity and specificity were high, at 95.2% (95% confidence interval 93.4–96.5%)

and 95.8% (93.1–97.5%) respectively, with an area under the curve of 0.955 (0.940–0.970).[7] What these figures cannot capture is performance when the source record is incomplete, which is the situation in most retrospective case material and in much of routine practice.

Delay between symptom onset and diagnosis remains substantial even in specialist hands. Among 46 children with Takayasu arteritis or polyarteritis nodosa managed at a tertiary paediatric rheumatology centre, the median diagnostic delay was 13 weeks.[8] Delay matters because a proportion of the damage that accrues is not reversible. Neurological events are among the most consequential. In a 24-year retrospective series, stroke occurred in 6.3% (26 of 411) of patients with Takayasu arteritis, and among those affected the common carotid artery was involved in 73.0% (19 of 26), a figure matched exactly by the subclavian artery, and the internal carotid artery in 57.7% (15 of 26).[9] A multicentre cohort that compared 108 patients with stroke complicating either giant cell arteritis or Takayasu arteritis found a striking territorial dissociation: the carotid territory accounted for 79.5% of strokes in Takayasu arteritis but only 35.3% in giant cell arteritis, whereas the vertebrobasilar territory predominated in giant cell arteritis (75% versus 20.5%, both p<0.001).[10] A case-control study of 35 patients with stroke and 50 without identified internal carotid artery involvement (odds ratio 5.98, p=0.004) and male sex (odds ratio 5.70, p=0.038) as independent predictors.[11] Carotid disease severity measured non-invasively carries the same message: among 295 patients, of whom 260 (88.14%) were female, 93 (31.53%) experienced neurological severe ischaemic events, and a left carotid intima-media thickness to diameter ratio of 0.55 or more conferred an odds ratio of 2.75 (95% confidence interval 1.24–6.07, p=0.01).[12]

Ocular involvement is common and frequently sight-threatening. In an Italian cohort, ocular manifestations were detected in 37.2% of patients (16 of 43) and were often responsible for severe visual deterioration, with visual acuity ranging from normal in 7 eyes through 20/40 to 20/200 in 20 eyes down to counting fingers in 2 eyes, hand motion in 2 eyes and no light perception in 1 eye.[13] The published ocular presentations of Takayasu arteritis are dominated by Takayasu retinopathy, ocular ischaemic syndrome, retinal arterial occlusion and arteritic anterior ischaemic optic neuropathy, and the outcomes are often poor: a patient with arteritic anterior ischaemic optic neuropathy complicating Takayasu arteritis presented with counting fingers close to the face and died of the illness within three months despite maximal immunosuppression,[14] while a young woman with simultaneous ischaemic stroke and anterior ischaemic optic neuropathy had no light perception with a relative afferent pupillary defect at presentation and improved only to counting fingers on follow-up.[15] Even after successful revascularisation, recovery is unpredictable and asymmetric between the two eyes.[16]

Two features of the present case sit awkwardly against this literature. First, the classical peripheral vascular signs on which clinicians and criteria alike rely were either absent or, more often, never recorded. A pre-pulseless phase is well described and is not uncommon: among 238 patients with angiographically proven disease, 91 (38.23%) were classified as pre-pulseless, and these patients more frequently had deranged renal function (adjusted odds ratio 4.43, 95% confidence interval 1.58–12.37) and Hata type IV disease (adjusted odds ratio 8.02, 95% confidence interval 2.61–24.65).[17] Second, the presenting syndrome was an orbital apex syndrome, in which the optic nerve and all three ocular motor nerves fail together. Series of orbital apex syndrome are dominated by structural and infectious causes rather than by large-vessel disease. Among 66 patients, inflammation accounted for 54.54% (36 of 66), neoplasm for 18.18% (12 of 66), trauma for 15.15% (10 of 66), fungal infection for 9.09% (6 of 66) and vascular causes for only 3.03% (2 of 66);[18] in a southern Taiwanese series of twenty patients, neoplasm led at 55% (11 of 20) with vascular causes accounting for 10% (2 of 20), and among the fourteen patients in whom both an initial and a final acuity were recorded, more than half remained legally blind and only 28.6% (4 of 14) improved.[19] A vasculitic cause of orbital apex syndrome is therefore a diagnosis that the base rates actively discourage.

The novelty of this report lies in the combination of an orbital apex syndrome as the sentinel manifestation of Takayasu arteritis, an angiographically unilateral long-segment carotid lesion extending without interruption from the common carotid artery to the supraclinoid internal carotid artery, a complete absence of documented peripheral vascular signs, and 58 months of longitudinal follow-up that exposes the safety consequences of maintenance immunosuppression. The aim of this study is threefold: to describe in detail a neuro-ophthalmic presentation of Takayasu arteritis that conventional structural imaging could not explain; to demonstrate, by explicit item-by-item scoring, why a validated classification instrument returns a near-zero score in a patient whose diagnosis is nevertheless secure, and what that implies for the use of such instruments on incomplete records; and to draw attention to two findings in the longitudinal record that were documented but not acted upon, namely a sustained grade 3 lymphopenia on thiopurine maintenance and an abdominal aortic angiogram that was planned but never performed in a hypertensive patient.

2. Case Presentation

Patient information and clinical examination

A 36-year-old man presented in August 2020 with a chronic right-sided headache that had been progressive for an extended period and had recently become markedly worse. The headache was followed by loss of vision in the right eye. On presentation the right eye was closed. Examination demonstrated right optic nerve dysfunction with complete loss of vision in that eye, together with paresis of the right oculomotor, trochlear and abducens nerves producing complete right ophthalmoplegia. Additional ophthalmic findings were ptosis, proptosis and absent pupillary reflexes on the right. The combination of optic neuropathy with total ophthalmoplegia in the same orbit corresponds to an orbital apex syndrome. The patient remained fully conscious throughout, with a Glasgow Coma Scale score of E4V5M6.

There was no reported fever, weight loss, night sweats or limb claudication, and no clinically significant difference in blood pressure between the arms was identified. Several elements of the vascular examination that are central both to the traditional clinical picture of Takayasu arteritis and to its classification were not documented: bilateral peripheral pulse assessment, carotid pulse and carotid tenderness, and auscultation for arterial bruits. On the ophthalmic side, formal Snellen or logMAR visual acuity, fundoscopy, optical coherence tomography, fluorescein angiography and visual field testing were likewise not performed or not recorded. Table 1 presents the complete presenting examination classified into three categories that are deliberately kept distinct throughout this report: findings documented as present, findings documented as absent, and findings never assessed. The distinction between the second and third categories is not pedantic. A recorded absence is evidence; an unrecorded item is not, and

treating the two as equivalent is the single most common source of error when historical records are scored retrospectively, as detailed in Table 1. The initial differential diagnosis entertained by the admitting team was an orbital apex lesion or an intracranial space-occupying process.

Structural imaging of the brain and orbits

Contrast-enhanced computed tomography of the brain and orbits was performed on 31 August 2020. There was no intracranial mass, no cerebral infarction, no haemorrhage and no pathological contrast enhancement. The globes, retrobulbar tissues, optic nerves, extraocular muscles, orbital walls, orbital apices and lacrimal glands were all reported as unremarkable. Two incidental abnormalities were noted: enlargement of the cisterna magna, reported as compatible with either a mega cisterna magna or a retrocerebellar arachnoid cyst, and inflammatory change in several paranasal sinuses. Neither adequately explained the combination of an optic neuropathy and a complete ophthalmoplegia, and neither was situated in the orbital apex or cavernous sinus. Figure 1 reproduces the four representative sections from that study.

As shown in Figure 1, the study was comprehensively negative for the structural pathology it was designed to detect. Panel A, at the orbital level, demonstrates symmetrical globes with normal retrobulbar fat planes, extraocular muscles and orbital apices, without the muscle enlargement of thyroid eye disease, the soft-tissue infiltration of an inflammatory pseudotumour or the bone destruction of an invasive process. Panel B, at the posterior fossa level, shows no infarct or haemorrhage and displays the enlarged retrocerebellar cerebrospinal fluid space, which is seen to best advantage on the midline sagittal reformat in panel C. Panel D, at the ventricular level, confirms normal ventricular calibre without parenchymal abnormality. The important negative in Figure 1 is the absence of any cavernous sinus or orbital apex lesion, since a mass, an inflammatory infiltrate or a thrombosed cavernous sinus at that site would readily have accounted for the entire clinical syndrome.

Table 1. Presenting clinical and neuro-ophthalmological findings at the August 2020 evaluation, classified as documented present, documented absent, or not assessed in the available record.

Domain	Finding	Status	Detail as recorded
Constitutional	Fever, weight loss, night sweats	ABSENT	Explicitly recorded as absent
Constitutional	Arm or leg claudication	ABSENT	Explicitly recorded as absent
Vascular examination	Inter-arm systolic blood pressure difference	ABSENT	No clinically significant difference identified
Vascular examination	Bilateral peripheral pulse assessment	NOT ASSESSED	Not documented in the available record
Vascular examination	Carotid pulse and carotid tenderness	NOT ASSESSED	Not documented in the available record
Vascular examination	Auscultation for arterial bruit	NOT ASSESSED	Not documented in the available record
Neurological	Level of consciousness	PRESENT	Glasgow Coma Scale E4V5M6
Neurological	Headache	PRESENT	Chronic progressive right hemicranial headache, recently worse
Cranial nerve	Optic nerve (CN II) dysfunction, right	PRESENT	Complete loss of vision in the right eye
Cranial nerve	Oculomotor, trochlear and abducens (CN III, IV, VI), right	PRESENT	Complete right ophthalmoplegia
Ophthalmic	Ptosis, right	PRESENT	Right eye closed on presentation
Ophthalmic	Proptosis, right	PRESENT	Recorded on inspection
Ophthalmic	Pupillary light reflexes, right	PRESENT	Absent
Ophthalmic	Snellen or logMAR visual acuity	NOT ASSESSED	No formal acuity recorded
Ophthalmic	Fundoscopy, OCT, fluorescein angiography, visual fields	NOT ASSESSED	None performed or documented

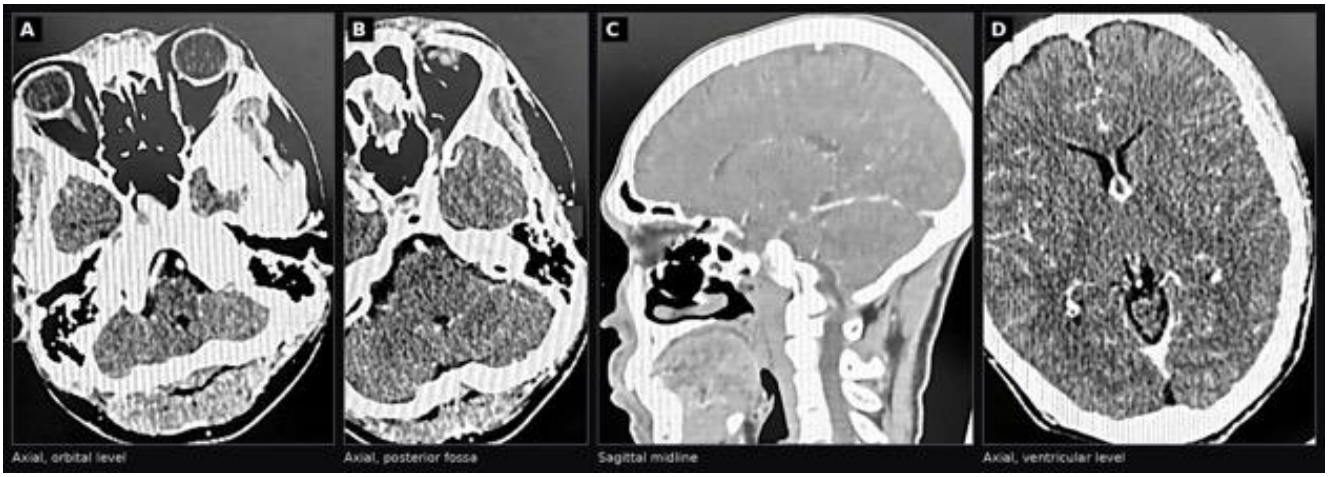

Figure 1. Contrast-enhanced computed tomography of the brain and orbits performed on 31 August 2020. (A) Axial image at the level of the orbits showing symmetrical globes, retrobulbar fat, extraocular muscles, optic nerves and orbital apices without mass, infiltration or abnormal enhancement. (B) Axial image at the level of the posterior fossa showing no infarct or haemorrhage and an enlarged retrocerebellar cerebrospinal fluid space. (C) Midline sagittal reformat demonstrating the enlarged cisterna magna, reported as either a mega cisterna magna or a retrocerebellar arachnoid cyst. (D) Axial image at the ventricular level showing normal ventricular calibre and no parenchymal lesion. No finding on this study accounted for the combined optic neuropathy and complete ophthalmoplegia.

Computed tomography angiography of the cervicocerebral arteries

Computed tomography angiography was performed on 7 September 2020, seven days after the structural study. It demonstrated irregularity and marked luminal narrowing of the right common carotid artery, extending continuously into the cervical, petrous, cavernous and supraclinoid segments of the right internal carotid artery. The left internal carotid artery, the anterior and middle cerebral arteries bilaterally, both vertebral arteries, the basilar artery and both posterior cerebral arteries were preserved, without significant plaque, stenosis, aneurysm or arteriovenous malformation. A small tubular hyperdense focus in the right temporal region was considered to represent vascular calcification. The volume-rendered reconstructions from this acquisition are reproduced in Figure 2. The erythrocyte sedimentation rate was reported as elevated; the numerical value was not recorded in the material available for this report, and no C-reactive protein result was documented.

Figure 2 illustrates the volume-rendered reconstructions from this acquisition. In panel A the asymmetry between the two carotid axes is immediately apparent: one side is represented by a sparse, faint and irregularly opacified vascular tree, while the contralateral axis and its branches are densely and continuously opacified. Panel B reproduces the same asymmetry at the intracranial level, where the affected carotid siphon and its distal branches are thin and poorly attenuating against a broad, brightly opacified contralateral siphon and middle cerebral artery. On the axial source images the attenuated axis corresponded to the right common and internal carotid arteries. The side-to-side difference in calibre and opacification is demonstrated in Figure 2 without any added annotation. The reconstructions in Figure 2 are deliberately presented without superimposed arrows or laterality markers so that the reader can judge the difference in opacification directly rather than through the authors' annotation.

Table 2 maps the imaging findings onto the nine arterial territories that the 2022 American College of Rheumatology/EULAR criteria use to score anatomical extent. One territory, the right carotid, was demonstrably involved. One, the left carotid, was demonstrably spared. The remaining seven were either not covered by the acquisition or were assessed only by a modality incapable of excluding arteritis. The territory-by-territory status of each vessel, together with the modality actually applied to it, is detailed in Table 2. This distribution is the anatomical counterpart of the clinical pattern in Table 1, and the two together define the boundary of what can honestly be asserted about the extent of disease in this patient.

Figure 2. Three-dimensional volume-rendered reconstructions from the computed tomography angiogram of 7 September 2020. (A) Volume-rendered projection of the cervicocerebral arterial tree showing marked side-to-side asymmetry, with a sparse, faint and irregularly opacified carotid axis on one side contrasted against a densely opacified contralateral axis and its branch tree. (B) Intracranial projection in which the affected carotid axis and its distal branches are thin and poorly attenuating, whereas the contralateral carotid siphon and middle cerebral artery are broad and brightly opacified. On the axial source images the attenuated axis corresponded to the right common and internal carotid arteries. Both reconstructions are shown as acquired, without added arrows or labels, so that the asymmetry of opacification can be judged directly.

Table 2. Arterial territory map derived from the 7 September 2020 computed tomography angiogram and the 18 September 2020 abdominal ultrasonogram, mapped onto the nine arterial territories used by the 2022 American College of Rheumatology/EULAR classification criteria.

Arterial territory	Modality applied	Finding	Counted as involved
Right carotid	CT angiography	Long-segment irregularity and marked luminal narrowing of the common carotid artery continuing into the cervical, petrous, cavernous and supraclinoid internal carotid segments	YES
Left carotid	CT angiography	Preserved; no significant plaque, stenosis, aneurysm or malformation	NO
Right subclavian	Not imaged	Not covered by the acquisition	NOT ASSESSED
Left subclavian	Not imaged	Not covered by the acquisition	NOT ASSESSED
Thoracic aorta	Not imaged	Not covered by the acquisition	NOT ASSESSED
Abdominal aorta	Ultrasonography only	No structural abnormality identified; wall thickness not measured; planned CT angiography not performed	NOT ASSESSED
Mesenteric	Ultrasonography only	No abnormality identified	NOT ASSESSED
Right renal	Ultrasonography only	No abnormality identified; arterial calibre and velocity not measured	NOT ASSESSED
Left renal	Ultrasonography only	No abnormality identified; arterial calibre and velocity not measured	NOT ASSESSED
Intracranial anterior circulation	CT angiography	Bilateral anterior and middle cerebral arteries preserved	Not a scored territory
Posterior circulation	CT angiography	Vertebral, basilar and posterior cerebral arteries preserved	Not a scored territory

Territories counted as involved: 1 of 9. CT, computed tomography.

Extracranial survey and laboratory evaluation

Abdominal ultrasonography was performed on 18 September 2020 as part of the evaluation for large-vessel vasculitis. The study was reported as showing mild hepatomegaly and minimal ascites, with no significant renal, para-aortic or other abdominal structural abnormality. Figure 3 shows the relevant panels. Panel A demonstrates the right hepatic lobe with the calliper trace used for biometry; the two recorded measurements were 14.25 cm and 11.18 cm. It should be noted openly that a longest recorded hepatic dimension of 14.25 cm lies within the range conventionally accepted as normal for the right lobe in an adult man, so the descriptive impression of mild hepatomegaly was not corroborated by the recorded biometry; the discrepancy is reported here as it appears in the source record rather than silently reconciled. Panel B shows the gallbladder and adjacent liver with a 0.35 cm calliper measurement recorded on the study. Panel C shows the portal vein interrogated with colour Doppler, with unobstructed hepatopetal flow and no evidence of portal hypertension. Panel D shows the para-aortic region, labelled by the operator as proximal and distal, in which no lymphadenopathy or focal abnormality was identified.

The limitation of the survey shown in Figure 3 is methodological rather than technical. B-mode ultrasonography of the para-aortic region evaluates the soft tissues adjacent to the aorta; it does not measure aortic wall thickness, does not interrogate renal arterial velocities and cannot exclude mural inflammation or ostial renal artery stenosis. Recorded as normal in Table 2, these territories are therefore better described as unassessed than as uninvolved. Complete angiographic evaluation of the aorta and its major branches, dedicated arterial wall imaging, fluorodeoxyglucose positron emission tomography, catheter angiography and histopathological examination were not performed at any point. What the abdominal survey did and did not cover is documented in Figure 3.

Diagnostic assessment and classification

Takayasu arteritis was diagnosed on the basis of the patient's age, the elevated inflammatory marker, the neuro-ophthalmic ischaemic syndrome and the long-segment stenotic involvement of the right common and internal carotid arteries in the absence of convincing atherosclerotic disease. The morphology and distribution of the lesion were considered compatible with large-vessel vasculitis and could not be attributed to an intracranial mass, a primary orbital disorder or an isolated cranial neuropathy, as Figure 1 and Figure 2 together demonstrate. The documented lesions involved the right common carotid artery and its internal carotid continuation, a distribution compatible with Hata angiographic type I disease; because imaging of the entire aorta and its major branches was not obtained, clinically silent involvement elsewhere could not be excluded, and the angiographic type assignment is therefore provisional.

Table 3 presents the 2022 American College of Rheumatology/EULAR criteria applied item by item to the documented record. Both absolute requirements were met: the patient was 36 years old at diagnosis, and vasculitis was evidenced on cross-sectional angiography. Of the seven additional clinical items, one was not met because the patient is male, two were documented as absent, one was not documented at all, and three of the highest-weighted items, each worth two points, were never assessed. Of the three imaging items, one territory of arterial involvement was demonstrated, yielding a single point; symmetric involvement of paired arteries was not present on the imaging performed; and the three-point item for abdominal aortic involvement with renal or mesenteric disease could not be scored because the relevant angiography was never obtained. The total was therefore one point against a classification threshold of five. As set out in Table 3, had any two of the three unassessed two-point examination items been positive and recorded, the score would have reached exactly five. The diagnosis in this patient rests on clinical and angiographic judgement, and the criteria are reported here for transparency rather than as corroboration. The alternative diagnoses that were formally considered, and the evidence for and against each of them, are set out in Table 4.

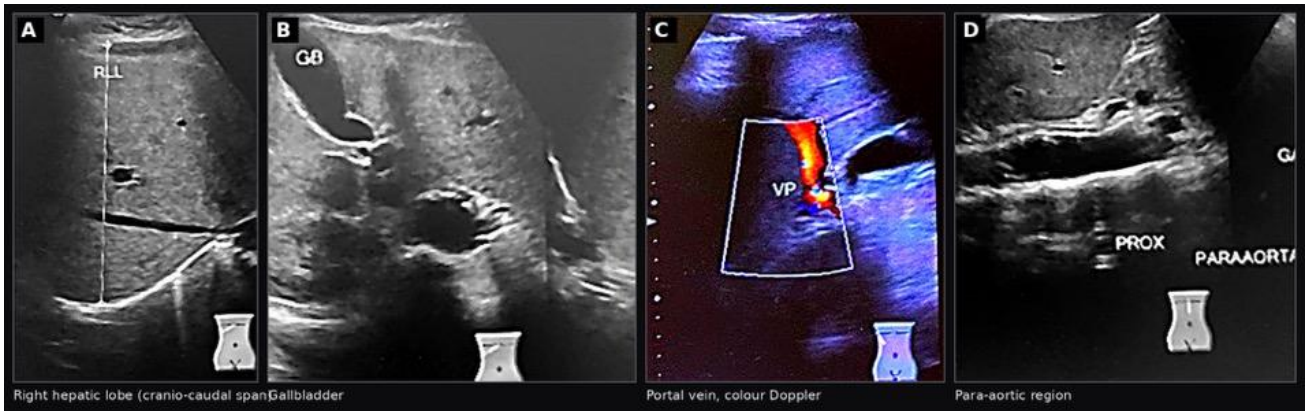

Figure 3. Abdominal ultrasonography performed on 18 September 2020 during the survey for extracranial large-vessel involvement. (A) Right hepatic lobe with the recorded calliper trace; the two recorded measurements were 14.25 cm and 11.18 cm. (B) Gallbladder and adjacent liver, with a 0.35 cm calliper measurement recorded on the study. (C) Portal vein interrogated with colour Doppler, showing unobstructed hepatopetal flow. (D) Para-aortic region, labelled by the operator as proximal and distal, in which no lymphadenopathy or focal abnormality was identified. Ultrasonography of this kind surveys the para-aortic soft tissues but does not measure aortic wall thickness and cannot exclude aortic or renal arterial involvement.

Table 3. The 2022 American College of Rheumatology/EULAR classification criteria for Takayasu arteritis applied retrospectively to the documented record of this patient.

Criterion	Weight	Status in this patient	Score
Absolute requirement: age 60 years or younger at diagnosis	Required	Met; 36 years at diagnosis	-
Absolute requirement: evidence of vasculitis on imaging	Required	Met; CT angiography of the cervicocerebral arteries	-
Female sex	+1	Not met; the patient is male	0
Angina or ischaemic cardiac pain	+2	Not documented	0
Arm or leg claudication	+2	Documented absent	0
Vascular bruit	+2	NOT ASSESSED	0
Reduced pulse in upper extremity	+2	NOT ASSESSED	0
Carotid artery abnormality (reduced pulse or tenderness)	+2	NOT ASSESSED	0
Systolic blood pressure difference between arms of 20 mmHg or more	+1	Documented absent	0
Number of affected arterial territories: one territory	+1	Met; right carotid only	+1
Symmetric involvement of paired arteries	+1	Not met; disease was unilateral on the imaging performed	0
Abdominal aorta involvement with renal or mesenteric involvement	+3	NOT ASSESSED; abdominal aortic angiography never performed	0
TOTAL	-	Threshold for classification is 5 points or more	1

Three examination items worth +2 each were never recorded. Had any two of them been positive and documented, the score would have reached the classification threshold of 5. CT, computed tomography.

Table 4. Differential diagnosis of long-segment unilateral carotid narrowing in a young adult, with the evidence for and against each alternative in this patient.

Alternative diagnosis	Features that would support it	Assessment in this patient
Cervical artery dissection	Abrupt onset, neck pain, intramural haematoma, tapered flame-shaped occlusion, Horner syndrome	Course was chronic and progressive over months; no history of trauma; no dedicated vessel-wall imaging was performed, so dissection was not formally excluded
Fibromuscular dysplasia	Beaded string-of-beads mid-to-distal cervical segments, often bilateral, frequently renal involvement	The lesion was a continuous long-segment stenosis rather than beaded, and extended intracranially; renal arteries were not formally imaged
Premature atherosclerosis	Focal bifurcation plaque, calcification, conventional vascular risk factors	One small tubular hyperdense focus in the right temporal region was considered vascular calcification; the diffuse continuous pattern and the patient's age favour arteritis
Moyamoya-spectrum arteriopathy	Progressive supraclinoid internal carotid stenosis with basal collateral network	Supraclinoid involvement was present, but there was no basal collateral network and disease began proximally in the common carotid artery
Giant cell arteritis	Age above 50 years, jaw claudication, temporal artery abnormality, polymyalgic symptoms	Markedly atypical at 36 years; none of the supporting features documented
Behcet disease with vascular involvement	Recurrent oral and genital ulceration, uveitis, venous thrombosis, pulmonary artery aneurysm	No mucocutaneous, ocular inflammatory or thrombotic features documented
Infection-associated arteritis (tuberculosis, syphilis)	Constitutional symptoms, positive serology or microbiology, basal meningeal enhancement	No constitutional symptoms; no pathological meningeal enhancement on contrast CT; targeted serology not documented
IgG4-related disease	Periarterial soft-tissue cuffing, elevated serum IgG4, other organ involvement	No periarterial soft-tissue mass described; serum IgG4 not measured
Radiation-induced arteriopathy	Prior head and neck irradiation in the affected field	No history of irradiation

Therapeutic intervention and early follow-up

The patient received high-dose systemic glucocorticoid therapy during the initial phase, followed by azathioprine as a glucocorticoid-sparing immunosuppressant and by antiplatelet therapy. The exact glucocorticoid induction dose, route of administration, duration and tapering schedule, the initial azathioprine dose and the timing of antiplatelet initiation were not documented in the available record. Following treatment the headache improved substantially and ultimately resolved. The complete loss of vision in the right eye persisted without any recovery, indicating that the neuro-ophthalmic injury sustained before treatment was already irreversible. During outpatient review on 24 September and 16 November 2020 the diagnosis was maintained. By November 2020 the patient reported no active symptoms, the blood pressure was 120/70 mmHg, and computed tomography angiography of the abdominal aorta was planned; the result of that study does not appear in any subsequent record.

Figure 4 presents the full clinical course as a timeline. The seven days separating the uninformative structural computed tomography of 31 August 2020 from the diagnostic angiographic study of 7 September 2020 is the interval in which the working hypothesis moved from a structural orbital or intracranial lesion to a vascular one, and it is the shortest interval in the whole record with any prognostic significance. The available documentation does not identify which clinician initiated that change of hypothesis, and no attribution is claimed here.

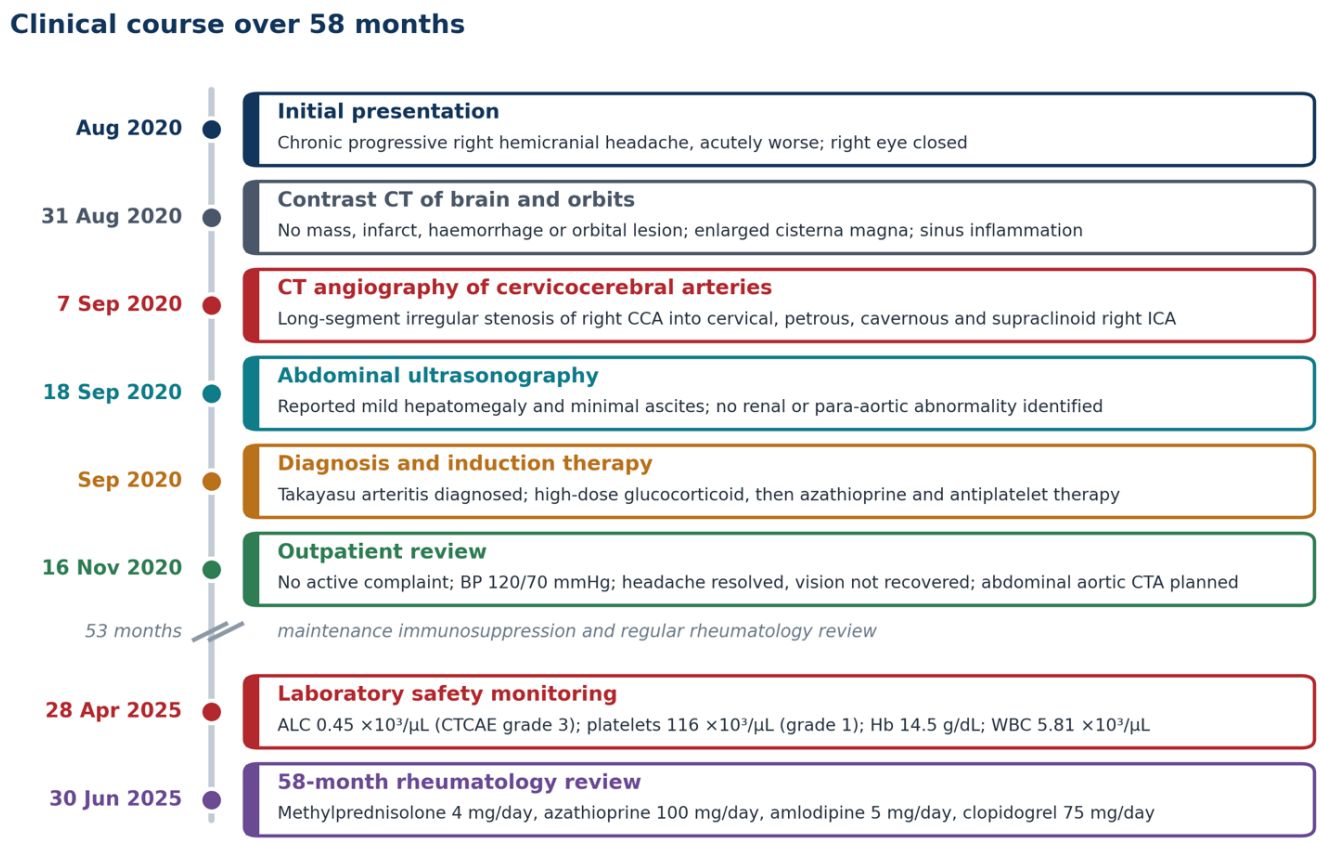

Figure 4. Clinical timeline of the 58 months between the first neuro-ophthalmic presentation in August 2020 and the most recent rheumatology review on 30 June 2025. The seven days that separated the unrevealing brain and orbital computed tomography from the diagnostic computed tomography angiogram are the interval in which the diagnostic pathway was redirected from a structural to a vascular hypothesis. ALC, absolute lymphocyte count; BP, blood pressure; CCA, common carotid artery; CTA, computed tomography angiography; CTCAE, Common Terminology Criteria for Adverse Events; Hb, haemoglobin; ICA, internal carotid artery; WBC, white blood cell count.

Long-term follow-up and safety surveillance

The patient remained under regular rheumatology follow-up for 58 months. In June 2025 his maintenance regimen comprised methylprednisolone 4 mg once daily, azathioprine 50 mg twice daily (100 mg per day), amlodipine 5 mg once daily and clopidogrel 75 mg once daily. Aspirin 80 mg daily had previously been discontinued following a fall in the platelet count, and clopidogrel was subsequently substituted after specialist evaluation. Hypertension remained controlled. A complete blood count obtained on 28 April 2025 showed a white blood cell count of 5.81 × 10^3/µL, an absolute lymphocyte count of 0.45 × 10^3/µL, a haemoglobin concentration of 14.5 g/dL and a platelet count of 116 × 10^3/µL. Serial complete blood count monitoring at two-month intervals was planned. At the most recent review the patient was clinically stable and reported no new symptoms suggestive of active disease. As detailed in Figure 4, the interval between the last documented outpatient contact of the diagnostic era and the 2025 laboratory panel was 53 months.

Table 5 lists the laboratory, pharmacological and vascular status at the 58-month review with formal toxicity grading. The absolute lymphocyte count of 0.45 × 10^3/µL corresponds to grade 3 lymphopenia by the Common Terminology Criteria for Adverse Events version 5.0 and represents 7.7% of the total leucocyte count, which was itself normal; the derivation is 0.45 ÷ 5.81 × 100 = 7.7%. The platelet count of 116 × 10^3/µL corresponds to grade 1 thrombocytopenia. The haemoglobin concentration and the total leucocyte count were both normal, so the cytopenias were lineage-selective rather than a global marrow effect. The methylprednisolone dose of 4 mg per day is equivalent to approximately 5 mg per day of prednisolone. Body weight was not recorded at any point, so the azathioprine dose cannot be expressed in mg/kg, and thiopurine S-methyltransferase and nudix hydrolase 15 genotyping had not been performed.

3. Discussion

A neuro-ophthalmic presentation without classical peripheral vascular signs

The diagnostic difficulty in this case was not the rarity of Takayasu arteritis but the near-total absence of the findings that ordinarily prompt clinicians to think of it. There was no fever, no weight loss, no claudication and no inter-arm blood pressure difference, and the three physical-examination findings most closely associated with the disease, namely a reduced peripheral pulse, an abnormal carotid pulse or carotid tenderness, and an arterial bruit, were never sought or at least never recorded, as detailed in Table 1. A pre-pulseless phase is a recognised and frequent stage of the disease rather than an anomaly: 91 of 238 patients with angiographically proven Takayasu arteritis in one cohort were classified as pre-pulseless, and these patients had a distinct clinical and angiographic profile while their outcomes were broadly similar to those of pulseless patients.[17] The practical corollary is that the absence of pulse deficits carries almost no negative predictive value at the time when the diagnosis would be most useful.

What did point towards a vascular process was the anatomy of the deficit. An orbital apex syndrome implies simultaneous failure of the optic nerve and of all three ocular motor nerves, structures that share a confined anatomical space and, as discussed below, a shared arterial supply. In published series such a syndrome is usually structural: inflammation accounted for 54.54% and neoplasm for 18.18% of 66 cases in one series, with vascular causes making up only 3.03%,[18] and neoplasm led at 55% in a second series of twenty patients, in which, among the fourteen with paired acuity measurements, more than half remained legally blind and only 28.6% improved.[19] When dedicated structural imaging of that space is entirely normal, as it was here and as Figure 1 documents, the prior probability of the remaining categories shifts sharply, and a perfusion-based explanation becomes the most economical hypothesis. Male sex, far from arguing against Takayasu arteritis, should have raised the index of suspicion for a severe course, since male sex was an independent predictor of stroke with an odds ratio of 5.70 in a case-control study[11] and was associated with substantially higher mortality among patients with coronary involvement.[4] The timeline in Figure 4 shows that the correct hypothesis was reached within seven days of the negative structural study, which compares favourably with the median 13-week diagnostic delay reported in specialist paediatric practice,[8] although by then the visual loss was already complete.

What the classification criteria could and could not do here

Table 3 shows that this patient scores one point on the 2022 American College of Rheumatology/EULAR criteria against a threshold of five. It is worth being precise about what that means. The criteria were developed and validated to classify patients for research once a diagnosis of medium- or large-vessel vasculitis has already been made and mimics have been excluded; they perform impressively in that role, with a validation sensitivity of 93.8% and specificity of 99.2%.[5] Independent validation in 504 patients confirmed high sensitivity at 95.83% while showing that specificity in real-world comparator populations may be considerably lower at 63.51%,[6] and a large Chinese validation reported both sensitivity and specificity above 95%.[7] None of these studies addresses the situation encountered here, in which the score is depressed not by the biology of the disease but by the completeness of the record.

The arithmetic makes the point unusually clearly. Three of the seven clinical items each carry two points, and all three were never assessed. As Table 3 sets out, if any two of them had been positive and documented, the total would have risen from one to exactly five and the patient would have classified. A fourth pathway to the threshold existed on the imaging side: the item for abdominal aortic involvement with renal or mesenteric disease is worth three points, and the abdominal aortic angiogram that would have adjudicated it was planned in November 2020 and never performed. In other words, two brief bedside manoeuvres and one cross-sectional study stood between a score of one and a score of five or more. This is not an argument against the criteria; it is an argument for treating a low score derived from an incomplete record as uninformative rather than as evidence against the diagnosis, and for recording explicit negatives at the bedside so that later scoring is possible at all. The distinction between documented absence and non-assessment that structures Table 1 exists precisely for this purpose.

One lesion, three deficits: the arterial substrate

Figure 5 summarises the anatomical argument that unifies the presentation. The stenotic process was continuous from the right common carotid artery through the cervical, petrous and cavernous segments to the supraclinoid internal carotid artery. A lesion occupying that entire course lies upstream of three functionally distinct vascular beds at once. The first is the hemispheric anterior circulation, reduced flow through which offers a plausible substrate for a chronic progressive hemicranial headache. The second is the group of cavernous branches of the internal carotid artery. The third is the ophthalmic artery, which arises from the supraclinoid segment and supplies the optic nerve, retina and choroid. These three beds and the deficit each of them explains are mapped in Figure 5.

The cavernous branches deserve particular emphasis because they explain the feature of this case that structural imaging could not. In a cadaveric study of ten head specimens comprising twenty sides, the meningohypophyseal trunk was present in 100% of specimens and the inferolateral trunk in 85%; the oculomotor nerve was supplied by branches of the inferolateral trunk in all specimens, the proximal trochlear nerve received its supply from the superior branch of the inferolateral trunk in 75%, and the proximal third of the abducens nerve within Dorello canal was supplied exclusively by the dorsal meningeal artery arising from the meningohypophyseal trunk in 100% of specimens.[20] A more recent anatomical and surgical study found the inferolateral trunk on 93% (38 of 41) of sides, dividing into two branches in 21% (8 of 38) and, more commonly, three branches in 74% (28 of 38), which together supply cranial nerves III, IV, V1, V2, V3 and VI and the Gasserian ganglion.[21] A single stenotic lesion of the cavernous internal carotid artery is therefore positioned to compromise the arterial supply of all three ocular motor nerves simultaneously, which is exactly what a mass lesion at the orbital apex would otherwise be required to do mechanically. As Figure 5 shows, the same lesion extends far enough distally to involve the origin of the ophthalmic artery, completing the syndrome.

Two caveats must be stated before the inference, not after it. First, the same anatomical study that documents this arterial supply also reports that the inferolateral trunk can be sacrificed surgically without permanent cranial nerve deficit because of collateral supply,[21] which means that stenosis of the parent vessel is not by itself sufficient to produce neuropathy and that the collateral reserve of the individual patient matters. Second, no perfusion imaging, selective catheter angiography or vessel-wall study was performed in this patient, so the mechanism proposed in Figure 5 is an anatomical inference from a demonstrated lesion and a demonstrated deficit, not a measured haemodynamic finding. The clinical action that follows from it is nonetheless concrete: in a young adult with an orbital apex syndrome and normal structural imaging, the cavernous and supraclinoid internal carotid segments should be interrogated specifically, and the request to the radiologist should say so. That instruction follows directly from the arterial arrangement illustrated in Figure 5.

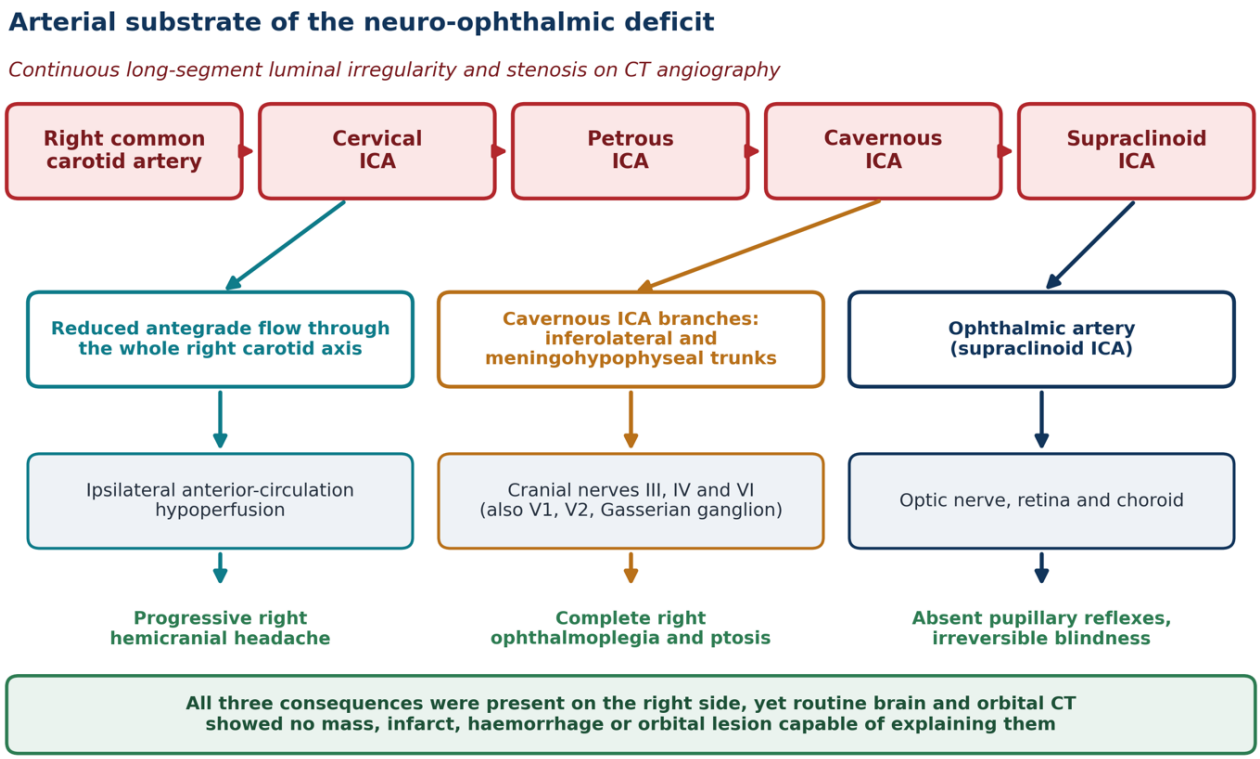

Figure 5. Proposed arterial substrate of the neuro-ophthalmic deficit. The stenotic process was continuous from the right common carotid artery to the supraclinoid internal carotid artery, so it lay upstream of three functionally distinct vascular beds simultaneously: the hemispheric anterior circulation, the cavernous branches that supply the ocular motor nerves, and the ophthalmic artery. This single-lesion explanation accounts for the coexistence of headache, complete ophthalmoplegia and optic neuropathy without requiring any mass, infarct or orbital lesion, none of which was present. The scheme is an anatomical inference rather than a demonstrated mechanism, because neither perfusion imaging nor selective catheter angiography was performed. ICA, internal carotid artery.

Ocular ischaemia and the irreversibility of the visual loss

The visual loss in this patient never recovered despite an inflammatory syndrome that otherwise responded well, and the distinction between controlling inflammation and reversing established ischaemic injury is central to counselling such patients. The published experience is consistent. In a series of patients with Takayasu arteritis and ocular involvement, acuity outcomes ranged from normal to no light perception and severe visual deterioration was common.[13] Arteritic anterior ischaemic optic neuropathy in this setting has been described as an ominous systemic sign, with one reported patient dying within three months of presentation despite maximal immunosuppression.[14] Where recovery does occur it is partial and unpredictable: a patient treated with endovascular intervention regained 6/18 in one eye after two months, while no acuity improvement was reported for the fellow eye, which had presented with no light perception.[16] A young woman with simultaneous stroke and anterior ischaemic optic neuropathy improved only from no light perception to counting fingers.[15]

The haemodynamic literature on carotid-related ocular ischaemia offers a mechanistic frame, provided its limitations are respected. Among 260 patients with ipsilateral internal carotid artery stenosis of 70% or more, hypoperfusion and reversed flow in the ophthalmic artery were associated with ocular ischaemic syndrome with an odds ratio of 3.536, and ocular ischaemic syndrome was 3.892 times more common with total occlusion than with severe stenosis.[22] That study was retrospective with pre-selected case and control groups, so it establishes association rather than a population incidence, and the authors themselves list sample size and ethnic specificity as limitations. In chronic carotid total occlusion of atherosclerotic origin, retrograde ophthalmic artery flow was present in 57.4% of patients compared with 8.3% of those with carotid stenosis, and eyes with retrograde flow showed lower retinal vessel length density in the superficial vascular complex (17.59 versus 18.48 mm^{-1}, p=0.049).[23] That cohort contained no patients with Takayasu arteritis and the design was cross-sectional, so the finding supports optical coherence tomography angiography metrics as candidate biomarkers of collateral-dependent perfusion and nothing stronger. Applied to the present case, this literature suggests that a fundus examination, optical coherence tomography and an assessment of ophthalmic artery flow direction at presentation would have been informative about the mechanism and about the fellow eye; none was performed, as Table 1 records, and the opportunity cannot be recovered retrospectively.

Differential diagnosis of long-segment unilateral carotid narrowing

Table 4 sets out the alternatives to Takayasu arteritis in a 36-year-old man with a continuous irregular stenosis extending from the common carotid artery to the supraclinoid internal carotid artery. Cervical artery dissection is the most important competitor in this age group, but the clinical course here was chronic and progressive over months rather than abrupt, there was no antecedent trauma or neck pain, and no Horner syndrome was described; it must nonetheless be conceded that no vessel-wall imaging was performed and the exclusion is therefore clinical rather than radiological. Fibromuscular dysplasia typically produces a beaded appearance in the mid and distal cervical segments and does not usually extend intracranially, whereas the lesion here was a continuous long-segment narrowing that began proximally in the common carotid artery. Premature atherosclerosis is argued against by the age, the diffuse rather than bifurcation-centred distribution, and the absence of documented vascular risk factors, notwithstanding the single tubular calcific focus

noted in the right temporal region. Moyamoya-spectrum arteriopathy involves the supraclinoid segment characteristically but is accompanied by a basal collateral network, which was not present, and does not begin in the common carotid artery. Each of these four alternatives is listed in Table 4 with the discriminating feature that argues against it.

As detailed in Table 4, the remaining alternatives are constrained mainly by what was not investigated. Giant cell arteritis would be markedly atypical at 36 years. Behcet disease was not supported by any mucocutaneous, ocular inflammatory or thrombotic feature in the record. Infection-associated arteritis, particularly tuberculous or syphilitic, is relevant in this geographical setting; there were no constitutional symptoms and no pathological meningeal enhancement on contrast-enhanced computed tomography, but targeted serological and microbiological testing was not documented, and this is a genuine residual uncertainty rather than a formality. IgG4-related disease can produce periarterial soft-tissue cuffing, which was not described, but serum IgG4 was not measured. Radiation arteriopathy was excluded by history. The complete list of alternatives, with the specific feature that would support each and the assessment actually available in this patient, is set out in Table 4. Supporting the working diagnosis are the age at onset, the continuous irregular stenotic morphology, the elevated erythrocyte sedimentation rate, the symptomatic response of the headache to anti-inflammatory therapy and the stable five-year course under immunosuppression. Isolated carotid vasculitis is itself a described entity with a substantial event rate: in a multicentre series the left internal carotid artery was most often involved at 39%, 81% of patients had stenosis, and 58% experienced a vascular event including stroke or transient ischaemic attack in 25%.[24] The distinction between isolated carotid vasculitis and Takayasu arteritis limited to one carotid territory is not resolvable without complete aortic imaging, which brings the discussion back to the gap identified in Table 2.

Adequacy and choice of vascular imaging

The decisive investigation in this case was computed tomography angiography, and its value lay in answering a question that the preceding structural study had not been designed to ask. Comparative work supports the diagnostic performance of cross-sectional angiography in this disease: in 36 patients evaluated with magnetic resonance and computed tomography angiography, imaging-defined disease activity correlated significantly with an elevated erythrocyte sedimentation rate and a positive C-reactive protein (p<0.0001).[25] Technique continues to improve; a prospective study of 53 patients with Takayasu arteritis, of mean age 33.8 plus or minus 10.2 years and of whom 49 were female, showed that dark-blood computed tomography angiography with deep learning reconstruction achieved significantly higher contrast-to-noise ratios than hybrid iterative reconstruction in all cervical arteries (all p<0.001), permitting direct assessment of the arterial wall rather than the lumen alone.[26] Had wall imaging of this quality been applied at presentation, the distinction between an inflammatory mural process and a dissection or a fibromuscular lesion, which Table 4 leaves open, might have been made non-invasively.

The corollary for surveillance is less intuitive. In a large multicentre cohort, radiological activity or progression was found in only 18 of the 418 imaging studies (4%) performed while the physician global assessment was inactive, whereas an active physician global assessment predicted radiological progression with an odds ratio of 31.766 (95% confidence interval 17.961–56.219, p<0.001).[27] Prospective longitudinal data point the same way: there was no change in angiographic findings over follow-up in 787 of 793 arterial territories, or 99%, that had no fluorodeoxyglucose positron emission tomography activity at baseline, and no angiographic progression occurred in patients who remained in clinical remission despite subclinical positron emission tomography activity at baseline.[28] Read together, these findings argue against reflex serial angiography in a clinically quiescent patient and in favour of symptom-driven and assessment-driven imaging. They do not, however, excuse the omission identified in Table 2, because the abdominal aorta and renal arteries in this patient were never characterised at baseline at all; the argument against repeating imaging in a quiescent patient presupposes that a baseline exists. The negative structural study reproduced in Figure 1 and the diagnostic angiogram reproduced in Figure 2 together illustrate that the modality must be matched to the hypothesis, and that a normal result excludes only what the technique was capable of detecting.

Two further considerations bear on how this patient should be followed. The first concerns the interpretation of acute-phase reactants. In a cohort of 113 patients of mixed racial background, of whom 51 were White and more than half of the non-White patients were Asian, White patients had substantially higher inflammatory markers at diagnosis than non-White patients, with a C-reactive protein of 33.6 mg/l versus 9.4 mg/l (p=0.033) and an erythrocyte sedimentation rate of 51 mm/h versus 24 mm/h (p=0.047).[29] An Indonesian patient whose only recorded inflammatory measure is a qualitative statement that the erythrocyte sedimentation rate was elevated therefore carries an unquantified and potentially modest inflammatory signal, and future assessments in this patient should record numerical values for both the erythrocyte sedimentation rate and C-reactive protein rather than a categorical impression. The second concerns a practical alternative to repeated cross-sectional angiography of the neck. In 68 patients with Takayasu arteritis contributing 120 common carotid arteries, compared against 120 control arteries, the residual inner diameter measured by ultrasound was the most reliable single parameter for grading stenosis, with areas under the receiver operating characteristic curve of 0.901 for stenosis of 50% or more and 0.969 for stenosis of 70% or more.[30] Ultrasound can also speak to activity and not only to calibre: in a cohort of 115 patients with carotid involvement, the specificity of extensive contrast-enhanced ultrasound enhancement for indicating active disease was 95% although sensitivity was only 67%, and 68% of patients whose active disease subsided after treatment showed a fall in enhancement.[31] A carotid duplex protocol reporting residual inner diameter would allow the affected axis in this patient to be followed without repeated radiation exposure, and would also supply, at last, the carotid examination findings whose absence is recorded in Table 1 and whose weight in Table 3 is two points.

Immunosuppression, disease control and arterial remodelling

The patient received high-dose glucocorticoid followed by azathioprine, which remains a reasonable conventional choice, although the comparative evidence is more nuanced than is often assumed. In a multicentre cohort of 301 patients in which 204 (67.8%) received methotrexate and 77 (25.6%) azathioprine as first-line steroid-sparing agent, remission, relapse, radiographic progression and adverse effect rates were similar between the two drugs, and drug survival did not

differ (at a median of 48 months, 32% versus 42%, p=0.34); however, the vascular surgery rate was significantly higher in the azathioprine group (23% versus 9%, p=0.001) and a greater proportion of methotrexate-treated patients were receiving 5 mg per day or less of glucocorticoid at the end of follow-up (76% versus 62%, p=0.034).[32] These are observational comparisons subject to confounding by indication, and they do not establish that azathioprine caused the excess vascular surgery. They do suggest that the choice is not neutral and that a patient maintained on azathioprine warrants attentive vascular follow-up. In this case, as Figure 4 shows, the clinical course over 58 months was stable, no new ischaemic event occurred, and the glucocorticoid dose was successfully reduced to a methylprednisolone equivalent of approximately 5 mg of prednisolone daily.

Whether medical therapy modifies the arterial lesion itself is a separate question. Serial computed tomography angiographic measurement across 75 treatment lines showed an overall reduction in arterial wall thickness in 73% of cases, with a relative decrease of more than 25% in 31%,[33] which indicates that mural inflammation is modifiable. Established luminal damage is a different matter, and where critical supra-aortic stenosis persists, revascularisation is feasible with acceptable risk: among 219 consecutive patients undergoing 265 endovascular procedures for 375 supra-aortic lesions, peri-operative neurological complications occurred in 8 of 219 cases (3.7%) and the rates of 30-day death and of stroke were both 0.5%.[34] In this patient no revascularisation was undertaken, and given that the visual loss was already complete and the headache had resolved, no clear target for intervention remained. The judgement would be different in a patient with progressive or fluctuating ipsilateral symptoms, and the possibility of ocular recovery after intervention, though limited, is documented.[16]

A documented but unaddressed safety signal: lymphopenia and thiopurine pharmacogenetics

The most actionable finding in the longitudinal record is one that appears in the notes without comment. As Table 5 details, the absolute lymphocyte count on 28 April 2025 was $0.45 \times 10^3/\mu L$, which is grade 3 lymphopenia by the Common Terminology Criteria for Adverse Events version 5.0, against a normal total white cell count of $5.81 \times 10^3/\mu L$, so that lymphocytes comprised 7.7% of circulating leucocytes. The platelet count of $116 \times 10^3/\mu L$ is grade 1 thrombocytopenia. Haemoglobin was normal at 14.5 g/dL. The pattern is lineage-selective rather than a global marrow suppression, which is characteristic of combined thiopurine and glucocorticoid exposure. All three counts, their reference intervals and their formal toxicity grades are listed in Table 5. Serious infection is not a theoretical concern in this population: of 238 patients with Takayasu arteritis, 38 (16%) developed serious infections across 50 episodes, three of which were fatal, with pneumonia (n=19) and tuberculosis (n=12) the commonest, a mean daily glucocorticoid dose of 13.03 (standard deviation 10.41) mg prednisolone equivalent across the 39 episodes that occurred while the patient was taking a corticosteroid, the other 11 having occurred off corticosteroid, and a mortality hazard ratio of 5.52 (95% confidence interval 1.75–17.39) associated with serious infection.[35] That cohort did not report lymphocyte counts, so it cannot be cited as evidence that lymphopenia specifically predicts infection in Takayasu arteritis; the inference here is that a patient carrying both a grade 3 lymphopenia and continuing glucocorticoid exposure sits in a population with a documented 16% serious infection rate and a fivefold mortality hazard when infection occurs.

The second omission is pharmacogenetic. Thiopurine S-methyltransferase and nudix hydrolase 15 genotyping had not been performed, as Table 5 records. Table 5 also records that no abdominal aortic imaging had been performed by the 58-month review. A genotype-first cohort study within a national bioresource found NUDT15 variant alleles in 2.0% (457 of 23,081) of participants overall but in 21.7% (10 of 46) of those of East Asian ancestry and 13.6% (151 of 1,113) of those of South Asian ancestry, against 1.3% (271 of 21,304) of Europeans; carriers of any NUDT15 variant experienced severe myelosuppression in 11.3% (27 of 239) compared with 1% (8 of 840) of wild-type controls, an odds ratio of 13.3 (95% confidence interval 6.2–31.6, p<0.001).[36] That study was conducted in a United Kingdom inflammatory bowel disease population and did not report a separate Southeast Asian ancestry group, so its allele frequencies cannot be transferred directly to an Indonesian patient. The direction of the finding is nonetheless clear: variant frequency rises steeply outside European ancestry, and the consequence of carriage is severe rather than trivial. The concrete recommendation arising from this case is that in an Indonesian patient established on long-term azathioprine who has developed a grade 3 lymphopenia, thiopurine pharmacogenetic testing should be requested, the dose should be reconsidered against body weight in mg/kg terms once weight is recorded, the two-monthly blood count schedule already planned should be adhered to, and prophylaxis against opportunistic infection should be discussed explicitly rather than left implicit. None of these steps requires new technology and all of them are absent from the record as it stands.

The antithrombotic history deserves a similar reading. Aspirin 80 mg daily was withdrawn because the platelet count fell, and clopidogrel 75 mg daily was substituted. The substitution is understandable but is not obviously protective: a grade 1 thrombocytopenia is a weak indication to stop one antiplatelet agent, and replacing it with another does not restore platelet number. Platelet indices in this disease are not only a safety readout but track activity: among 215 consecutively enrolled newly diagnosed patients the overall baseline plateletcrit was 0.32% (0.24–0.38), a high level was present in 43 of 215 (20.00%), and patients with a normal baseline plateletcrit were more likely to achieve complete remission (hazard ratio 4.65, 95% confidence interval 2.38–19.08, p<0.01).[37] Read against that literature, a falling platelet count in a patient who is otherwise quiescent is more likely to reflect drug effect than disease activity. The more useful question, which the record does not address, is whether the thrombocytopenia is a thiopurine effect, in which case dose adjustment guided by genotype and weight would address both the cytopenia and the lymphopenia at once. Antiplatelet therapy in Takayasu arteritis is individualised rather than universal, and the decision in a patient with established critical cranial arterial disease and a mild thrombocytopenia is a genuine trade-off that merits explicit documentation, as summarised in Table 5.

Table 5. Laboratory, pharmacological and vascular status at the 58-month review, with Common Terminology Criteria for Adverse Events version 5.0 grading of the haematological abnormalities.

Parameter	Value (28 April 2025)	Reference interval	CTCAE grade	Interpretation
White blood cell count	5.81 × 10^3/μL	4.0–10.0	0	Normal
Absolute lymphocyte count	0.45 × 10^3/μL	1.0–4.0	3	Grade 3 lymphopenia; 7.7% of the total leucocyte count
Haemoglobin	14.5 g/dL	13.0–17.0	0	Normal
Platelet count	116 × 10^3/μL	150–400	1	Grade 1 thrombocytopenia; aspirin previously withdrawn on this basis
Erythrocyte sedimentation rate	Elevated; value not recorded	-	-	Only inflammatory marker documented at diagnosis
Methylprednisolone	4 mg once daily	-	-	Approximately 5 mg/day prednisolone equivalent
Azathioprine	50 mg twice daily (100 mg/day)	-	-	Body weight not recorded, so mg/kg dosing cannot be derived
Amlodipine	5 mg once daily	-	-	Blood pressure controlled; 120/70 mmHg in November 2020
Clopidogrel	75 mg once daily	-	-	Replaced aspirin 80 mg/day after the platelet count fell
Thiopurine pharmacogenetics	Not performed	-	-	TPMT and NUDT15 genotype unknown
Abdominal aortic imaging	Not performed	-	-	Planned in November 2020; still outstanding at 58 months

CTCAE, Common Terminology Criteria for Adverse Events; NUDT15, nudix hydrolase 15; TPMT, thiopurine S-methyltransferase. Cells marked in bold red fall outside the reference interval. Reference intervals are those of the reporting laboratory as conventionally applied to adult men.

Unresolved vascular territories, hypertension and the abdominal aorta

Table 2 shows that seven of the nine scored arterial territories were never adequately assessed, and the omission is not neutral in a patient who requires an antihypertensive agent. As mapped in Table 2, only the right carotid was demonstrably involved and only the left carotid was demonstrably spared. Renal artery involvement is common in this disease: of 215 patients in a matched cohort, 117 (54.42%) had renal artery involvement, bilateral in 66 (56.41%), and although the mortality risk was not increased (hazard ratio 2.32, 95% confidence interval 0.61–8.78), morbidity was.[38-40] A separate analysis of the 567 patients registered in a large Chinese cohort confirmed renal artery stenosis in 172 of 567 (30.34%), with left renal artery involvement in 73 of 172 (42.44%).[39] Male patients specifically carry more of this burden, with systemic hypertension in 62.1% versus 42.4% and renal artery involvement in 62.7% versus 53.9% compared with women.[2] This patient is a hypertensive man with Takayasu arteritis whose renal arteries have been evaluated only by B-mode abdominal ultrasonography, a modality that, as Figure 3 illustrates, images the para-aortic soft tissues but neither measures aortic wall thickness nor interrogates renal arterial velocity. Attributing his hypertension to essential causes and treating it with amlodipine is therefore a provisional decision resting on an untested assumption.

The record contains the correct plan and not its execution. Computed tomography angiography of the abdominal aorta was planned in November 2020, as the timeline in Figure 4 records, and no result appears at any point in the subsequent 53 months. Beyond the clinical implications for blood pressure management, this single unperformed study is also what prevents the three-point classification item for abdominal aortic involvement with renal or mesenteric disease from being adjudicated, as Table 3 shows, and what leaves the angiographic type assignment provisional. The recommendation that follows is narrow and inexpensive: a single cross-sectional angiographic survey of the aorta and its major branches should be completed, both to establish the baseline that surveillance strategy presupposes[27] and to allow the hypertension to be attributed rather than assumed.

Strengths and limitations

The strengths of this report are the completeness of the imaging record at diagnosis, reproduced here rather than merely described in Figure 1, Figure 2 and Figure 3, and the duration of longitudinal follow-up, which allows the safety consequences of maintenance therapy to be observed rather than predicted. The limitations are substantial and are stated deliberately. The report is retrospective and depends on a clinical record that was not assembled with publication in mind, so the distinction between a finding that was absent and one that was never sought is preserved throughout Table 1 rather than resolved. No histopathological confirmation was obtained, and none of vessel-wall magnetic resonance imaging, fluorodeoxyglucose positron emission tomography, catheter angiography or cerebral perfusion imaging was performed, so the diagnosis is clinical and angiographic and the mechanism proposed in Figure 5 is an inference rather than a measurement. The ophthalmological evaluation was incomplete: no formal visual acuity, fundoscopy, optical coherence tomography, fluorescein angiography or visual field testing was recorded, which prevents the ocular injury from being classified as anterior ischaemic optic neuropathy, ocular ischaemic syndrome or retinal arterial occlusion. The unaffected fellow eye was likewise never formally assessed, so a subclinical contralateral abnormality cannot be excluded. Glucocorticoid dosing details, the tapering schedule and body weight were not documented, so exposure cannot be quantified and the azathioprine dose cannot be expressed in mg/kg. Inflammatory markers were limited to a single qualitative statement that the erythrocyte sedimentation rate was elevated, without a numerical value and without a C-reactive protein. The hepatic biometry discrepancy identified in Figure 3 is reported as found rather than reconciled. Finally, this is a single patient, and the mechanistic and management inferences drawn here require confirmation in series with

complete neuro-ophthalmological and vascular characterisation.

4. Conclusion

Takayasu arteritis can announce itself as an orbital apex syndrome, with chronic progressive hemicranial headache, complete ophthalmoplegia and irreversible monocular blindness, in a young man who has no pulse deficit, no inter-arm blood pressure difference, no constitutional symptoms and a wholly normal computed tomogram of the brain and orbits. In this patient the diagnosis turned on a single decision, taken seven days after the negative structural study, to image the arteries rather than the parenchyma. Computed tomography angiography then demonstrated a continuous long-segment stenosis running from the right common carotid artery to the supraclinoid internal carotid artery, a lesion positioned upstream of the hemispheric circulation, of the cavernous branches that supply the ocular motor nerves and of the ophthalmic artery simultaneously, and therefore capable of producing the entire syndrome without any mass, infarct or orbital lesion.

Three lessons generalise beyond this patient. First, the absence of classical peripheral vascular signs has little negative predictive value, because a pre-pulseless phase is a normal stage of the disease rather than an exception, and male sex should raise rather than lower concern about the severity of the course. Second, a validated classification instrument scored on an incomplete record returns a number that reflects documentation quality as much as biology: this patient scored one point of a required five, and two unrecorded bedside manoeuvres together with one unperformed cross-sectional study stood between that score and the threshold. Recording explicit negatives costs nothing at the time and cannot be recovered afterwards. Third, long-term care in this disease is not only about disease activity. A grade 3 lymphopenia and a grade 1 thrombocytopenia on maintenance azathioprine, an unassessed thiopurine genotype in a patient of Asian ancestry, an undocumented body weight that prevents weight-based dosing, and an abdominal aortic angiogram that was planned but never performed in a hypertensive patient are all correctable, and each of them represents a decision that has been deferred rather than made. Early vascular imaging may prevent further ischaemic injury, but once the optic nerve has failed the loss is permanent, and the remaining work of management is the systematic elimination of the avoidable risks that accumulate over the years that follow.

Declarations

Ethical approval and consent to participate. This report describes routine clinical care. Written informed consent for publication of this case report and the accompanying anonymised images was obtained from the patient. All identifying information has been removed from the clinical record and from every image reproduced here.

Consent for publication. Written informed consent for publication was obtained from the patient.

Availability of data and materials. All data supporting the findings of this report are contained within the article. Further anonymised detail is available from the corresponding author on reasonable request.

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