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Solitary Sinonasal Neurofibroma in an Elderly Male: A Rare Presentation and Surgical Management

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A B S T R A C T

Solitary sinonasal neurofibromas are rare peripheral nerve sheath tumors whose diagnosis is frequently confounded by non-specific symptoms and radiological features that mimic common inflammatory conditions. In the elderly, a unilateral sinonasal mass necessitates a high index of suspicion for neoplasia, yet diagnostic pitfalls remain a significant clinical challenge. A 65-year-old male with no stigmata of neurofibromatosis type 1 presented with a three-year history of progressively worsening unilateral nasal obstruction. Endoscopy revealed a large, pale, firm, non-friable mass. Computed tomography (CT) demonstrated an extensive, non-enhancing soft tissue mass originating in the left maxillary sinus, causing significant expansile bone remodeling and extending into multiple adjacent sinuses. The initial radiological impression was extensive sinonasal polyposis. However, an incisional biopsy followed by a comprehensive morphological analysis confirmed the diagnosis of a benign spindle cell tumor consistent with neurofibroma. The patient underwent complete tumor excision via a left lateral rhinotomy. The postoperative course was uneventful, with no recurrence at 12-month follow-up. In conclusion, this case underscores the critical importance of a thorough diagnostic workup for unilateral sinonasal masses in the elderly, where radiological findings can be misleading. Histopathological analysis is indispensable for the definitive diagnosis of spindle cell tumors in this location. For massive, maxillary-based neurofibromas with extensive lateral and anterior involvement, the lateral rhinotomy remains a vital and superior surgical approach, providing the necessary exposure to uphold the fundamental principle of complete oncologic resection and maximize the probability of a curative outcome.

1. Introduction

Neoplasms of the sinonasal tract, while accounting for only 3-5% of head and neck malignancies, encompass a remarkably diverse spectrum of pathologies. Among these, tumors of neurogenic origin are particularly infrequent, with benign peripheral nerve sheath tumors (PNSTs) like neurofibromas representing a true clinical rarity. A neurofibroma is a complex, unencapsulated tumor arising from the nerve sheath, composed of a heterogeneous population of Schwann cells, perineurial-like cells, fibroblasts, and mast cells embedded in a myxoid-collagenous matrix.[1] While they are a hallmark of the genetic disorder neurofibromatosis type 1 (NF1), the vast majority of sinonasal neurofibromas occur as solitary, sporadic lesions in individuals with no other systemic manifestations. The clinical course of a solitary sinonasal neurofibroma is notoriously

indolent.[2] Its slow, insidious growth means that symptoms, when they finally appear, are often vague and non-specific, primarily related to mass effect. Patients typically present with a long history of unilateral nasal obstruction, rhinorrhea, or intermittent epistaxis. This symptomatology creates a significant diagnostic challenge, as it closely mirrors that of far more common inflammatory conditions, such as chronic rhinosinusitis with nasal polyposis.[3] This diagnostic ambiguity is particularly pronounced as the clinical and even radiological features can closely mimic benign inflammatory processes, a pitfall this case report vividly illustrates.[4] Consequently, patients often experience substantial diagnostic delays, presenting only when the tumor has achieved a considerable size, causing significant bony remodeling and compressing adjacent critical structures.[5]

The diagnostic evaluation relies heavily on cross-sectional imaging and, ultimately, histopathological analysis. Computed tomography (CT) is excellent for delineating the associated bony changes, which are typically expansile rather than infiltrative.[6] Magnetic resonance imaging (MRI) is the standard of care for soft tissue characterization, helping to narrow the differential diagnosis and assess for potential perineural spread.[7] However, as this case demonstrates, even with imaging, a definitive preoperative diagnosis is often elusive. Therefore, tissue biopsy with comprehensive morphological analysis is not merely helpful but absolutely mandatory for an accurate diagnosis, as the features can overlap with a wide range of other benign and malignant spindle cell tumors.[8] The definitive treatment is complete surgical excision. Over the past two decades, the field of rhinology has been revolutionized by the advent of endoscopic endonasal surgery.[9] However, the enthusiasm for minimally invasive techniques must be balanced against the fundamental oncologic principle of complete tumor removal (R0 resection). While the literature contains reports of sinonasal neurofibromas, there is a lack of detailed analysis regarding the specific radiological pitfalls in elderly patients and a lack of robust debate on the limits of endoscopic surgery for maxillary-origin tumors with multi-sinus extension.[10]

The novelty of this report, therefore, lies in its critical analysis of the diagnostic labyrinth presented by a massive, solitary sinonasal neurofibroma that masqueraded as an inflammatory disease in an elderly patient. The aim of this study is to underscore the critical importance of maintaining a high index of suspicion for neoplasia in this clinical context, to dissect the misleading radiological features and demonstrate the indispensable role of histopathology, and to present a rigorous, evidence-based analysis reaffirming the indications for traditional open surgery in an era dominated by endoscopic techniques.

2. Case Presentation

A 65-year-old Balinese male was referred to our tertiary Otorhinolaryngology department with a three-year history of progressively worsening left-sided nasal obstruction. The patient, a farmer by occupation, presented with a clinical narrative classic for an indolent, slow-growing sinonasal mass, a journey meticulously detailed in Figure 1. His chief complaint, as highlighted, was a progressive and unremitting blockage of the left nasal passage, a symptom that had evolved from a minor nuisance to a significant impediment to his quality of life over a 36-month period. This extended timeline, graphically represented in the figure, underscores the insidious nature of the underlying pathology, allowing it to reach a substantial size before prompting the patient to seek specialized medical care. Complementing the primary complaint of obstruction was a secondary symptom of intermittent, self-limiting epistaxis. While not constant, these episodes of nasal bleeding represented a concerning feature in the patient's history, suggesting a lesion with some degree of vascularity or one causing significant pressure-related changes to the surrounding mucosa. The patient's history also revealed a crucial therapeutic detail: a 2-month course of empiric medical therapy, likely targeting a presumed diagnosis of inflammatory rhinitis or

sinusitis, had yielded no improvement. This failure of conservative management is a significant diagnostic clue, strongly suggesting that the etiology of his symptoms was not inflammatory but rather structural or neoplastic. The unresponsiveness to standard medical treatment served to differentiate his condition from more common sinonasal ailments and was a key factor in his eventual referral for a higher level of care. Equally informative were the pertinent negatives meticulously documented in his clinical history. A comprehensive review of his family history was negative for Neurofibromatosis type 1 (NF1), a critical piece of information that steered the differential diagnosis away from a syndromic cause and towards a sporadic, solitary lesion. This finding is vital, as the management and long-term surveillance for patients with NF1-associated tumors can differ significantly from those with isolated neoplasms. The absence of a genetic predisposition, as indicated in Figure 1, framed the diagnostic challenge as one of identifying a primary sinonasal pathology unique to the patient himself. The patient profile, clearly laid out, provides the demographic context for this clinical puzzle: an elderly male from a specific ethnic background whose occupation may or may not have contributed environmental factors in his condition, though no direct link was established. In essence, Figure 1 does more than just list facts; it constructs a compelling clinical narrative of a patient with a long-standing, progressively worsening unilateral nasal mass that was unresponsive to initial therapy and lacked any syndromic association, thereby setting the stage for the detailed diagnostic workup and surgical management that would follow. The graphical representation of these key data points provides a clear, concise, and scientifically informative summary that encapsulates the entirety of the patient's journey up to his presentation at the tertiary care facility, effectively highlighting the key features that would guide the subsequent clinical investigation. The figure serves as a visual abstract of the case's preamble, providing all the necessary information to understand the diagnostic and therapeutic challenges that lay ahead.

Figure 1. Patient demographics and clinical history.

On physical examination, the patient was in good general health. The external nasal framework and facial contours were symmetric, with no evidence of proptosis or palpable masses. However, internal examination revealed a significant unilateral pathology. A comprehensive assessment, visually summarized in Figure 2, confirmed that while the patient's systemic health was stable, a significant localized disease process was present within the sinonasal cavities. The general physical examination revealed a patient in good overall condition, with stable vital signs and a cooperative demeanor, findings that initially belied the extent of the internal pathology. The centerpiece of the clinical findings was the endoscopic view of the left nasal cavity, which was the site of the patient's primary complaint. As depicted in Figure 2, the examination revealed a complete obstruction of the nasal passage by a large, pale, whitish mass. The surface of the mass was noted to be smooth, a characteristic that can be associated with various submucosal or encapsulated lesions. Critically, upon gentle palpation with a probe, the mass was found to be firm and non-friable, and it did not bleed on contact. These features are of paramount diagnostic importance. They strongly argue against a diagnosis of simple inflammatory polyposis, which typically presents as soft, edematous, and often friable tissue. Furthermore, the lack of bleeding on contact made a highly vascular tumor, such as a juvenile nasopharyngeal angiofibroma or a hemangioma, less likely. This constellation of findings—a firm, pale, avascular-appearing solid mass—immediately raised the suspicion of a benign neoplastic process, such as a fibro-osseous lesion or, as was ultimately the case, a peripheral nerve sheath tumor. Examination of the contralateral and distal sites provided crucial context, as detailed in Figure 2. The right nasal cavity, though patent, was narrowed due to a significant deviation of the nasal septum. This was not an incidental finding but a direct consequence of the mass effect exerted by the large lesion on the left side, which was physically pushing the septum across the midline. This observation confirmed the significant size and pressure effect of the tumor. Further examination of the nasopharynx was revealing; it was clearly visualized and found to be free of pathology when viewed through the patient's right nasal cavity. However, the view from the left was completely obscured by the posterior extension of the mass. The oropharynx and larynx were unremarkable, indicating that the disease was confined to the upper sinonasal tract. A thorough neurological examination, with a particular focus on the cranial nerves innervating the face and sinonasal region, was performed to assess for any neurological compromise. As highlighted in Figure 2, all cranial nerves were found to be fully intact. Specific testing of the trigeminal nerve (CN V), which is responsible for facial sensation, and the facial nerve (CN VII), which controls the muscles of facial expression, revealed no deficits. The absence of any neurological signs, such as facial numbness, paresthesia, or weakness, was a significant finding. It suggested that despite its large size and the extensive bony remodeling it had caused, the tumor was not actively invading or infiltrating the major nerve trunks. This is a key feature that helps to differentiate slow-growing, expansile benign tumors from more aggressive, infiltrative malignant processes, which often present with early-onset neurological symptoms. In summary, Figure 2 provides a comprehensive and multi-faceted overview of the physical and endoscopic findings, painting a clear picture of a large, unilateral, solid, non-vascular sinonasal mass causing significant local mass effect but without evidence of neurological invasion, thereby steering the clinical diagnosis strongly towards a benign neoplasm.

A contrast-enhanced computed tomography (CT) scan of the paranasal sinuses was performed. The imaging revealed an extensive soft tissue mass with a low average density of approximately 25-35 Hounsfield Units (HU). The tumor demonstrated significant mass effect, causing smooth, expansile remodeling of the surrounding bony structures rather than aggressive, permeative destruction. Critically, the mass showed no significant internal or peripheral contrast enhancement.

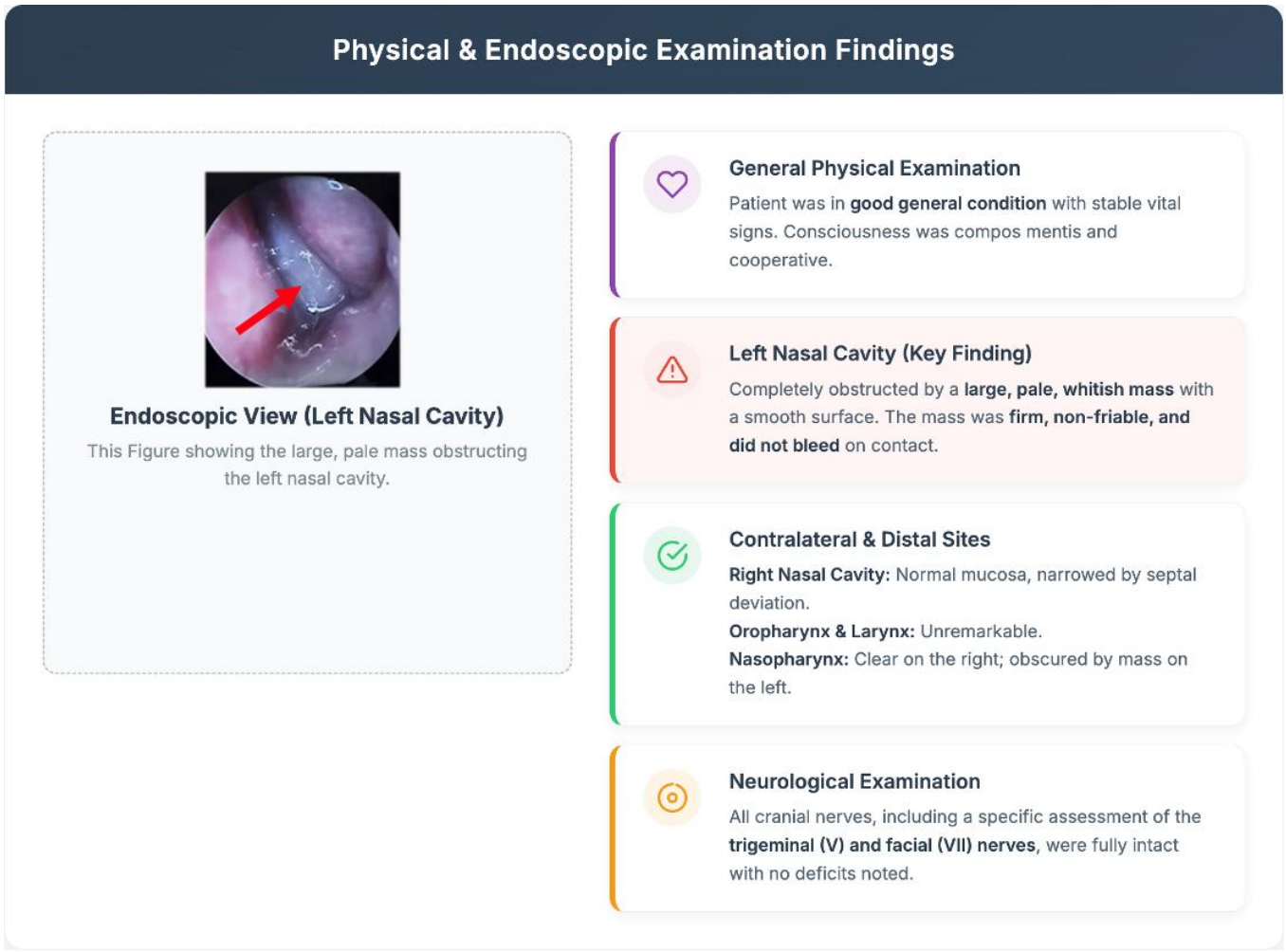

Figure 2. Physical & endoscopic examination findings.

While the skull base and orbital floors were intact, the tumor's extension into the sphenoid sinus brought it into close proximity to the carotid prominence, though a clear fat plane was maintained. The comprehensive radiological findings, summarized schematically in Figure 3, provide a detailed anatomical map of the pathology and offer crucial insights into its benign, slow-growing nature. The investigation into the tumor's origin and characteristics identified its epicenter within the left maxillary sinus, which was completely opacified by a large, homogeneous soft tissue mass. The lack of significant contrast enhancement is a pivotal finding, suggesting a lesion with low vascularity. This characteristic helps to differentiate it from highly vascular neoplasms, such as angiofibromas or certain malignancies, which would typically demonstrate avid enhancement. Furthermore, the low average density of 25-35 HU is consistent with a composition rich in myxoid or fluid content, a feature common in neurogenic tumors but also seen in inflammatory polyps, which foreshadows the diagnostic challenge to come. The sheer scale of the lesion is best understood through its extensive, multi-directional tumor extension, as detailed in Figure 3. The tumor's growth was not contained; it followed the path of least resistance, methodically expanding into contiguous spaces. Medially, it had eroded through the medial maxillary wall to completely fill the left nasal cavity, providing a direct anatomical explanation for the

patient's primary complaint of unilateral nasal obstruction. Superiorly, it extended into the left ethmoid sinuses, placing it in close proximity to the lamina papyracea and the floor of the orbit. Posteriorly, the mass extended beyond the choana into the nasopharynx, a finding that could potentially lead to Eustachian tube dysfunction or obstructive sleep apnea symptoms if left untreated. Perhaps most strikingly, the tumor demonstrated contralateral extension, crossing the midline by eroding the nasal septum. This dramatic finding, clearly visible on the coronal CT images presented in Figure 3, powerfully illustrates the immense pressure effect exerted by the mass over a prolonged period. The pattern of bony changes provides the most compelling radiological evidence of the tumor's benign etiology. Figure 3 explicitly describes "significant expansile remodeling with smooth, scalloped erosion." This is the radiological signature of a slow, indolent growth process. Unlike a malignant tumor that would typically infiltrate and destroy bone in a permeative, aggressive fashion with irregular margins, this lesion gently and persistently pushed against the bony confines of the sinuses. Over the years, this chronic pressure stimulated osteoclastic activity, causing the bone to gradually resorb and remodel around the expanding mass. The resulting smooth and scalloped edges are characteristic of a benign process that has grown in place for a long time, consistent with the patient's three-year clinical history. Finally, the section on the initial radiological impression reveals a critical teaching point. The tumor was initially misdiagnosed as "extensive sinonasal polyposis with secondary mucocele formation". This misinterpretation, as noted in Figure 3, represents a significant diagnostic pitfall. The overlap in features—sinus opacification, low density, and lack of enhancement—can easily lead to this conclusion. However, this initial impression failed to give appropriate weight to the key features arguing for a neoplasm: the strictly unilateral nature of the extensive disease and, most importantly, the frank, large-scale erosion of thick bone like the medial maxillary wall and nasal septum, which is highly atypical for simple inflammatory polyposis. This discrepancy between the initial impression and the subtle but critical neoplastic features underscores the importance of a meticulous and systematic approach to interpreting sinonasal imaging, particularly in cases of unilateral disease.

Given the stark incongruity between the solid endoscopic appearance and the radiological impression of polyposis, a diagnostic incisional biopsy was performed. This pivotal step shifted the investigation from the macroscopic world of imaging to the microscopic realm of cellular pathology, providing the definitive evidence required for an accurate diagnosis. The histopathological findings, meticulously detailed in Figure 4, resolved the clinical and radiological ambiguity and established the tumor's true identity. On gross examination, the tissue fragments obtained from the sinonasal mass presented as a firm, rubbery, grayish-white substance. This macroscopic appearance was the first piece of hard evidence that aligned with the endoscopic findings of a solid tumor, further distancing the diagnosis from that of edematous, translucent inflammatory polyps. The firm and rubbery consistency is characteristic of a lesion rich in stromal components, such as collagen and extracellular matrix, rather than one filled with inflammatory fluid. The microscopic description, however, provided the conclusive and defining features of the lesion. The analysis revealed a poorly circumscribed, hypocellular proliferation of bland spindle cells. The term "poorly circumscribed" is significant, as it reflects the unencapsulated and infiltrative growth pattern characteristic of a neurofibroma, which distinguishes it from its encapsulated cousin, the schwannoma. The proliferation was composed of "bland spindle cells," meaning the cells were uniform in size and shape, lacking the worrisome features of malignancy such as significant pleomorphism, hyperchromasia, or mitotic activity. Delving deeper into the cellular morphology, the nuclei were described as characteristically wavy or "serpentine" with scant cytoplasm. This serpentine

nuclear morphology is a classic, almost pathognomonic, feature of cells with Schwannian differentiation and is a hallmark of neurofibromas. The cells were arranged in intersecting fascicles within a distinctive background, or stroma, that was both myxoid (a pale, mucoid substance) and collagenous (containing fine collagen fibers). This mixed stroma is responsible for the tumor's unique consistency and its appearance on imaging. Furthermore, a key feature noted was the presence of numerous interspersed mast cells scattered throughout the lesion. While not exclusive to neurofibromas, the prominent presence of mast cells is a well-recognized and characteristic feature, reflecting the complex microenvironment of the tumor. Ultimately, the combination of these distinct histological features—the bland spindle cells with wavy nuclei, the unencapsulated growth, the mixed myxoid and collagenous stroma, and the presence of mast cells—allowed for a confident and unequivocal final diagnosis of benign neurofibroma.

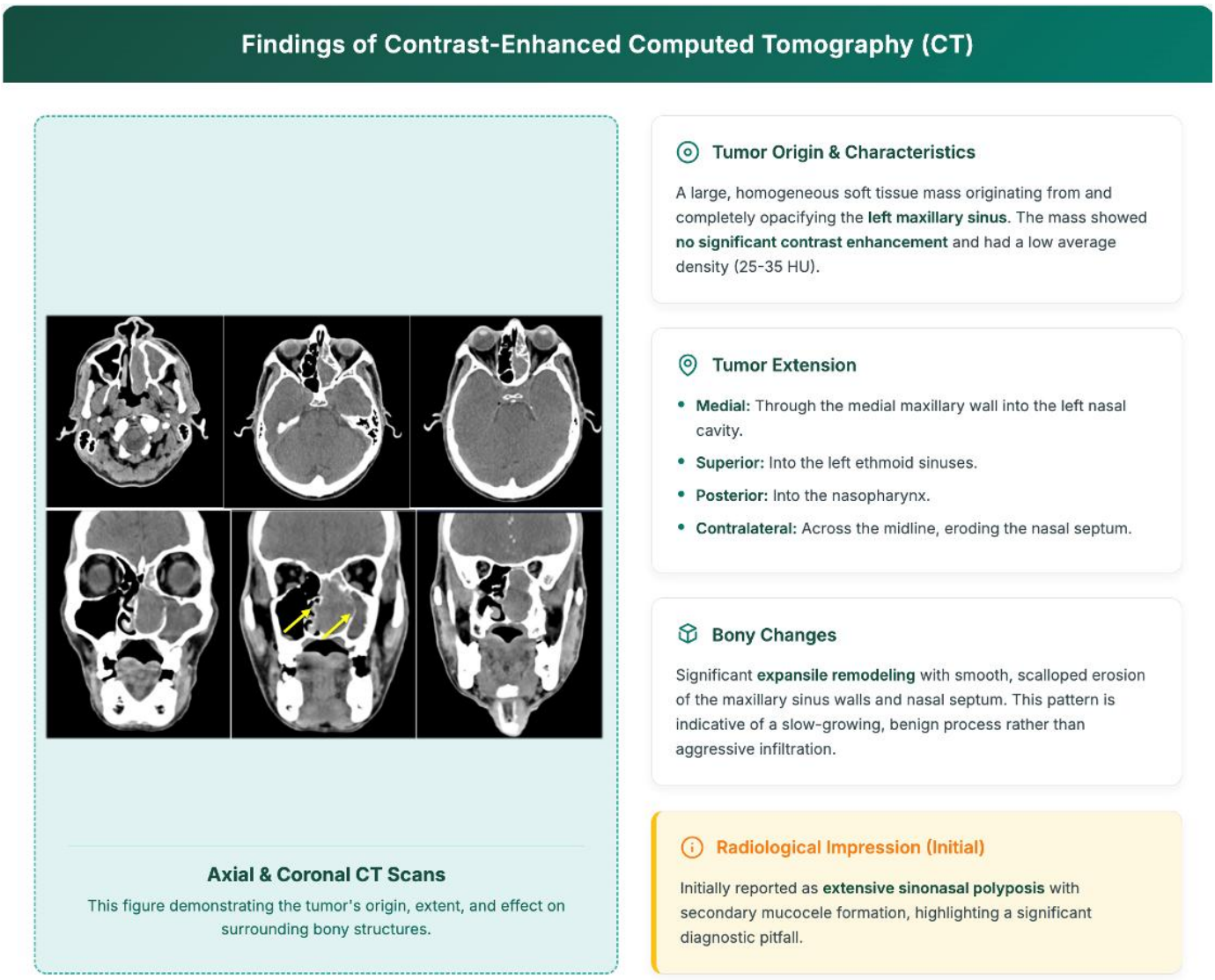

Figure 3. Findings of contrast-enhanced computed tomography (CT).

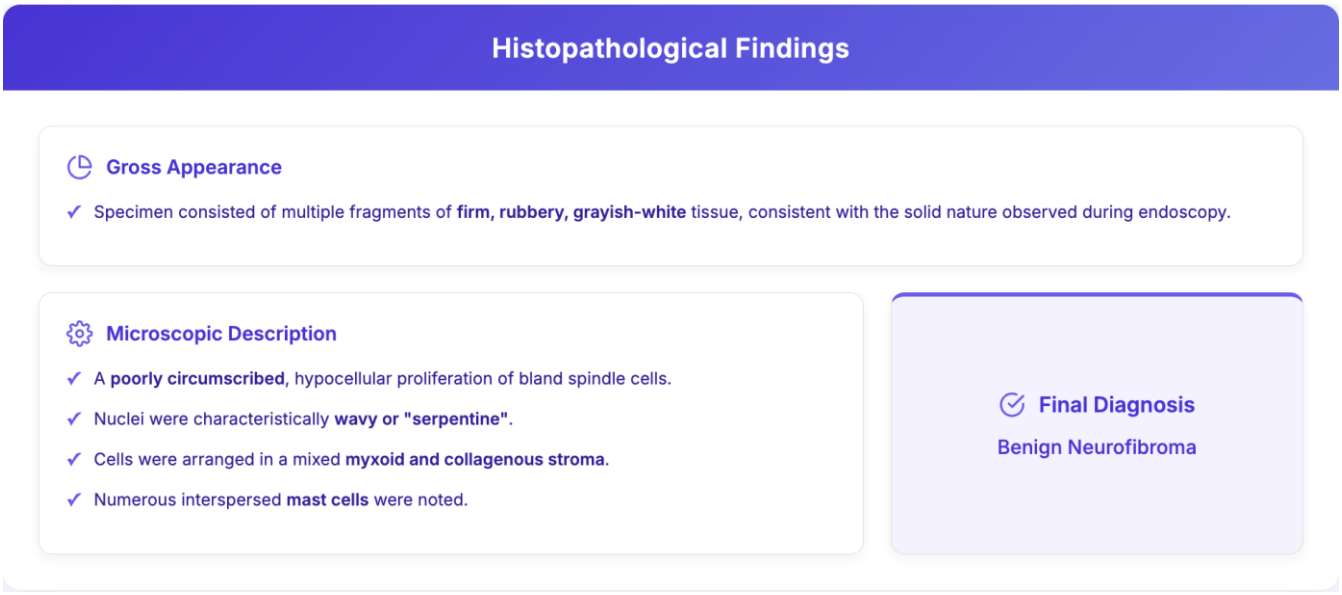

Figure 4. Histopathological findings.

Following the definitive diagnosis, a multidisciplinary discussion was held to determine the optimal surgical strategy. Based on the tumor's massive size, its origin within the maxillary sinus with diffuse involvement of the lateral and anterior walls, and the extensive multi-sinus extension, a left lateral rhinotomy via a Moure incision was selected. This decision, and its ultimate success, are comprehensively summarized in Figure 5. The chosen surgical approach represents a classic, time-tested technique for accessing the mid-facial skeleton. A left lateral rhinotomy involves a carefully placed incision along the side of the nose, which can be extended as needed (the Moure incision variant) to provide unparalleled exposure to the nasal cavity, maxillary sinus, and ethmoid sinuses. As stated in Figure 5, the primary goal of this approach was to achieve wide exposure. In an era increasingly dominated by minimally invasive endoscopic techniques, the selection of a traditional open procedure was a deliberate and critical choice. The preoperative imaging had demonstrated a tumor that not only filled the entire maxillary sinus but was broadly attached to its lateral and posterior walls—areas that are notoriously difficult to visualize and instrument through a purely endoscopic, transnasal corridor. The lateral rhinotomy effectively converts the closed "box" of the maxillary sinus into an open field, as suggested by the intraoperative view, allowing the surgeon to directly visualize the tumor's attachments and dissect it from surrounding structures under direct vision. The intraoperative findings, detailed in Figure 5, confirmed the wisdom of this approach. Upon entering the maxillary sinus, the surgical team confirmed that the tumor was as extensive as the CT scan had suggested, completely filling the left maxillary, ethmoid, sphenoid, and frontal sinuses. This finding underscored the significant challenge of the resection. The most crucial intraoperative discovery, however, was that the tumor's attachment was diffuse across the lateral and posterior walls of the maxillary sinus. This detail is paramount; it retrospectively validates the decision to forego an endoscopic approach. A tumor with a narrow pedicle or a medial attachment may be amenable to endoscopic removal, but a lesion with a broad, diffuse base on the far lateral wall cannot be safely and completely resected from a medial viewpoint. The ability to directly access and meticulously dissect this broad attachment is the unique advantage of the lateral rhinotomy. The

surgical team was able to meticulously follow the tumor into all its extensions, achieving what is noted as "macroscopic clearance (R0 resection)." This term signifies that, to the naked eye, all visible tumor was successfully removed, which is the single most important predictor of a successful outcome and the primary goal of any oncologic surgery. The success of the surgical strategy is ultimately measured by the final outcome, which was assessed at a 12-month follow-up and is highlighted as the concluding point in Figure 5. The patient experienced a complete resolution of all presenting symptoms, including the debilitating nasal obstruction and epistaxis. This clinical success was corroborated by endoscopic examination, which revealed a well-healed, widely patent sinonasal cavity with no evidence of tumor recurrence. This excellent outcome is the direct result of the successful R0 resection, which in turn was made possible by the selection of an appropriate surgical approach that provided the necessary exposure to address the tumor's specific anatomical challenges. In essence, Figure 5 masterfully connects the surgical plan, the operative reality, and the long-term patient benefit, providing a clear and compelling narrative of successful management for a complex and rare sinonasal tumor.

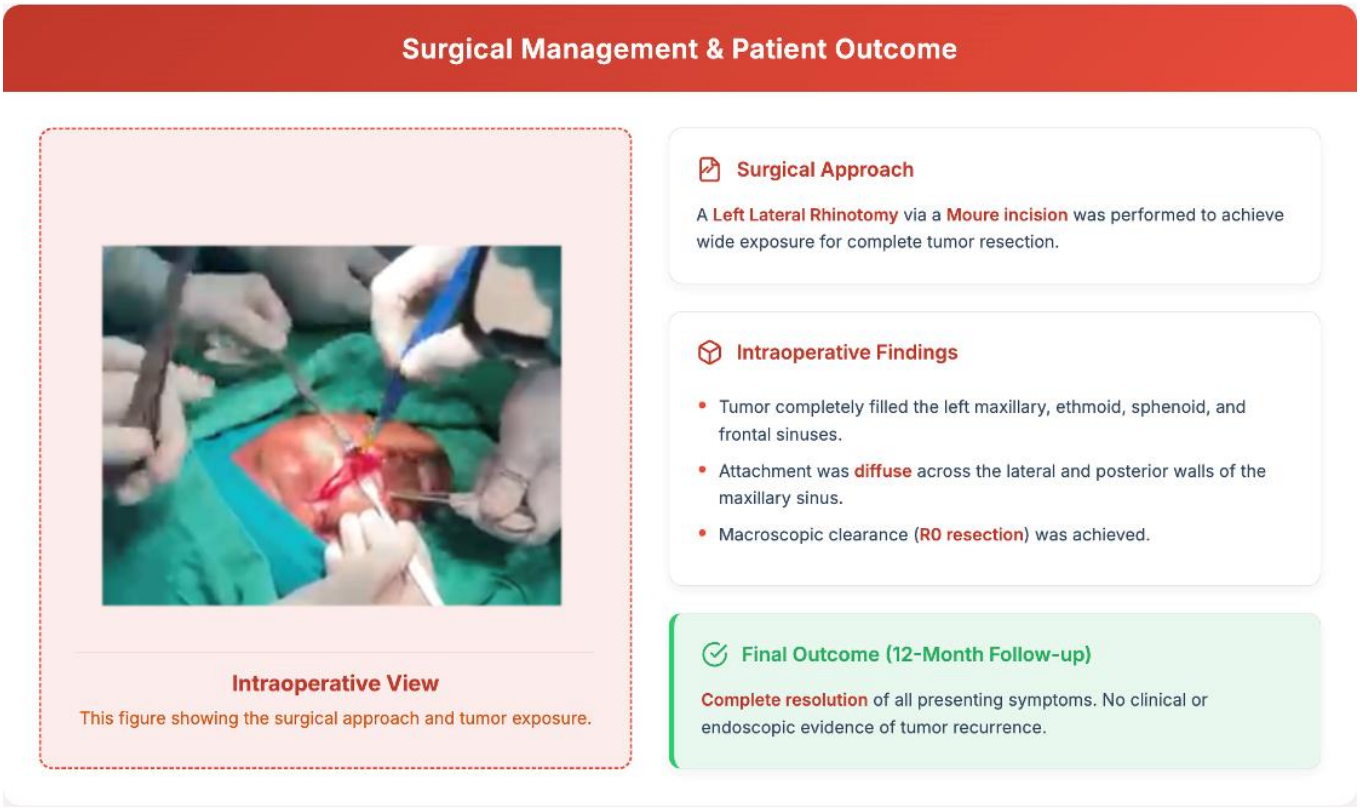

Figure 5. Surgical management & patient outcome.

3. Discussion

The diagnosis and management of the 65-year-old male presented in this report provide a valuable opportunity to conduct an in-depth exploration of a rare pathological entity. This case is particularly instructive due to its occurrence in an older individual without the genetic stigmata of neurofibromatosis type 1 (NF1), its clinical and radiological mimicry of a common inflammatory condition, and the necessity of a traditional open surgical approach for definitive

treatment.[11] To fully appreciate the clinical challenges posed by this tumor, one must first understand its fundamental biology. A neurofibroma is not a simple, monoclonal proliferation of a single cell type; rather, it is a complex, disorganized, benign neoplasm that represents a veritable microcosm of a peripheral nerve.[12] It is a true hamartoma of the nerve sheath, containing a heterogeneous population of cells essential for nerve function and maintenance. The primary neoplastic element is the Schwann cell, which has lost its tumor suppressor function.[13] However, these neoplastic Schwann cells recruit and are intimately admixed with a variety of other non-neoplastic cells, including perineurial-like cells, fibroblasts, endothelial cells of the microvasculature, and a significant number of resident mast cells. This complex cellular interplay is crucial to the tumor's structure and behavior.[14] The entire cellular milieu is embedded within an extracellular matrix rich in myxoid ground substance and haphazardly arranged collagen fibers, which contributes to the tumor's characteristic soft, rubbery consistency. Unlike its close relative, the schwannoma, which grows by eccentrically displacing the parent nerve axon, the neurofibroma is unencapsulated and grows intraneurally, meaning it infiltrates and splays apart the nerve fascicles, incorporating the native axons within its bulk. This intrinsic relationship with the nerve makes it impossible to resect the tumor without sacrificing the nerve of origin. The molecular pathogenesis of neurofibromas is one of the best-understood pathways in tumor biology and centers on the NF1 gene located on chromosome 17q11.2. This gene encodes a large, complex protein called neurofibromin. Neurofibromin functions as a critical tumor suppressor by acting as a GTPase-activating protein (GAP).[15] Its primary role is to accelerate the conversion of the active, signal-promoting form of Ras (Ras-GTP) to its inactive form (Ras-GDP). By inactivating Ras, neurofibromin effectively applies the brakes to the Ras/MAPK signaling pathway, a central cascade that controls cellular proliferation, differentiation, and survival. In the context of the genetic disorder NF1, individuals are born with one mutated, non-functional allele of the NF1 gene in every cell of their body (a germline mutation). A sporadic somatic mutation in the remaining healthy allele within a Schwann cell precursor—the "second hit" in Knudson's classic hypothesis—leads to a complete loss of neurofibromin function in that cell. With the brakes removed, the Ras pathway becomes constitutively active, driving the uncontrolled proliferation that initiates the formation of a neurofibroma.

The patient in this report, however, had a solitary neurofibroma without any signs of NF1. In these sporadic cases, the pathogenesis is believed to follow the same "two-hit" model, but both mutations occur somatically within a single Schwann cell precursor during the individual's lifetime. The first hit is a random mutation in one NF1 allele, and the second hit is a subsequent mutation or loss of the other allele. The end result is the same: a focal, clonal proliferation of neurofibromin-deficient Schwann cells that gives rise to a single, isolated tumor. The fact that our patient developed such a large tumor at an advanced age is consistent with this model, which requires two independent, random mutational events to occur over a lifetime. The activated Ras signaling in the neoplastic Schwann cells fundamentally alters the tumor microenvironment. These cells secrete a host of chemokines and growth factors, such as CXCL12 and Kit ligand, which act as powerful chemoattractants for other cell types. This explains the heterogeneous cellular composition seen on histology: the neoplastic Schwann cells are intimately admixed with a large population of recruited, non-neoplastic cells, including fibroblasts and a striking number of mast cells. This process is clearly demonstrated in our patient's pathology. These recruited mast cells are not passive bystanders; they degranulate and release their own potent mediators, including histamine, tryptase, vascular endothelial growth factor (VEGF), and transforming growth factor-beta (TGF-β). These factors, in turn, promote angiogenesis, further fibroblast proliferation, and the deposition of the

myxoid-collagenous extracellular matrix that gives the tumor its characteristic bulk and consistency.[16] This complex interplay between the neoplastic cells and their microenvironment creates a self-perpetuating cycle of growth. The extensive bony erosion seen in this case is a direct consequence of the tumor's slow, indolent, yet relentless growth. It is not a sign of malignant infiltration but rather a process of pressure-induced osteolysis. As the tumor gradually expands within the rigid confines of the sinonasal skeleton, it exerts constant pressure on the surrounding bone. This chronic pressure is thought to stimulate local osteoclast activity, leading to a gradual resorption and remodeling of the bone.[17] The tumor essentially carves out a space for itself over the years, which explains the smooth, scalloped margins of the bony defects seen on CT scans, as opposed to the permeative, destructive pattern typical of high-grade malignancies. This biological behavior is a key radiological feature that, while not specific, should raise the possibility of a slow-growing benign neoplasm.

The misinterpretation of the CT scan likely stemmed from a combination of factors, including the tumor's low density (25-35 HU), its complete lack of contrast enhancement, and its multi-sinus involvement, all of which can be seen in extensive inflammatory disease with retained secretions or mucocele formation. However, this interpretation overlooks several crucial red flags. The initial radiological interpretation of sinonasal polyposis highlights a significant cognitive pitfall. In any patient, particularly an elderly individual, with a unilateral sinonasal mass causing significant bony erosion, neoplasia must be the leading diagnosis until proven otherwise, irrespective of the lack of contrast enhancement. Inflammatory polyposis is typically bilateral. While unilateral presentation occurs, it is less common and should prompt a search for an underlying cause, such as a neoplasm causing unilateral obstruction. Furthermore, the pattern of bone change was more than simple remodeling; the frank erosion of the thick medial maxillary wall and nasal septum is atypical for polyposis and strongly suggestive of a neoplastic process. This case underscores the importance of maintaining a high index of suspicion and resisting the cognitive bias of attributing common findings (sinus opacification) to the most common cause (inflammation) without critically evaluating all the evidence. Schwannomas are the most common sinonasal PNST. Unlike the unencapsulated, infiltrative neurofibroma, schwannomas are well-encapsulated. On imaging, they often show cystic degeneration and more avid, heterogeneous enhancement. Histologically, they are composed purely of Schwann cells arranged in characteristic hypercellular (Antoni A) and hypocellular (Antoni B) patterns, and they lack the mixed cellular population of a neurofibroma. Inverted Papilloma, a benign but locally aggressive tumor, typically arises from the lateral nasal wall. On CT, it is known to cause focal hyperostosis at its site of attachment. On MRI, it classically displays a "convoluted cerebriform" pattern on T2-weighted images. These features were absent in our case. Malignant tumors (SCC, Adenocarcinoma, MPNST) were a prime concern in this 65-year-old. However, the three-year history without the development of "red flag" symptoms like severe pain, cranial neuropathies, or skin invasion argued against a high-grade malignancy. Radiologically, malignancies typically cause permeative bone destruction and show avid, often necrotic, contrast enhancement. Histologically, a low-grade malignant peripheral nerve sheath tumor (MPNST) can be a challenging differential. However, MPNSTs would be expected to show a higher cell density, nuclear atypia, and mitotic activity, all of which were absent in our patient's tumor. The definitive diagnosis of a sinonasal spindle cell tumor cannot be made on morphology alone. The histopathological features in this case were essential. The poorly circumscribed, hypocellular proliferation of bland spindle cells with wavy, "serpentine" nuclei and scant, indistinct eosinophilic cytoplasm, arranged in short, intersecting fascicles and loose clusters within a mixed myxoid and collagenous stroma containing numerous interspersed mast cells, are all classic

feature of a neurofibroma. This comprehensive morphological assessment represents the modern standard of care and is a non-negotiable step in the diagnostic process.[18]

The definitive treatment for this tumor was complete surgical excision. The central debate in this case revolves around the choice of surgical approach. While the endoscopic era has rightfully relegated many open procedures to a secondary role, this case provides a powerful argument for the continued, albeit selective, use of the lateral rhinotomy. The decision was not based on a reluctance to embrace modern techniques, but on a rigorous, anatomy-based analysis of the tumor's specific characteristics. A standard endoscopic approach, centered on a middle meatal antrostomy, would have been fundamentally inadequate. This approach provides excellent access to the medial and posterior walls of the maxillary sinus but offers a very limited view and reach to the anterior and, most critically, the lateral walls. Given that this tumor had a diffuse attachment across all walls of the maxillary sinus, attempting a resection through this limited corridor would have made achieving an R0 resection a matter of guesswork rather than surgical certainty. One must also consider whether more advanced, multi-portal endoscopic techniques could have succeeded. Endoscopic Medial Maxillectomy with a Prelacrimal Recess Approach. This technique involves removing the medial maxillary wall and drilling down the bone anterior to the nasolacrimal duct, providing improved access to the anterior and medial aspects of the sinus.[19] However, it would still have offered a poor angle of attack for the tumor's broad attachment to the far lateral wall. Canine Fossa Puncture / Endoscopic Modified Caldwell-Luc involves creating a small accessory portal through the canine fossa to allow for the introduction of a second instrument for triangulation and dissection of the lateral wall. While useful, the sheer bulk of the tumor in this case would have filled the entire sinus, leaving no working space between the tumor and the sinus walls for instrument manipulation. The tumor itself would have obstructed the view from the endoscope. Multi-Sinus Involvement, the extensions into the frontal and sphenoid sinuses could have been addressed endoscopically (requiring a Draf III and wide sphenoidotomy, respectively). However, this would have added significant time and complexity to an already challenging procedure, with the core problem of the maxillary sinus origin remaining unresolved. The lateral rhinotomy was chosen because it uniquely solved all of these challenges. The Moure incision and elevation of the cheek flap provided an unparalleled, wide-field view of the entire surgical field. The removal of the anterior maxillary wall converted the sinus from a keyhole into an open box, allowing for dissection of the tumor from all its attachments under direct, binocular vision.[20] This approach facilitated absolute certainty in achieving a macroscopic complete resection, which is the single most important determinant of long-term success. The lateral rhinotomy should not be viewed as an archaic technique, but as an essential and superior tool for ensuring the definitive treatment and long-term cure of the patient in this specific anatomical context.

Figure 6 showed a comprehensive and elegant schematic detailing the molecular and cellular pathophysiology of a solitary sporadic neurofibroma, illustrating the cascade from a single genetic event to its ultimate clinical manifestation. The diagram meticulously charts a linear, cause-and-effect progression, beginning with the foundational genetic mutation that initiates the entire neoplastic process. The first step, labeled Genetic Event, identifies the root cause as somatic "two-hit" mutations within the NF1 gene, occurring in a lone Schwann cell precursor. This is the cornerstone of the tumor's pathogenesis in a sporadic context, distinguishing it from the syndromic neurofibromatosis type 1, where an individual inherits one faulty allele. In this case, both mutational events happen randomly within the same cell over the course of the patient's life, a model consistent with the development of a large, isolated tumor at an advanced age. This initial step underscores the clonal origin of the neoplasm, stemming from a single, unfortunate cellular event.

Figure 6. Pathophysiology of a solitary sporadic neurofibroma.

Following this genetic insult, the diagram transitions to the immediate biochemical consequence. The second step, Molecular Consequence, explains that the biallelic inactivation of the NF1 gene leads to the complete loss of its protein product, neurofibromin, within the affected Schwann cell. This is the pivotal molecular switch that triggers the subsequent dysregulation. Neurofibromin is a critical tumor suppressor protein whose primary function is to act as a GTPase-activating protein (GAP). In its healthy state, it serves as a crucial "brake" on cellular signaling by facilitating the conversion of the active Ras-GTP to its inactive Ras-GDP form. The loss of this protein, as depicted in the figure, is therefore not a passive event but the removal of a fundamental regulatory mechanism, setting the stage for uncontrolled cellular activity. This loss of function is the central molecular lesion that directly drives the formation of the neurofibroma. The narrative then flows logically to the third step, Pathway Dysregulation, which illustrates the downstream effect of neurofibromin's absence. With the molecular brakes removed, the Ras/MAPK signaling pathway becomes constitutively active. This pathway is a central command-and-control cascade that governs fundamental cellular processes, including proliferation, differentiation, and survival. Its persistent activation, as shown in the figure, creates a powerful and unrelenting signal for the cell to grow and divide. This step is crucial because it links the upstream molecular defect to the downstream cellular behavior. The Schwann cell is now fundamentally altered, hardwired for continuous proliferation due to the uninhibited signaling flowing through this critical pathway. It is this sustained, abnormal signaling that transforms the once-healthy Schwann cell into the primary neoplastic element of the neurofibroma. The fourth step, Proliferation & Recruitment, details the dual consequences of this dysregulated pathway. Firstly, the neoplastic Schwann cells begin to undergo uncontrolled proliferation, initiating the physical formation and growth of the tumor mass. However, the figure astutely highlights that a neurofibroma is not merely a monoclonal proliferation of a single cell type. The neoplastic Schwann cells actively remodel their surroundings, becoming architects of a complex tumor microenvironment. They achieve this by secreting a variety of chemokines, such as CXCL12, and growth factors. As the schematic indicates, these signals act as powerful chemoattractants, recruiting a host of non-neoplastic cells—most notably fibroblasts and mast cells—into the growing tumor. This recruitment is a defining characteristic of neurofibromas, transforming them from a simple collection of tumor cells into a complex, heterogeneous tissue composed of multiple interacting cell types. This cellular complexity leads directly to the fifth step, Amplification & Growth, which describes how the recruited cells become active participants in the tumor's expansion. The mast cells, in particular, are not passive bystanders. Upon entering the tumor microenvironment, they degranulate and release a potent cocktail of their own mediators, including vascular endothelial growth factor (VEGF) and transforming growth-factor beta (TGF-β). These factors create a positive feedback loop; VEGF promotes angiogenesis, providing the tumor with the necessary blood supply for its expansion, while TGF-β stimulates further fibroblast proliferation and the deposition of the myxoid-collagenous extracellular matrix that gives the tumor its characteristic bulk and rubbery consistency. This process, as the figure describes, establishes a self-perpetuating cycle of tumor expansion, where the interplay between neoplastic Schwann cells and their recruited microenvironment drives relentless, albeit slow, growth over a period of years. The slow, indolent, yet inexorable growth of the tumor mass within the rigid confines of the sinonasal tract exerts a chronic, low-grade pressure on the surrounding bone. This is not an aggressive, malignant invasion but rather a process of pressure-induced osteolysis. The constant pressure is believed to stimulate local osteoclast activity, leading to the gradual resorption and remodeling of the bone as the tumor carves out space for itself over years. This specific biological behavior explains the characteristic

findings on a CT scan: smooth, scalloped, and expansile bony changes rather than the permeative destruction typical of malignancy. Thus, Figure 6 masterfully encapsulates the entire disease process, providing a clear and scientifically robust narrative that links a microscopic genetic event to the macroscopic radiological findings that define the clinical challenge of this rare tumor.

4. Conclusion

This case of a massive, solitary sinonasal neurofibroma masquerading as an inflammatory disease in an elderly male is a powerful clinical lesson. It highlights that in the evaluation of any persistent, unilateral sinonasal mass, neoplasia must be the primary consideration, and a definitive diagnosis requires a comprehensive histopathological analysis. Most critically, this case serves as an analytical reaffirmation of a core surgical principle: the surgical approach must be tailored to the unique anatomy of the tumor to uphold the paramount goal of complete oncologic resection. For massive, benign neoplasms with diffuse attachments across the lateral and anterior walls of the maxillary sinus, the lateral rhinotomy should not be viewed as a procedure of last resort, but as a deliberately chosen, superior surgical strategy that provides the certainty of exposure required to achieve a definitive cure.

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