SOLITARY NASAL NEUROFIBROMA: A CASE REPORT

NEUROFIBROME ISOLÉ DE LA FOSSE NASALE: A PROPOS D'UN CAS

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ABTRACT

Introduction: Neurofibroma of the nasal septum is a rare benign tumor with non-specific clinical and radiological features, frequently mimicking malignant lesions (e.g., chondroma or sarcoma) and leading to diagnostic uncertainty. This poses significant challenges in preoperative planning and surgical approach selection.
Case presentation: A 63-year-old woman presented with chronic nasal obstruction. Imaging suggested chondroma or sarcoma. Complete endoscopic resection led to a diagnosis of neurofibroma (PS100+, CD34-). The postoperative course was favorable, with no recurrence after 22 months of follow-up.
Conclusion: This case underscores the diagnostic dilemma of solitary nasal neurofibromas and demonstrates that transnasal endoscopic resection is an effective solution, ensuring complete excision while preserving function and aesthetics.
Keywords: nasal neoplasm, neurofibroma, endoscopic surgery

RESUMÉ

Introduction: Le neurofibrome du septum nasal est une tumeur rare aux manifestations cliniques et radiologiques non spécifiques. Nous rapportons un cas traité par voie endoscopique endonasale.
Cas clinique: Une femme de 63 ans se présentait pour une obstruction nasale chronique. L'imagerie évoquait un chondrome ou un sarcome. L'exérèse endoscopique a permis un diagnostic de neurofibrome (PS100+, CD34-). L'évolution était favorable sans récidive après 22 mois de suivi.
Conclusion: L'exérèse endoscopique endonasale est une approche efficace permettant une résection complète tout en préservant la fonction nasale et l'esthétique.
Mots-clés: tumeur nasale, Neurofibrome, Chirurgie endoscopique.

INTRODUCTION

Neurofibroma is a neurogenic benign tumor, commonly found in the head and neck and originates from the vestibular nerve. However, neurofibroma arising from the nose or paranasal cavities are rare, especially from the nasal septum [1-4].
The aim of this study is to describe clinical, radiologic and therapeutic specificity of the nasal neurofibroma. Through this study, we report a rare case of a solitary neurofibroma of the nasal septum treated by transnasal endoscopic approach. The challenges encountered in the management of such cases and a review of the literature were discussed.

OBSERVATION

63 years old north african woman with no personal medical history nor Von Recklinghausen's disease nor a family history of neural tumors was referred by a general practician to our department. The patient consulted for chronic and bilateral nasal obstruction with anosmia, that worsened progressively. No other rhinological or otological symptoms were reported. The endoscopic examination showed a process depending on the posterior part of the nasal septum and occupying the entire nasopharynx, with a hard-stony consistency. Facial mass CT scan revealed a suspicious locally aggressive tumor of the nasopharyngeal postero-superior walls, measuring 40 mm in length, with osteolysis and spiculated periosteal reaction of the body of the sphenoid and the vomer (Figure 1). Nasopharyngeal MRI showed a tumoral process invading the clivus (Figure 2) near the sella turcica (Figure 3), the sphenoid sinuses and filling the oropharynx (Figure 4), without intracranial or parapharyngeal extension. The tumor had necrotic areas and measured 45 mm in length. The mass was initially suggestive of chondroma or sarcoma.

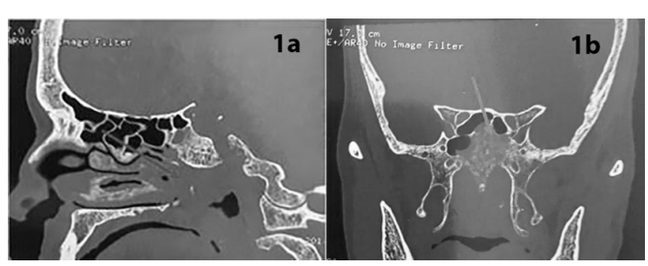

Figure 1: 1a: Facial mass CT scan in sagittal section revealing extensive nasal tumor (red arrow)
1b: Facial mass CT scan in coronal section revealing the tumor with spiculated periosteal reaction of the sphenoid bone (arrow)

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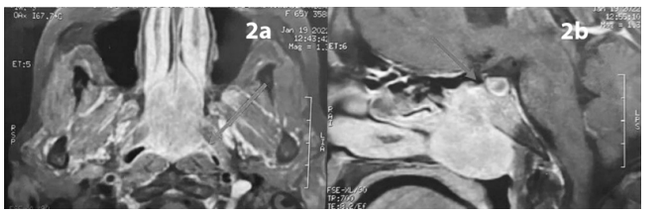

Figure 2: 2a: Nasopharyngeal MRI in axial section, Tl-weighted image: showing the enhanced nasal tumor, filling the nasopharynx without parapharyngeal extension (arrow)
2b: Nasopharyngeal MRI in sagittal section, T1-weighted image: showing the enhanced nasal tumor invading the clivus (arrow)

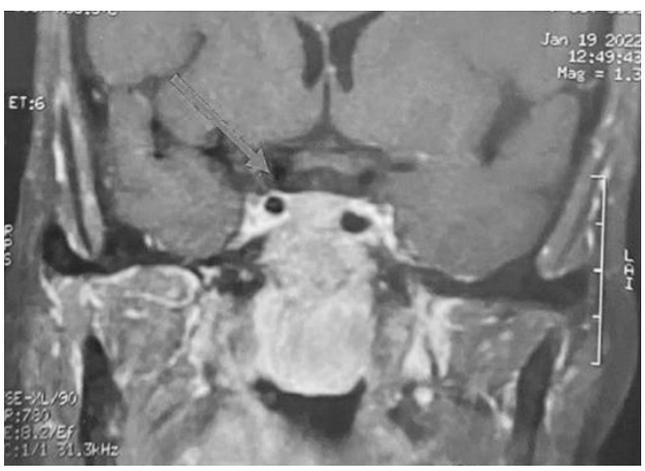

Figure 3: Nasopharyngeal MRI in coronal section, T1-weighted image: showing the enhanced nasal tumor near the sella turcica (arrow)

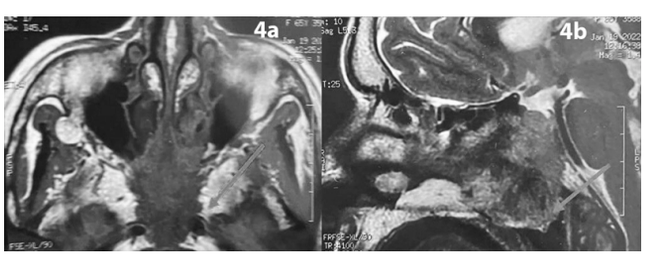

Figure 4: 4a: Nasopharyngeal MRI in axial section, T2-weighted image: showing the unenhanced nasal tumor extending to the sphenoidal sinuses (arrow)
4b: Nasopharyngeal MRI in sagittal section, T2-weighted image: showing the unenhanced nasal tumor filling the nasopharynx and the oropharynx (arrow)

The mass biopsy, performed under local anesthesia, revealed at the anatomo- pathological examination chondromyxoid debris but did not allow the diagnosis.
A surgical treatment, consisting of complete excision of the process and the posterior part of the nasal septum, was performed by transnasal endoscopic approach.
Anatomo-pathological examination revealed an infiltrating fuso-cellular proliferation with a myxoid background. The immunohistochemical study showed an intense and diffuse positivity to PS100 and a negative CD34. The diagnosis of neurofibroma was established.
The evolution was marked by the relief of symptoms and no postoperative complications were noted. After a 22 months period of follow up, the endoscopic examination and the postoperative imaging remain normal and confirm the absence of tumor recurrence (Figure 5).

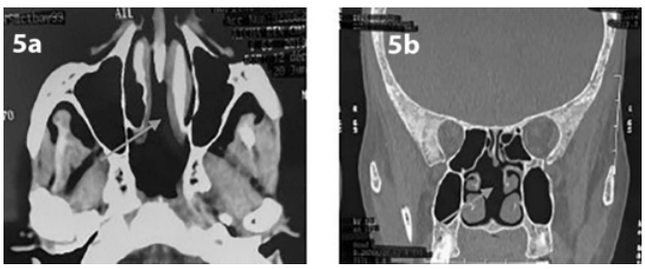

Figure 5: Facial mass CT scan in axial (a) and coronal (b) sections: showing a nasal fossa free of tumor, the nasal septum was removed (arrow)

DISCUSSION

Neurofibromas are localized slow-growing, benign tumors, arising from peripheral nerves sheath
[3, 5]. They can therefore develop in every possible anatomic location in the body. These tumors are commonly found in the head and neck and originate from the vestibular nerve. However, their occurrence in the nose is rare, especially in the nasal septum [1– 3].
Almost all neurofibromas are associated with the autosomal dominant disorder Neurofibromatosis type 1, also known as Von Recklinghausen's disease. Isolated neurofibromas without this association are extremely rare [1,2,5,6].
Because of how rare solitary nasal neurofibromas are, most authors report cases and we lack epidemiological data in literature. Some authors reported that there was no gender predilection [2,7,8] while other authors noted a female predominance [1,10].
Although isolated peripheral nerve-sheath may develop at any age [2,7], its incidence is higher in the fourth [2,8,10], fifth [1,9,10] and sixth [1,2,9,10] decades of life.
Clinical findings of nasal neurofibromas are nonspecific and similar to others nasal tumors
[2,6,10]. The symptoms and signs depend on the location of the tumor and the surrounding structures involved [8, 10]. Nasal neurofibroma usually presents with a mass lesion causing nasal obstruction, pain, and epistaxis [6,8-10].
Radiological findings of neurofibromas are non- specific and depend on the tumor extension [5]. CT scanning and MRI are the main imaging methods used [10]. Preoperative evaluation CT reveals a tissular mass with heterogeneous contrast enhancement, characteristic of neurofibroma [10].
It detects the pushing borders of the tumor, instead of bone lysis or cerebral involvement that are typical of malignancy [8,9].
Therefore, neurofibroma of the nasal cavity are frequently misdiagnosed preoperatively because of the non-specific nature of their symptoms, clinical findings and radiological constatations [2,5,7]. Even histological

appearances in biopsy tend to be non-specific [10].
The diagnosis is most often unexpected and reached postoperatively by histopathological and immunohistochemical studies [1–3,7]. Histologically, neurofibroma is non- encapsulated, poorly circumscribed [1,9-10]. In contrast, schwannoma is an encapsulated tumor [5].
Ultra-structurally, neurofibromas are often associated with nerves, showing a centripetal distribution of the tumor cells and a characteristic blending of Schwann cells, perineurial fibroblasts with collagen and myxoid stroma [1,7,8]. Neurofibromas with myxoid changes may be difficult to differentiate from myxomas that involve facial bones [8].
Immunohistochemical examination shows positivity for S100 protein, indicating that the mass originates from Schwann [2, 7, 8, 10].
Complete surgical excision of the tumor is the only radical treatment [1, 2, 8]. Surgical approach techniques depend on the extent and the location of the tumor [8]. Moreover, functional and cosmetic effects of the surgery should be considered [8].
Until several years ago, most surgeons recommended external approaches (midfacial degloving, lateral rhinotomy or rhinoplasty), but recently with the development of endoscopic nasal surgery, transnasal endoscopic excision of benign nasal tumors is widely emerging [6].
In the literature, few surgeons opted for transnasal endoscopic surgery for total resection of neurofibroma of the nasal cavity. They reported a complete excision of the tumor and no recurrence during follow up [6, 9, 10].
Transnasal endoscopic resection should be considered especially when faced to a solitary, small, and localized nasal neurofibroma with an identified origin [8, 10].
Treatment of nasal neurofibromas is challenging because of their location, high recurrence and more importantly potential esthetic outcomes, making complete excision a challenge. In fact, A high local recurrence rate is typical with incomplete surgical excision [5]. However, if the tumor is completely removed, prognosis is excellent without recurrence in most series [1, 2, 6, 7].
Malignant transformation of neurofibroma is rare, around 10% to 15%, unless the patient has neurofibromatosis [1,6,8,10]. Patients should be closely monitored given the possibility of malignant transformation of solitary neurofibroma even though it is rare.

CONCLUSION

Solitary neurofibromas are slow-growing benign tumors that are rarely located in the nasal septum.
Due to their non-specific clinical and radiological features, they often pose a significant diagnostic challenge, mimicking malignant tumors such as chondromas or sarcomas. This diagnostic uncertainty complicates preoperative planning and raises therapeutic concerns regarding the optimal surgical approach.
Nasal neurofibromas are curable by complete surgical excision, which is critical to prevent recurrence. The transnasal endoscopic approach emerges as a promising technique in this context, offering a conservative yet effective solution. It enables complete tumor resection while preserving nasal function and aesthetics, addressing both the diagnostic dilemma and therapeutic challenges associated with these rare tumors.

ETHICAL APPROVAL

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

DECLARATION OF CONFLICTING INTERESTS

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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