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Case Report

Nasal Septal Schwannoma: a Case Report and Review of the Literature

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Abstract

The authors report the story of a schwannoma implanted on the left side of the nasal septum in a 46- year-old female patient complaining of nasal obstruction. A sinus CT-scanner showed a non- encapsulated 32 x 28 x 18 mm soft-tissue mass in the left nasal fossa, associated with a left anterior sinusitis. Biopsy didn't show any sign of malignancy. The tumour was resected endoscopically associated to a middle antrostomy and an ethmoidectomy under general anaesthesia. Pathological and immunohistochemical analysis gave the final diagnosis. With a follow up of 4 years, there was no recurrence. Interestingly, the patient reported the resection of another schwannoma originated from the median hard palate 28 years earlier. The authors report the case and review the worldwide literature on this rare lesion.

Introduction

Schwannomas are rare, mostly benign neoplasms constituted of proliferative differentiated Schwann cells from myelinated nerve sheaths, derived from the neural crests. They usually present as well-circumscribed and frequently encapsulated slow-growing solitary tumors. 90% of schwannomas are sporadic. They can on occasion be multiple or recurrent, either in cases of incomplete resection, or in certain genetic conditions.

They originate from any myelinated nerve (somatic, autonomic and cranial nerves) except for olfactory and optic nerves who don't have any Schwann cell, but very similar olfactory unsheathing cells derived from the olfactory placode. On the nasal septum, this concerns autonomic fibres heading to the submucosal glands and vessels or trigeminal maxillary and ophthalmic branches mediating somatosensory signals from the mucosa through the naso- palatine, great palatine and naso-ciliary nerves. Schwannomas constitute approximately 5% of all tumors of the head and neck, which hosts 25 to 45% of them. Less than 4% of schwannomas are found in the sinonasal area1, while up to 80% arise from the vestibular nerve. In the nose and paranasal sinuses, the most frequent sites are the ethmoidal sinus, the maxillary sinus, and finally the sphenoid sinus. Schwannomas originating from the nasal septum are exceedingly rare.

Case Report

A 46-year old Mediterranean woman first consulted in pneumology for sleeping disturbance. She had unilateral nasal obstruction with an important rhinolalia and left muco-purulent

rhinorrhea. There was no complaints of epistaxis, facial pain, facial swelling or hyposmia. In her medical history, we noticed an allergic asthma and allergic rhinitis, no chronic sinusitis, nasal surgery or smoking history. She reported the resection of a schwannoma from the median hard palate 28 years before. A non-contrast-enhanced computed tomography (CT) of the paranasal sinuses revealed a 32 x 28 x 18 mm homogenous non- encapsulated soft tissue density in the left nasal cavity and olfactory fossa (Figure 1). The nasal septum deviated on the right side. The left middle turbinate was lateralized and there was an associated left anterior pansinusitis. There was no bone erosion. The patient was referred to the Otorhinolaryngology outpatient clinic. On examination, a well-circumscribed submucosal mass with light and smooth overlying mucosa was observed occupying the left nasal cavity. A biopsy was performed endoscopically under local anaesthesia. Microscopical analysis of the sample didn't show any malignant characteristic, despite not being formally identified.

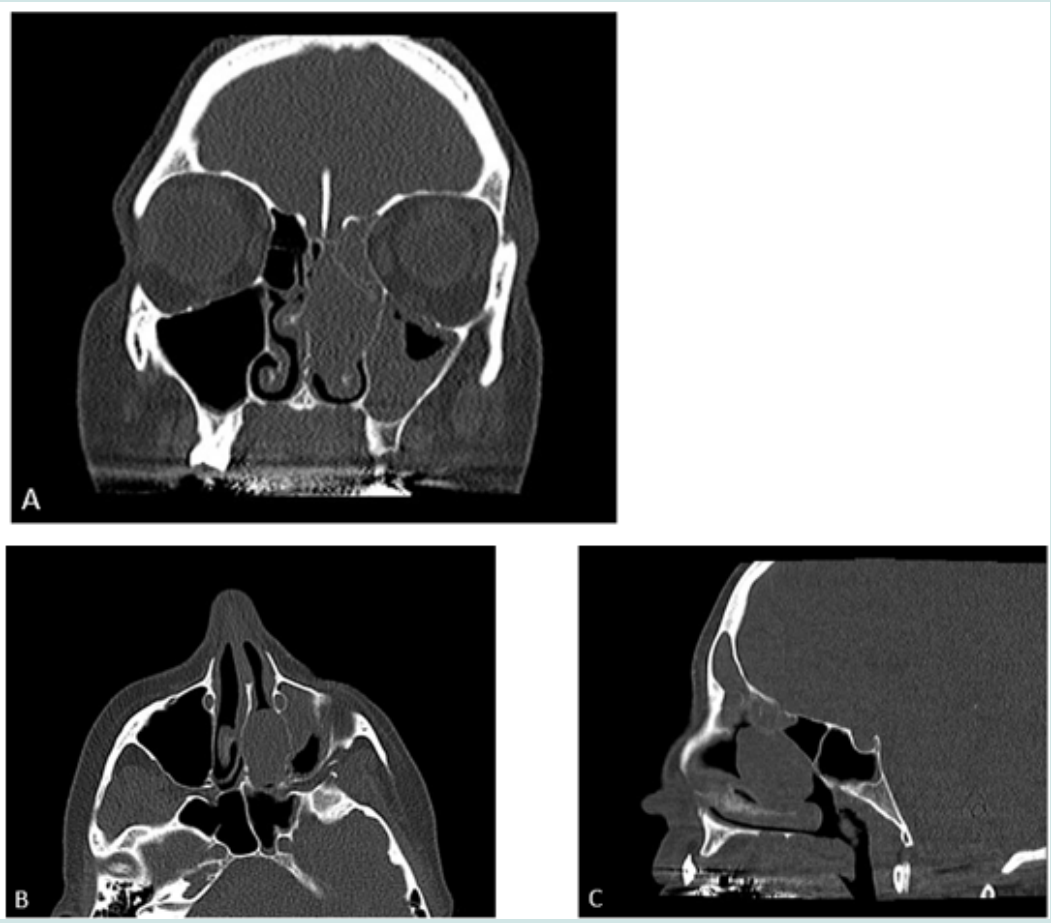

Figure 1: Non contrast-enhanced CT scanner of the paranasal sinuses in bony window. A - coronal image : well- demarcated tissular mass in the left nasal fossa, osteomeatal complex obstruction, no orbital or cranial damage. B - axial image : no bone erosion, free lachrymal duct. C - sagittal image : posterior situation of the lesion. Left anterior sinusitis.

The mass was then removed endoscopically under general anesthesia with resection of the septal mucoperiochondrium. The resection was complete with safe surgical margins. The site of insertion was the posterior portion of the nasal septum. The authors combined the resection to a left middle antrostomy and left ethmoidectomy. The histological analysis of the specimen (Figure 2) showed bundles of spindle-shaped cells with fusiform nuclei showing focal atypia, arranged into a dense organized cellular cytoplasm with a discrete palisading pattern. There were also alternating areas of normal respiratory and sinusal epithelia with a slightly inflammatory lympho-plasmocytic infiltrate in a looser hypocellular chorion. No evidence of necrosis was found into the analyzed specimen. Immunohistochemical staining was diffusely and strongly positive for S100 protein and negative for desmin, actin and epithelial membrane antigen (EMA). The final diagnosis was a benign schwannoma of the nasal septum. Post-operative evolution was uneventful. A postoperative MRI performed 6 months after surgery confirmed the absence of residual tumour. 4 years after the intervention, the patient is still free of disease. The authors wanted to refer the patient to a geneticist because this was the second resection of a schwannoma in this patient, but she refused.

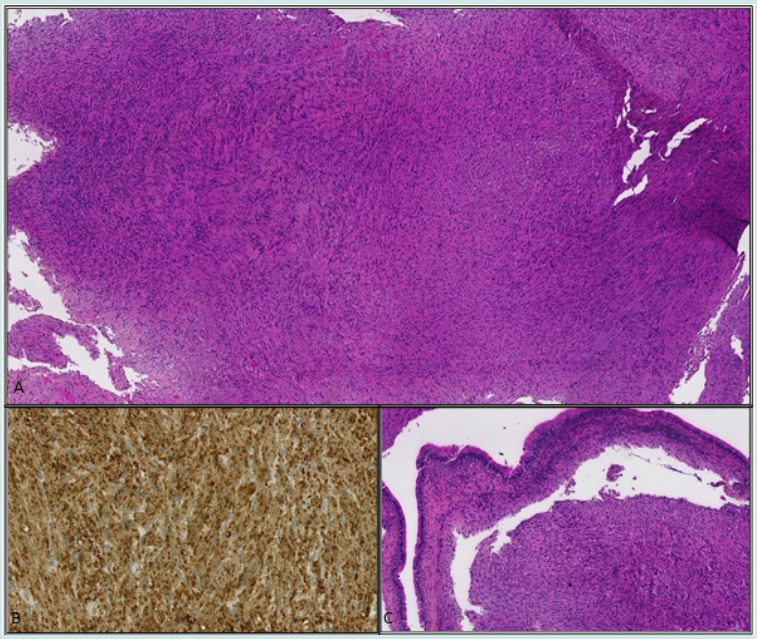

Figure 2: Anatomopathological analysis of the specimen. A - Antoni A palisading pattern forming Verocay bodies dispersed in a loose chorion (Antoni B). B - Immunohistochemical staining shows diffuse positivity for S-100 protein. C - clear delineation between the nasal respiratory epithelium and the submucosal mass.

Literature Review

The authors reviewed different databases (PubMed, Research gate, Europe PMC) with the terms: "nasal septal schwannoma", "nasal septum schwannoma" and "nasal septal neurinoma". They found 35 reported cases between January 2000 and September 2020 in the available English scientific literature. Sino- nasal schwannomas are more commonly reported. In this review, there was no gender preponderance. The tumour arises in adults with ages ranging from 16 to 82 years old. The diagnosis is generally suspected by nasal endoscopy and the extension is defined by imaging: typically a contrast-enhanced CT scan and sometimes an MRI. Bone erosion is often seen, despite the benign character of these tumours, leading to an erroneous suspicion of malignancy [1] (Table 1).

Table 1: Literature review : all nasal septal schwannomas reported in the available English literature between 2000 and 2020.

Authors	year	Age / sex	side	Imaging	Extention to vital structures + size	Surgical procedure	Follow-up
Berlucchi & al.	2000	29/M	L	Contrast CT	4x3x1.5 cm	Degloving En bloc medial maxillectomy	NED 7 yr
Wada & al.	2001	62/F	R+L	Contrast CT MRI	Anterior ethmoid sinuses To the skull base	Lateral rhinotomy En bloc tumorectmoy	NED 15 mo
Cakmak & al.	2003	33/M	R	-	0.5x1 cm	Local + margin	NED 6 yr
Wang & al.	2004	55/M	R	CT	Bone erosion medial wall of maxillary sinus	Endoscopy Piecemeal + margin	NED
Park, Lee & Byun	2007	51/F	R	Contrast CT MRI	Bone erosion : nasal septum, medial wall of maxillary, sphenoid	endoscopy - 10mm margin Piecemeal	NED 1 yr
Dharia & al.	200707	19/F	L	CT	Septal deviation 3.6x2.5x2.1 cm	Endoscopy	NED 4 yr
Pagella & al.	2009	82/M	L	CT	sphenoethmoid recess	Endoscopy En bloc + free margins	NED 5 yr
kodama & al	2010	41/F	L	Contrast CT	posterior nasal cavity, superior meatus and sphenoid sinus with sphenoidal mucocele	endoscopy + Harmonic sphenopalatine cautery + septoplasty + sphenoidectomy	NED 2 yr
				MRI	inhomogeneous enhancement	en bloc	

Zhou & al.	2012	20/M	R	CT		endoscopy	NED 6 Mo
					7x10 mm	en bloc + free cartilage cuff	
Mitra & al.	2012	24/M	L	CT		endoscopy	NED 6 Mo
					3.0x1.6x1.5 cm	en bloc + free cartilage cuff	
Hu & al.	2012	51/M	L	Contrast CT	maxillary sinus : medial wall dehiscence	lateral rhinotomy	NED 12,3 yr
					5.0x6.0 cm		
		48/F	R	Contrast CT	nasopharynx, vomer 3.0x2.0x2.1 cm	lateral rhinotomy	NED 4.8 yr died of heart disease
		56/M	L	Contrast CT	- 1.0x1.0	Endoscopy en bloc +free mucoperiostum margin	NED 7.8 yr
Gulia & al.	2013	35/F	L	- -	1.5x1.0x1.0 cm	En bloc tumorectmoy local anesthesia	NED 6 Mo
Furlan Pauna & al.	2013	78/M	R	CT	Bone thining in maxillary sinus medial wall	Endoscopy en bloc +free mucoperiostum margin	-
Yilmaz & al.	2014	65/M	R	-	2.0x3.0 cm	septoplasty - local anesthesia en bloc - free margin	NED 4yr
Tati & al.	2014	40/M	R	CT	nose deformity, hypoesthesy R cheek	endoscopic	NED
					30x30x15 mm		
Dhingra & al.	2014	28/F	R	non contrast CT	ethmoid sinuses, maxillary sinus ostium, deviated thinned septum	endoscopic	NED
Lee & al	2014	53/M	R	non contrast CT	right choana and nasopharynx and some erosion of the septal bone	endoscopic	NED 1 yr
				MRI	2.5x1.5 cm	Piecemeal + mucosal margin	
Cadd & al.	2014	31/M	R	non contrast CT	-	midfacial degloving	NED
				MRI		en bloc - free margin	
Forer & al.	2015	52/F	R	non contrast CT MRI	-	posterior nasal septectomy	NED 96 Mo
Gawarle & al.	2015	70/F	L	Contrast CT	oropharynx up to nasal vestibule, deviated septum	endoscopic	NDE 10 Mo
					85x35x21	piecemeal	
Vallamreddy & al	2016	35/F	L	non contrast CT	both choanae, nasopharynx	endoscopic	NED 6 Mo
						piecemeal	
Sathe & al	2016	38/F	L	contrast CT	bilateral nasal cavities, lateral splaying of both medial maxillary walls, ethmoidal, sphenoidal sinus and nasopharynx, eroded and septum deviated to the right	endoscopic	NED 3 Mo
				MRI	54x42x54 mm	ostia + post septectomy	
Karatas & al	2016	31/F	R	Non contrast CT	through perforation from right to left nasal cavity, remodelling middle and inferior conchae	midfacial degloving + endoscopic	NED 8 Mo
					47x17 mm		

Valsamidis & al	2016	54/M	R	CT	posterior nasal septum, right nasal cavity, nasopharynx,	midfacial degloving en bloc	NED 6 Mo
Gupta & al	2017	40/F	L	Contrast CT	anterior nasal septum reaching external nares	lateral rhinotomy	NED 1 yr
Canevari & al	2017	74/M	R	Contrast CT	erosion of inferior and middle turbinates and ethmoid lamelle, septum displaced, thickening of antero-medical corner of orbital wall	endoscopic + diode laser	NED 1 yr
				MRI		piecemeal "disassembling technique" + margin + intraopertive frozen section	
Gerritsen & al	2017	16/M	L	CT	no bony erosion	endoscopic	NED 1 Mo
					1.0x1.5x2.1	cm en bloc	
Devaraja & al	2018	57/M	R	non contrast CT	-	endoscopic	NED 6 Mo
						piecemeal	
sadiya	2018	50/F	R	CT	no bone destruction, smooth margins	piecemeal	NED 1 yr
Kaushal & al	2019	17/F	R	non contrast CT	bone remodelling, left maxillary sinusitis	endoscopic + septoplasty	-
Alrasheed & al	2019	64/F	L	Contrast CT	mild heterogeneous enhancement and bone remodeling	endoscopic	NED 3 Yr
Mohamad Umbaik & al	2019	48/F	L	Contrast CT	left nasal cavity and left middle meatus + septum deviation	endoscopic	NED 8 Mo
					3.7x1.8x3.7cm		
Bie & al	2019	47/M	L	non contrast CT	left nasal cavity and left maxillary sinus, nasal septum deviated to right	endoscopic + plasma knife	NED 6 Mo
						5 mm margins	
Jamme & al	2023	46/F	L	non contrast CT	left nasal cavity and left middle meatus, nasal septum deviated to right, fronto-ethmoido-maxillary retention	endoscopic	NED 4 yr
						piecemeal + mucosal margin sinusitis	

M = male ; F = female ; L = left ; R = right ; CT = computed tomography ; MRI = magnetic resonance imaging ; NED = no evidence of disease.

Discussion

Although schwannomas are benign tumours, they have a significant comorbidity, mostly due to a mass effect leading to a bone erosion onto neighboring structures. The symptoms depend on the anatomical location of the lesion and on its size. In the nasal cavity, the most frequent symptoms are unilateral nasal obstruction, epistaxis, facial pain and sometimes nasal discharge. In long-evolving cases, more debilitating symptoms can occur, such as exophthalmos, diplopia, facial swelling, cranial nerve deficit, skull base fistula, hyposmia, epiphora or hypopituitarism [2]. In the nasal cavity and nasal septum, they tend to become symptomatic more quickly than in the paranasal sinuses. However, when it originates from the posterior part of the nasal septum the extension usually progresses towards the sphenoethmoidal recess, or the nasopharynx with a long asymptomatic period, which can cause a delay in diagnosis. Because the symptomatology and the macroscopic and endoscopic aspects of most intranasal masses are not specific, imaging and histology are mandatory to make the definitive diagnosis.

Imaging

Imaging techniques for sinonasal tumours include mainly cross-sectional CT (to evaluate thickening, remodeling, erosion, sclerosis or destruction of adjacent bone) and MRI (to characterize the tissular components of the mass, its extent, to rule out the presence of pathologic lymph nodes and to differentiate the lesion itself from retention sinusitis)[3]. Intravenous contrast medium injection for both imaging techniques is recommended in nasal tumours evaluation protocols. Schwannomas appear in tomography as well- delineated, often capsulated soft-tissue tumours with a local extension that can induce mucous retention and sometimes bone deformation or bone destruction by mass effect [1]. In gadolinium-enhanced multiplanar MRI, schwannomas are hypointense or isointense to skeletal muscles in T1-weighted images, and heterogeneously hyperintense in T2-weighted acquisitions, due to the alternance of Antoni A and Antoni B areas [3]. This pattern is not specific of nerve sheath tumours, but this category shares other specific features :

a. The "target sign", characterized by low- signal intensity with low contrast enhancement centrally and high-signal intensity with high contrast enhancement peripherally on T2-weighted and contrast- enhanced T1-weighted images, has a high specificity but a low sensitivity [4].

b. The "fascicular sign" shows a fascicular bundles pattern as in normal large diameter nerves.

c. If the affected nerve trunk is large, it can sometimes be seen at the periphery of the mass for schwannomas, but by far less frequently in neurofibromas, in which the affected nerve passes through the tumour and is often indistinguishable [5]. In the nose and paranasal sinuses, the nerve of origin is seldom identified due to their relatively small diameter.

In the present case, MRI wasn't performed preoperatively because it wouldn't have added any important information after CT. The latter was sufficient to rule out involvement of any vital structure and it gave clues towards a benign mass, easily removable in toto.

PET-CT can be used complimentarily in selected cases suspicious for malignancy, for N- and M- staging, to monitor the response to a treatment or to screen for residual or recurrent tumours. It is, however, not recommended for routine diagnosis [3].

Histology

Histopathological examination, often helped by immuno-histochemistry (IHC), is the only way to obtain a definitive diagnosis for tumours of the nose and paranasal sinuses. A biopsy of the mass is recommended before surgery. For significant tumours the biopsy must be performed under general anaesthesia. Typically these tumours have a trend to bleed. Schwannomas stained by haematoxylin and eosin show under optic microscopy a binary aspect of alternating Antoni A and Antoni B areas in various combinations. The former is made of compact indistinguishable spindle cells with twisted nuclei arranged in bundles of interlacing fascicles. This nuclear palisading pattern around a central acellular hypereosinophylic area defines a "Verocay body" [6]. Antoni B areas are by far less cellular with a loose myxoid stroma and occasionally glands or benign epithelial structures [2]. In addition, there is thickening and hyalinization of blood vessels' walls.

Immunohistochemistry

IHC of schwannomas shows a diffuse and intense positivity of the cytoplasm and nuclei for S-100 protein : a neural-crest antigen of the supporting cells of the nervous system which is specific to Schwann cells and melanocytes [7]. Thus, S-100 positive staining also applies to melanoma, meningioma or neurofibroma. Vimentin, an ubiquitarian protein of mesenchymal origin, is negative for schwannomas but not for other mesenchymal (sarcomas) and spindle-cell neoplasms. Smooth muscle actin (SMA) negativity helps to rule out leiomyoma. Desmin is a mesenchymal marker of myogenic origin to which smooth muscle tumours are inconstantly positive. EMA (Epithelial membrane antigen) and PgR (progesterone receptor) are both good markers for meningiomas [8]. The S-100 marker usually stains less diffusely and less intensely into neurofibromas than in schwannomas. Calretinin is expressed in 96% of schwannomas versus only 7% of neurofibromas. In peripheral nerves, CD34 is expressed by dendritic and endoneural fibroblastic cells. In schwannomas, CD34 staining underlines the biphasic pattern by being mostly present in Antoni B areas. On the contrary, CD34 is more diffuse in neurofibromas with no biphasic pattern [9].

Differential diagnosis

Although they are not exhaustive, these case series published by Bist [10] and Yildirim [11] give a broad overview of the different tumours often encountered in the nose. Histological assessment is therefore of utmost importance to make a differential diagnosis. The most important lesions to rule out are neurofibromas and malignant transformation. Neurofibromas are more prone to malignant evolution (incidence of 2,4% to 16,5%) than schwannomas and are more locally aggressive. Complete resection of schwannomas is easier as they usually have a true capsule, and their originating nerve stays at the periphery. On the contrary, neurofibromas are less prone to have a capsule and frequently englobe the nerve [12]. Malignant schwannomas can occur either as an evolution of a previously benign schwannoma, of a neurofibroma or de novo. Malignant transformation is extremely rare except in Von Recklinghausen's disease [13], where their incidence raises up to 10-15%. Until now, no case of malignant schwannoma from the nasal septum has been reported. Microscopically, signs of malignancy are focal hyperchromasia and dense packing with increased cellularity, indistinct cytoplasm, slender wavy or curled nuclei with numerous mitoses, anaplastic cells and invasiveness. IHC of Malignant schwannomas shows positive staining for S-100 antigen, Leu-17, vimentin and CD56 and negative staining for CK, LCA, CD34, SMA, desmin, chromogranin, and synaptophysin [14].

Genetics

Some genomic mutations associated with schwannomas are responsible for a group of genetic neurocutaneous disorders characterized by a predisposition to nerve sheath tumours called neurofibromatosis (type 1, type 2 and schwannomatosis) and Carney complex. Despite sharing some phenotypical similarities, their genotypical, molecular and clinical features distinguish those diseases. Nevertheless, schwannomas are often solitary and sporadic. An integrative genetic analysis of 125 vestibular and spinal schwannomas assessed their genomic basis [15]. It put in light that NF2 gene was altered or deleted in 76% of cases. In our patient, NF1 and NF2 didn't seem very likely. Indeed, given the absence of family history, her age and the full penetrance of these clinically diagnosed conditions, it seems reasonable to consider that these affections would already have been diagnosed. It was, however, impossible to tell if both schwannomas were the expression of a schwannomatosis trait, two unrelated sporadic events or a recurrence of the same incompletely resected lesion on her naso- palatine nerve, 28 years

later. Nevertheless, this seems quite long for a recurrence, even for these slow-growing tumours.

Treatment

The gold standard treatment for sinonasal benign schwannoma is surgery: a complete resection with safe margins. Nowadays, the endoscopic endonasal approach is the modality of choice in the majority of cases. In fact the lesions are frequently of moderate size and well circumscribed. In very extensive lesions with extra-nasal extension, an external approach can be considered. The dissection should include the muco-perichondrium or the mucoperiosteum [16]. In case of very extensive lesions, inoperable patient or malignant transformation radiotherapy can be considered even if schwannomas are typically radioresistant.

Follow-up

There are no definitive guidelines concerning the follow-up of these lesions in the literature due to their rarity. However, it is reasonable to follow up the patient by nasal endoscopy during the 3 first years following the surgery. An additional imaging (CT or MRI) can be necessary in extensive lesions or when nasal endoscopy is not conclusive. In the literature, no recurrence has been reported in case of complete resection with safe margins. No malignancy transformation was reported either.

Conclusion

Schwannomas are peripheral nerve sheath tumours seldom encountered in the sinonasal area and exceptionally on the nasal septum. A proper clinical and radiological assessment is of utmost importance to make a preoperative mapping of the tumour. CT and MRI are the best imaging modalities. As these tumours are well circumscribed, endoscopic endonasal surgery is nowadays the gold standard treatment in the majority of cases. Anatomopathology with IHC is essential to make a precise and definitive diagnosis. Neurofibromas and sarcomas are the most common differential diagnosis.

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