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LITERATURE REVIEW

Considerations on nerve tumors of the nasal fossae

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ABSTRACT

Nerve tumors of the nasal fossae are very rarely found in current practice. Tumors of the peripheral nerves can arise from the branches or roots of these nerves. There are two categories of nerve tumors: schwannoma and neurofibroma. They can be solitary or plexiform, and can hardly become malignant. If they are not diagnosed in time, they can reach large dimensions and cause major disorders in the nose and even in the paranasal sinuses. The diagnostic protocol is simple and precise, but the histopathological examination establishes the diagnostic of certainty. Treatment of nerve tumors of the nasal fossae is essentially surgical. The approach of the tumor must take into account its extension and the obligation of leaving no residual pathology in the nasal fossae.

KEYWORDS: nerve tumor, nasal fossae, schwannoma, neurofibroma, endoscopic surgery

INTRODUCTION

Although they occupy a relatively small anatomic region, the nasal cavity and the paranasal sinuses represent the site of origin of a complex group of tumors with a great histological variety. These include tumors that arise from the epithelium, seromucous glands, soft tissue, bone, cartilage, neuroectodermal tissue, hematolymphoid cells or from the odontogenic apparatus.

Nerve tumors of the nasal fossae are very rarely found in current practice. Diagnosis of certainty and histopathological form is determined by the histopathological examination.

Tumors of the peripheral nerves can arise from the branches or roots of these nerves. There are two categories of nerve tumors: neurilemmoma (schwannoma) and neurofibroma. Schwannomas are solitary and develop from the nerve sheath, which allows a relatively easy removal during surgery. On the contrary, neurofibromas tend to be multiple and develop from the endoneural substance. They are difficult to dissect and may be the sign of the Recklinghausen neurofibromatosis.

Neurofibromatosis or Von Recklinghausen disease is classified into two types:

• **Type I neurofibromatosis** - is a disease with autosomal dominant character, transmitted by chromosome 17. It is a neurocutaneous disorder, characterized by café-au-lait epidermal spots, Lisch nodules of the iris, mental retardation, sometimes hydrocephalus and presence of other tumors of the central nervous system.

• **Type II neurofibromatosis** - is transmitted by chromosome 22; it is also autosomal dominant, and neurofibromas are located on the acoustic nerve (VIII), usually bilaterally. Sometimes, they can be associated with meningiomas and astrocytomas.

Benign nerve tumors of the nasal fossae are very rare, representing about 5% of all benign tumors of the nose. They can be solitary or plexiform, and can hardly become malignant. If they are not diagnosed in time, they can reach large dimensions and cause major disorders in the nose and even in the paranasal sinuses.

Schwannoma develops from the Schwann cells of the peripheral nerves sheath. It is an uncommon tumor which can be found in any part of the body, in 25-45% of the cases being located in the head and the neck. Less than 4% of the schwannomas affect, in descending order of frequency, the ethmoid, the maxillary sinus, the nasal fossae and the sphenoid[1].

Rhinosinusal schwannomas develop in the ophthalmic and maxillary branches of the trigeminal nerve, but may also originate at the level of sympathetic fibers of the carotid plexus or of parasympathetic fibers of the pterygopalatine ganglion [2]. In case of large, extended tumors, their nervous origin cannot be identified.

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Symptoms presented by patients are nasal obstruction, rhinorrhea, epistaxis, anosmia, headache, hearing loss, facial or orbital tumefaction.

Neurofibromas are extremely rare in the sinonasal tract, even less frequent than schwannomas. In the case of neurofibromas associated with type II neurofibromatosis, patients are young, predominantly male[3]. In common type of neurofibromas, the disease can occur at any age, with an even gender distribution.

The tumor develops in the ophthalmic or maxillary branch of the trigeminal nerve and it is most commonly located in the maxillary sinus or ethmoid, and/or in the nasal cavity[4]. The plexiform neurofibroma can be found in the nasosinusal region, especially in the maxillary sinus associated with type I neurofibromatosis[5].

Symptoms include epistaxis, rhinorrhea, tumefactions, nasal obstruction and pain[6,7]. Most of the times, neurofibromas are diagnosed late, when they are of considerable dimensions and create difficulties in the therapeutic approach. Even though malignization is rare, the question usually arises about the symptoms caused by the tumor and its extension.

The **diagnosis protocol** for the nerve tumors of the nasal fossae is the following:

• **ENT clinical examination** which can reveal: in case of a schwannoma, a well-defined, non-encapsulated, globular, firm, yellow-brown tumor[8], with a network of capillaries on the surface, which may suggest a hypervascular lesion, or a firm, fusiform, submucosal, gray-brown tumor, sometimes with polypoid appearance and intact surface epithelium[6,9], in the case of neurofibromas. Sometimes, the tumor may be mistaken for nasal polyps, while other times, only aspiration of secretions and anemisation of the mucosa can offer a better picture of the tumor in the nasal fossa.

• **Rigid nasal endoscopic examination** provides details on tumor penetration, its extension, the relationship to the elements of the nasal fossae and, possibly, on the bone destruction caused by the tumor.

• **Imaging examination** and especially the cranio-facial CT scan with axial and coronal slices provide details regarding tumor extension and its relationship to the nasosinusal bone walls. In the case of schwannomas, the cranio-facial MRI reveals the histological features of the tumor. Thus, lesions with predominant Antoni A areas have intermediate signal intensity on both T1 and T2 images, unlike lesions with predominant Antoni B areas, corresponding to a myxoid stroma of low grade, characterized by hypersignal on T2[10,11].

• **Tumor biopsy and histopathological examination** of the sample establish the diagnosis of certainty of the tumorous form. I have to mention that biopsy can be performed preoperatively or examination of the sample completely removed from the nasal fossa can be made after resection. Immunohistochemical examinations may be also required.

Histopathological examination of the *schwannoma* (Figure 1) reveals densely cellular areas with wavy, fusiform, palisading nuclei, which alternate with loose, myxoid, poorly cellular areas. There are cellular Antoni A areas with Verocay bodies and hypocellular myxoid Antoni B areas. Tumor cells are strongly and diffusely immunoreactive for S100 protein. Moreover, one can observe areas of cystic degeneration and vessels with recent thrombosis. The *neurofibroma* (Figure 2) is like a benign tumoral proliferation with fusiform cells and wavy or comma-shaped nuclei. The stroma is myxoid and mitoses are absent. From the immunohistochemical point of view, the tumor is diffusely immunoreactive for S100 protein, but the proportion of positive cells is lower than in schwannoma.

Treatment of nerve tumors of the nasal fossae is essentially surgical. The approach of the tumor must take into account its extension and the obligation of leaving no residual pathology in the nasal fossae. I per-

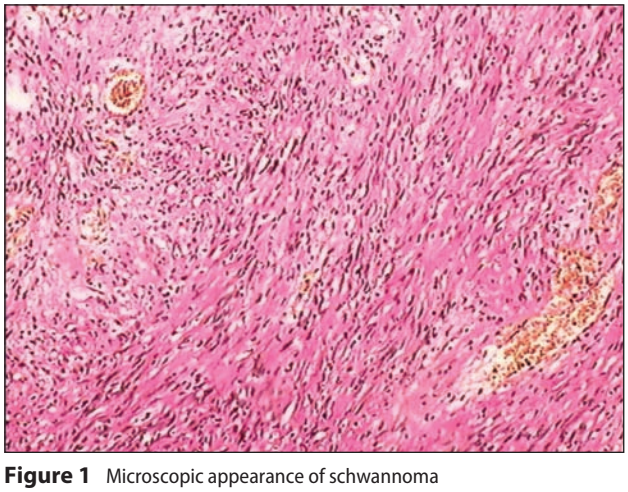

Figure 1 Microscopic appearance of schwannoma

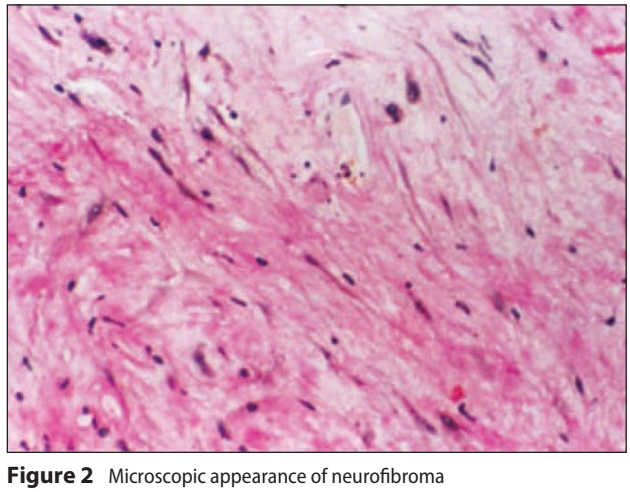

Figure 2 Microscopic appearance of neurofibroma

sonally have always opted for the endoscopic approach and, even if the tumor dimensions were larger, I practiced the so-called "piece meal resection", that is piece-by-piece resection of the tumor. The advantages are: the possibility of endoscopic visualization of the surgical area, which enables prevention of complications (CSF fistulas, damage to surrounding structures), faster patient recovery due to minimal surgical trauma, preservation of nasal functions, and last but not least, the aesthetic reasons.

Obviously, if tumor extension and the associated pathology require it, we will opt for a mixed or external approach.

We will refer to three cases from the personal casuistry, operated in the ENT & HNS Clinic, "Sfânta Maria" Hospital.

CASE PRESENTATION

The first case we refer to presented a large tumor of the right nasal fossa, which was occupying the entire upper third of the fossa, reaching and even destroying the posterior part of the septum and the anterior wall of the homolateral sphenoid sinus (Figure 3). The patient was prior diagnosed with nasal polyposis without histopathological examination (!!!). Nasal endoscopic examination revealed tumor penetration along the ceiling of the nasal cavity (sessile tumor), destruction of the posterior septum and penetration into the anterior sphenoidal wall. Despite the tumor size and due to the histopathological examination which showed that it was a solitary neurofibroma, endoscopic approach was chosen, that is "piece-meal resection" with complete ablation of the tumor.

The cranio-facial CT examination and the nasal endoscopy performed one month after surgery revealed healing, with no residual pathology.

The second case was an accidental finding. In a female patient with unilateral nasal obstruction, hypertrophy of the tail of the right inferior turbinate was diagnosed. Endoscopic examination revealed a pseudotumoral appearance of the inferior turbinate tail, which occupied 80% of the choanal orifice and extended up to the medium third of the nasal fossa. The cranio-facial CT examination revealed no other changes than occupation of the concha and extension in the medium third of the nasal fossa to the upper edge of the inferior concha. We performed complete ablation of the tumor together with the tail of the inferior nasal turbinate. Detachment from the inferior concha was easy, with minimal bleeding. The histopathological result revealed the presence of a schwannoma (Figure 4).

The third case presented with a right pansinusitis. The cranio-facial CT scan with contrast substance showed a change in tissue consistency in the posterior superior third of the nasal fossa and in the region of the middle meatus, as compared with the appearance of the sinus contents. Endoscopic nasal examination revealed, in the previously described region, the presence of a tumor covered by purulent secretions. Granulation tissue and oedematous polyps partially occupied the middle meatus, obstructing the infundibulum.

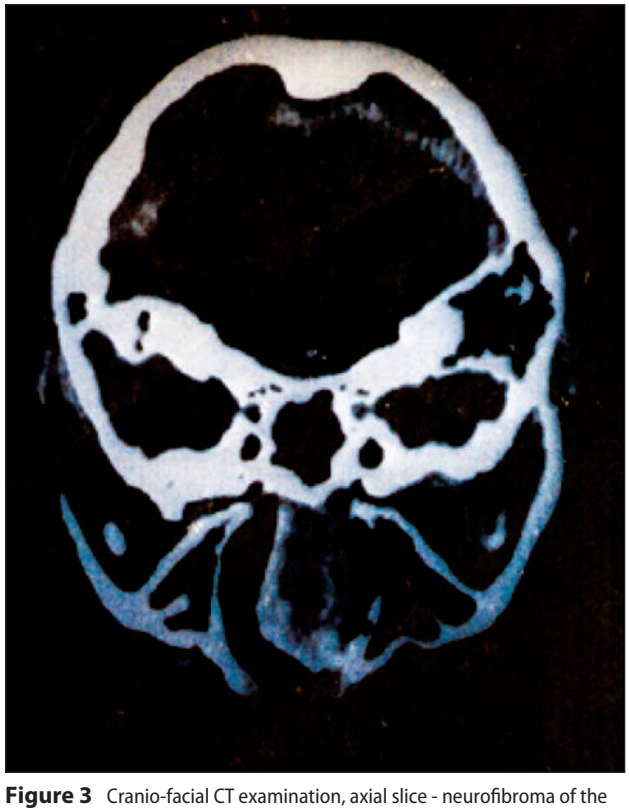

Figure 3 Cranio-facial CT examination, axial slice - neurofibroma of the right nasal fossa

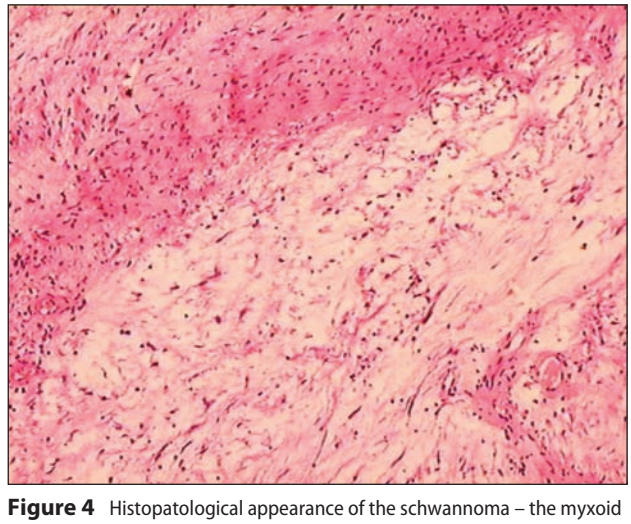

Figure 4 Histopatological appearance of the schwannoma – the myxoid area

We chose radical surgery of the maxillary sinus with drainage of a mucocele and endoscopic ablation (piece-meal resection) of the tumor. During the same surgical intervention, we also performed antero-posterior ethmoidectomy and wide-open drainage of the sphenoid sinus at the expence of the anterior wall. The content of these sinuses was similar to that of the maxillary sinus.

Histopathological examination showed the presence of a plexiform neurofibroma. It is interesting that the large dimensions of the tumor have led to an exudative polysinusitis, whose symptoms became dominant. Despite the large dimensions of the tumor, it could be approached endoscopically, but the old lesions of the maxillary sinus imposed a radical approach.

None of the operated patients showed clinical signs that could create any confusion with type I neurofibromatosis.

Endoscopic evaluation and CT scan performed three months postoperatively did not show any relapse or residual pathology. The average duration of hospitalization for the patients was 4 days. No intra or postoperative complications were recorded.

CONCLUSIONS

• Nerve tumors of the nasal fossae are very rare.

• The diagnostic protocol is simple and precise, but requires appropriate equipment.

• Histopathological examination establishes the diagnosis of certainty and of tumorous form. We recommend biopsy prior to surgery in order to operate knowing the accurate diagnosis.

• Endoscopic approach is preferable, but indication of this type of intervention must be properly determined. However, the advantages of endoscopy should not make us give up external approach when appropriate. It is preferable to perform a block resection of the tumor, but ablation of "piece meal resection" type provides undeniable advantages.

• Malignization of these tumors is rare. Complications of tumor extensions can be extremely dangerous. It is worth mentioning: erosion of bony septum or sinus walls, blocking drainage ostia with secondary sinusitis, vascular compressions or extension to neighboring regions.

• In order to diagnose a nerve tumor of the nasal fossae, we must first of all think of it!

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