

Unusual association of sphenoidal schwannoma with nasosinus polyposis

Abstract

Schwannomas or neurilemmomas are tumors of benign nerve sheath, and between 25 and 45% of all arise in the head and neck region. Less than 4 % are found in the sinonasal tract and their diagnosis may have difficulty due to non-specific symptoms and image characteristics. In several cases, complete surgical removal by endoscopic approach is the treatment of choice in these tumors. We report the management of an unusual case of sphenoid sinus schwannoma in a 70-years-old male along with massive bilateral nasal polyposis and severe symptoms. The association with severe sinonasal polyposis has not been previously described.

Keywords: schwannoma, neurilemmoma, sinonasal polyposis

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Oscar Álvarez Montero,¹ Marta Mancheño Losa,² María Antonia Rodríguez-Castejón,² Mayte Pinilla Urraca,¹ Margarita Gimeno-Arangüez³

¹ENT Department Puerta de Hierro Majadahonda University Hospital, Spain

²ENT Department Infanta Leonor U, Spain

³Histopathology Department Infanta Leonor U Hospital, Spain

Correspondence: Oscar Álvarez-Montero, ENT Department, Puerta de Hierro-Majadahonda University Hospital, Madrid, Spain, Calle Manuel de Falla, 1; 28222 Majadahonda, Madrid, Spain, Tel +34649821996, Email olamaontero@movistar.es

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Introduction

Schwannoma is a benign tumour originated from the sheath of myelinated peripheral nerves. The most frequent location is the VIII cranial nerve. Between 25 to 45% of schwannomas arise in head and neck region, and less than 4% are found in the sinonasal tract.¹ These arise from the Schwann cells in the ophthalmic and maxillary branches of the trigeminal nerve, in the III, IV and VI cranial nerves, in the autonomic nervous system, in the carotid plexus or sphenopalatine ganglion.¹ Whether sinonasal schwannomas can be derived from the olfactory nerve components is still a controversial issue,² but recent studies point more likely to its origin in the meningeal or ethmoidal branches of the trigeminal nerve.³ The most frequent locations in the sinonasal tract are: the ethmoidal sinus, maxillary sinus, nasal cavity and nasal septum. The sphenoid and frontal sinuses are rarely affected.¹

The non-specific signs and symptoms of sinonasal schwannomas are the same of those of any mass of the anterior or posterior nasal tract,¹ and their imaging findings are rather unspecific.^{4,5} Differential diagnosis should be made with other more common skull base tumours or space-occupying mass lesions such as sphenoid mucocoele, angiofibroma, inverted papilloma, clival chordoma, chondrosarcoma, meningioma, pituitary adenomas, craniopharyngiomas and plasmacytoma.^{4,5}

Complete surgical excision is considered the preferred treatment and most lesions may be removed by standard endoscopic techniques,^{1,6,7} although different approaches must be done if intracranial extension is present.^{1,7}

Here in we report an unusual case of sphenoidal schwannoma coupled with sinonasal massive polyposis, and we focus the role of computed tomography (CT) and magnetic resonance image (MRI) on the differential diagnosis and the surgical endoscopic approach.

Case report

A 70-year-old male was attended at the Emergency Department of our hospital, with nasal obstruction, hyposmia, severe headache,

ocular pain and diplopia. He had medical history of arterial hypertension, deep venous thrombosis and nasal polyposis in treatment with hydrochlorothiazide, acenocumarol and topical intranasal corticosteroids. There was no personal or family history of neurofibromatosis disease. The physical examination showed a VI left cranial nerve palsy. Non-contrast emergency CT revealed a complete sinonasal polyposis opacification with a 5 x 3.5cm sphenoid sinus expansive lesion. There was no calcification or bone destruction (Figure 1).



Figure 1 N=57; Epidemiological distribution of the pathological fractures, traumatic fractures, and nonunion.

The initial suspicion was sphenoidal mucocoele, and MRI was performed. The MRI showed an expansive mass of the 4.1 x 4.4 x 4.2cm sphenoid sinus expansive mass, heterogeneous on T2 weighted images and isointense on T1 weighted images, with intense heterogeneous contrast enhancement (Figure 2). The diagnosis of radiological suspicion was of chordoma or even macroadenoma.

The initial intravenous antibiotic (Clavulanic-Amoxicillin) and high doses steroid (methylprednisolone 1mg/kg of weigh per day)

treatment relieved the ocular pain, headache and diplopia. Then, following the resection of the polyps from the left nasal cavity, endoscopic biopsies of the mass were taken under general anesthesia in the operating theatre.

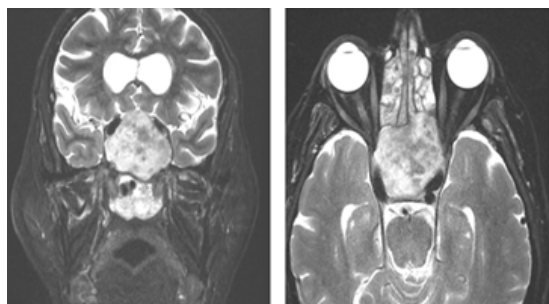


Figure 2 N=57; Epidemiological distribution of the pathological fractures, traumatic fractures, and nonunion.

The histopathology results showed a tumour with a fascicular pattern and elongated-undulating spindle cells in the myxoid matrix, nuclear polymorphisms and atypias, although without any abnormal necrosis or mitosis. No Verocay bodies were seen. The immunohistochemistry for S-100 protein showed strong nuclear and cytoplasmic reactivity, CD34 +, Actin -, EMA (epithelial membrane antigen) -, CEA (CD66e) - and Ki67 < 5%. The diagnosis was of a low-grade spindle-cell type of tumour in the peripheral nerve sheath, such as Schwannoma / neurofibroma but not a meningioma type (Figure 3).

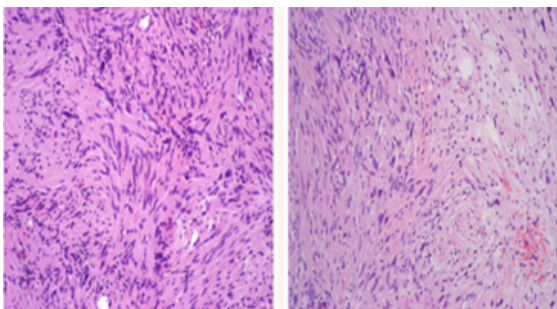


Figure 3 N=57; Epidemiological distribution of the pathological fractures, traumatic fractures, and nonunion.

The surgery was performed and, after the resection of the nasal polyposis, a bilateral sphenoidal approach was taken to perform a complete extradural resection of the lesion. We used a right pedicled nasoseptal flap (Hadad-Bassagasteguy flap(8)) to prevent any cerebrospinal fluid leaks. The final pathological examination confirmed the diagnosis of a benign schwannoma. After surgery, there was no any hormonal or neurological deficiency.

The patient has been disease-free, tumour and polyposis, for five years of follow-up (Figure 4 & 5).

Discussion

Sinonasal schwannomas usually present themselves with nasal obstruction, rhinorrhea, epistaxis, hyposmia or anosmia and pain.⁹ The poor specificity of these symptoms and slow progression of tumour often delays the correct diagnosis for about months to years, or until neurological symptoms began to occur. Diplopia secondary to, III, IV or VI cranial nerve palsy is a sign, which can allow us to suspect the diagnosis.¹ Other symptoms like exophthalmos, facial swelling and epiphora are less frequently observed.^{9,10}

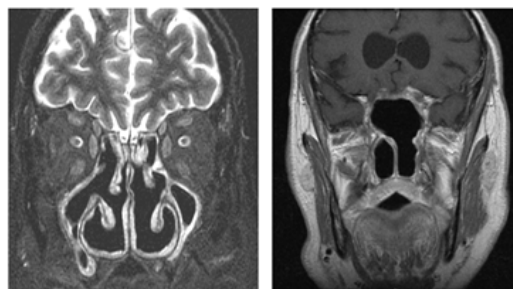


Figure 4 N=57; Epidemiological distribution of the pathological fractures, traumatic fractures, and nonunion.

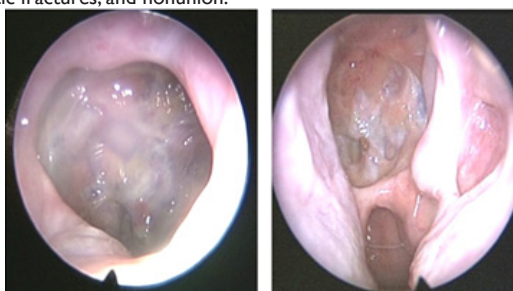


Figure 5 N=57; Epidemiological distribution of the pathological fractures, traumatic fractures, and nonunion.

Owing to their slow growth, radiological studies show an expansive soft tissue mass coupled with the preservation of most bony margins (thinning and / or swelling of the bony sinus walls).^{11,12} This finding can be helpful in differentiating schwannomas from other malignant tumours, such as chondrosarcoma, plasmacytoma, carcinoma, lymphomas, melanomas, etc., which tend to destroy the bone aggressively.

The lesions usually have a mottled central lucency with peripheral intensification on contrast enhanced CT. Heterogeneous appearance is related to areas of increased vascularity, cystic or necrotic regions.⁹ The isoattenuated compared with that of the brain stem and relatively homogeneous mild (masseter muscle) or marked enhancements on CT^{1,4} may provide the differential diagnosis in case of the most common causes namely expansive soft-tissue lesions on paranasal sinuses and mucocoele, which will not enhance at all.^{10,11}

Both, the MR imaging and CT scans show an intense heterogeneous contrast enhancement.^{9,11} The MRI may be useful in obtaining information about the nature of the lesion and in detecting any skull-base or intracranial issues¹⁰. Generally, neurilemmomas are isointense or hypointense on T1 -Weighted (T1-W) images and isointense or hyperintense on T2-W images.^{4,5}

Benign schwannomas are solitary and encapsulated masses attached to or surrounding their presumed nerve of origin, while neurofibromas are not encapsulated and may be multiple when seen with neurofibromatosis disease.⁵ Malignant transformation may occur in exceptional cases. Clival chordomas are slow growing tumours heterogeneously hyperintense or bright on T2-W images, and also show foci of haemorrhages as bright spots and dark areas on T1-W sequences, with marked contrast enhancement which may be heterogeneous.^{11,12} Preoperative correct judgement can aid to perform appropriate surgical approach.⁵ Schwannoma diagnosis can only be made by histopathological analysis.

The definitive treatment is the surgical complete excision. Endoscopic surgery provides a safe and minimally invasive treatment for these benign tumours. Surgical decompression followed by a radiosurgery is an acceptable option if there is unresectable.^{1,5,7} Recurrence is uncommon after total removal.¹⁰

The surgery can also involve a variety of combinations of lateral rhinotomy, Caldwell-Luc as well as external frontoethmoidectomy, trans-sphenoid approaches.^{5,10} Some extensive intracranial tumours require total transfacial or combined transfacial and intracranial approaches.¹⁰

Conclusion

Skull base issues caused by sphenoid sinus tumours can produce III, IV and VI cranial nerve palsies, facial and ocular pain, cephalgia and even loss of vision or other neurological symptoms. The clinical and radiological differential diagnoses include several types of benign and malignant neoplasms and the final diagnosis must be confirmed by the histological and immunohistochemical analysis. The association with massive bilateral polyposis is exceptional. Complete surgical resection is the only curative treatment for schwannomas, and an endoscopic approach may be useful in selected cases.

Acknowledgments

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Conflicts of interest

Author declares there are no conflicts of interest.

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