

Sinonasal Schwannoma: A Case Report and Updated Review of the Literature

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Abstract

Sinonasal schwannomas are rare, benign and slow-growing peripheral nerve sheath tumors without age, race, or gender predilection. We present the unusual case of a 59-year-old female who presented with right-sided retro-orbital pressure and rhinorrhea. Radiographic imaging revealed a mass centered in the right superior nasal cavity with extension through the cribriform plate into the right more than the left subfrontal region. Biopsy confirmed the diagnosis of a sinonasal schwannoma, which was resected via nasal endoscopic surgery and ethmoidectomy. These tumors, albeit rare, should be considered in the differential diagnosis for patients presenting with a paranasal sinus mass to ensure inclusion of this benign lesion and avoid unnecessary aggressive intervention.

Keywords

Neurilemmoma, Paranasal Sinus, Sinonasal Tract, Schwannoma

1. Introduction

Schwannomas, also known as neurilemmomas, are benign and slow-growing peripheral nerve sheath tumors composed of Schwann cells. Approximately 25% - 45% of schwannomas occur in the head and neck, and most frequently involve the vestibular division of cranial nerve VIII [1] [2]. Only about 4% of head and neck schwannomas arise in the sinonasal cavity. These lesions are typically asymptomatic until they grow large enough to

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exert a mass effect on surrounding structures, thereby producing clinical symptoms. Occasionally, these lesions can undergo malignant transformation. Surgical resection is the treatment of choice [2] [3]. We present a case of a relatively uncommon sinonasal schwannoma with symptoms of headache, eye pain, and visual disturbances successfully treated with right nasal endoscopic surgery and ethmoidectomy.

2. Case Report

A 59-year-old female presented with a 3-month history of right retro-orbital pressure and rhinorrhea. She denied fever, cough, and changes in vision or smell. After failing conservative therapy for presumed sinusitis, a computed tomography (CT) scan of the sinuses was obtained. Contrast-enhanced CT demonstrated a rounded soft tissue mass with mild patchy enhancement centered within the right superior nasal cavity. There was secondary benign bony remodeling and dehiscence of the cribriform plate and lamina papyracea with intracranial extension (**Figure 1**). Magnetic resonance imaging (MRI) obtained to further characterize the lesion demonstrated a well-circumscribed $2.9 \times 2.4 \times 2.2$ cm mass centered at the junction of the right anterior ethmoid air cells and frontal sinus with extension through the cribriform plate into the right more than left subfrontal region, abutting the right inferior frontal gyrus and obscuring the right gyrus rectus (**Figure 2**). This mass appeared hyperintense to brain on T2-weighted images and markedly enhanced on T1-weighted images following gadolinium administration. A biopsy was performed, which revealed an S-100 positive, epithelial membrane, antigen-negative tumor, consistent with schwannoma. The patient underwent a right nasal endoscopic surgery and ethmoidectomy with an uneventful postoperative course and no evidence of recurrence on follow-up MRI.



Figure 1. Sinonasal schwannoma. Unenhanced coronal (a) maxillofacial CT image demonstrates a right ethmoidal sinonasal schwannoma with soft tissue attenuation. Enhanced coronal (b) and axial (c) images show heterogeneous enhancement of the mass. Unenhanced sagittal (d) CT shows intracranial extension of the mass with surrounding benign bony remodeling.

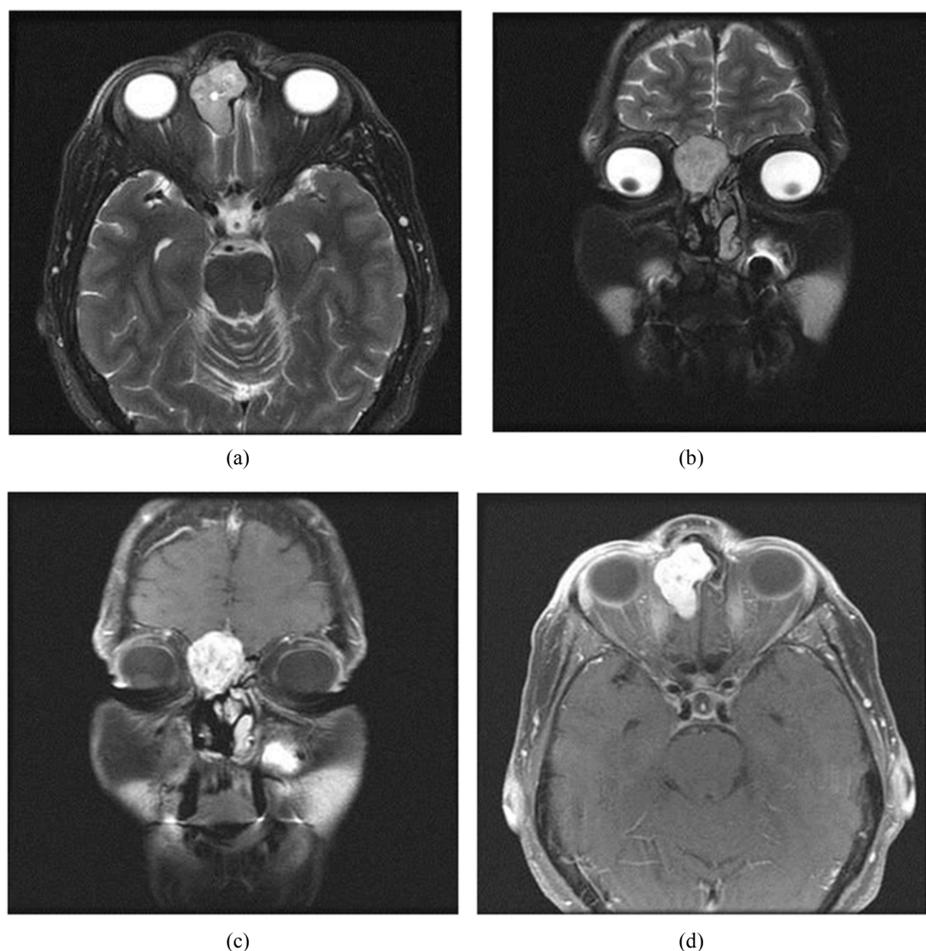


Figure 2. Right ethmoidal sinonasal schwannoma. Axial (a) and coronal (b) T2-weighted facial MRI images demonstrate a hyperintense mass centered in the right anterior ethmoid air cells. The mass characteristically enhances intensely on post-gadolinium axial (c) and coronal (d) T1-weighted images. Biopsy confirmed the diagnosis of sinonasal schwannoma.

3. Discussion

Sinonasal schwannomas are uncommon tumors that have only been described in case reports or small case series. These masses arise from the ophthalmic and maxillary branches of the trigeminal nerve or from autonomic nerves that innervate the septal vessels and mucosa [4]. Kim *et al.* showed that these lesions involve the nasal cavity and ethmoid sinus more frequently than other paranasal sinuses, likely due to the greater abundance of regional nerve innervations [5].

Sinonasal schwannomas have no age, race, or gender predilection. Common presenting symptoms include nasal obstruction, nasal discharge, and anosmia. Extension into the cavernous sinus may cause diplopia and retro-orbital pain. Epistaxis is another commonly-reported symptom, which may complicate the clinical picture by raising the possibility of a juvenile nasal angiofibroma [3]. Other differential diagnostic considerations for a superior nasal cavity mass include angiomatous polyps, squamous cell carcinoma, esthesioneuroblastoma, sinonasal undifferentiated carcinoma, lymphoma, adenocarcinoma, meningioma, neurofibroma, and hemangioma [2] [6].

The CT and MRI features of sinonasal schwannomas are generally nonspecific and may be insufficient for establishing a definitive diagnosis [5] [7]. Nevertheless, initial imaging with CT provides valuable information in the workup for nasal masses. Sinonasal schwannomas appear as unilateral, isoattenuating, tubular soft tissue masses with mild to marked enhancement with iodinated contrast. As demonstrated in our case, when present, secondary bony changes have a benign appearance with remodeling and smooth erosion often associated with

intracranial and/or intra-orbital tumor extension. Sinonasal schwannomas usually lack internal calcifications or blood products. The tubular configuration of the mass is thought to be a result of growth along the alignment of the ethmoid lamellae and is a feature that may help distinguish this lesion from other sinonasal malignancies [5]. On MRI, sinonasal schwannomas are well-defined lesions with intermediate T1 and variable T2 signal intensity, as well as homogenous intense enhancement following gadolinium contrast administration [5] [6]. While these imaging features are non-specific, the well-defined margins, secondary osseous remodeling, lack of internal calcification or blood products, and homogeneous intense enhancement suggest a benign process rather than a more commonly-occurring sinonasal malignancy. Furthermore, imaging delineates the lesion's intracranial and intra-orbital extension required for operative planning.

The imaging characteristics of other paranasal sinus lesions are broad. However, several sinonasal masses, including angiomatous polyps, meningiomas, carcinomas, and neurofibromas typically demonstrate soft tissue attenuation on CT with enhancement after iodinated contrast administration. On examination with MRI, these masses often have low signal intensity on T1WI and variable signal intensity on T2WI [8]-[11]. Sinonasal lymphoma possesses similar imaging findings with a tendency towards homogeneous high T2 signal intensity. Carcinoma and lymphoma demonstrate increased fluorodeoxyglucose (FDG) uptake on positron emission tomography (PET) imaging. These lesions also cause aggressive bony destruction, whereas benign lesions such as meningiomas and angiomatous polyps typically incite benign bone remodeling. Juvenile nasopharyngeal angiofibromas, which are benign vascular lesions, produce broadening of the pterygopalatine fossa from which they arise, with associated enlargement of the sphenopalatine foramen and Vidian canal on CT. On MR imaging, these lesions demonstrate flow voids and intense enhancement given the high vascularity [10].

Biopsy and pathologic examination is critical for confirming the diagnosis of schwannoma given the overlapping features with other sinonasal lesions [12]. These tumors are composed of uniformly-shaped spindle cells with variable cellularity. A combination of cellular proliferation and loosely-structured myxomatous regions known as Antoni A and Antoni B areas, respectively, is characteristic of the lesions [13]. Sinonasal schwannomas are highly cellular with a predominance of Antoni A areas, which contain tightly-organized nuclear palisades called Verocay bodies [13]. Immunohistochemistry staining plays an important role in accurate diagnosis. Sinonasal schwannomas demonstrate intense, diffuse S-100 protein immunoreactivity [14] [15]. These tumors may also demonstrate positive staining for neuron-specific enolase and vimentin and negative staining for epithelial membrane antigen (EMA), glial fibrillary acidic protein, and CD56 [3]. The tumor in our patient was S-100 positive and EMA negative.

Surgery is the treatment of choice for sinonasal schwannomas. Traditionally, transcranial and transfacial techniques were used to remove these lesions. However, these approaches now tend to be reserved for schwannomas that have undergone malignant transformation. Endoscopic and microscopic endonasal surgery approaches have recently demonstrated excellent outcomes for resection of benign paranasal sinus tumors [2] [3] [16]. These procedures have the benefit of lower morbidity, no external incision, and shorter hospital stay [17]. Radiotherapy may be considered for adjuvant therapy for malignant nerve sheath tumors [18]. Our patient underwent minimally-invasive nasal endoscopic surgery and ethmoidectomy without complication.

4. Conclusion

Although the sinonasal region is an uncommon location for schwannomas to present, they should be considered in the differential diagnosis of a benign-appearing nasal cavity mass. Radiographic imaging, in combination with histopathologic examination, is critical for characterizing sinonasal schwannomas and guiding treatment, which is often minimally-invasive surgery.

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