

Atypical Presentation of Sinonasal Cellular Schwannoma: A Nonsolitary Mass with Osseous, Orbital, and Intracranial Invasion

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Abstract

Objective Sinonasal cellular schwannoma represents < 4% of head and neck schwannomas. These benign tumors are typically confined to the nasal cavity or ethmoid sinus. We describe an atypical case of sinonasal cellular schwannoma with diffuse paranasal sinus involvement and both intraorbital and intracranial extension.

Results A 62-year-old woman presented with a 6-month history of right orbital proptosis and right-sided headache. Subsequent imaging revealed an invasive paranasal sinus mass extending through the skull base and displacing the right orbit. Preoperative biopsies were not diagnostic but revealed a spindle cell lesion suspicious for malignancy based on lack of encapsulation, infiltration of the sinonasal submucosa, and osseous invasion. The patient underwent open skull base surgery, and pathology confirmed a S100-positive nonencapsulated cellular schwannoma.

Conclusion An atypical case of sinonasal cellular schwannoma with intracranial extension is reported. Its presentation is contrary to the common view that these are isolated solitary lesions of the nasoethmoid region. We suggest that sinonasal cellular schwannoma be considered in the differential diagnosis of a poorly defined invasive paranasal sinus mass, particularly following biopsy.

Keywords

- ▶ cellular schwannoma
- ▶ nasal cavity
- ▶ sinuses
- ▶ paranasal
- ▶ intracranial

Background

Schwannoma is a benign tumor originating from the Schwann cell of the neural sheath, and 25 to 45% occur in the head and neck.¹ Although rare, these tumors may originate in the sinonasal tract. Cellular schwannomas of this area represent

< 4% of head and neck schwannomas; only an estimated 100 cases of this type have been reported in the literature.^{2,3}

Sinonasal schwannomas cause a variety of clinical symptoms that depend on the location and size of the mass.⁴ Patients often present quite late because there is significant room for the tumor to grow in the air-filled sinonasal tract.⁴

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Published descriptions of the radiologic appearance of these benign tumors indicate that they present as a well-demarcated solitary soft tissue mass, most commonly in the nasal cavity or ethmoid sinus.^{4,5} Over time, constant pressure by the mass on the surrounding structures may lead to bone remodeling and erosion.⁶

Cellular schwannoma was first described in 1981 as an uncommon but well-recognized variant of schwannoma.⁷⁻¹² The most common sites of occurrence include the posterior mediastinum and the retroperitoneum. Like conventional schwannomas, these are typically, but not always, encapsulated. Unlike conventional schwannomas, cellular schwannomas demonstrate worrisome clinical and histologic features including bone erosion and both increased cellularity and mitotic activity. Prior to the recognition of this subtype of schwannoma, approximately a third of cellular schwannomas were diagnosed as malignant based on these aggressive features.⁷⁻¹¹

We describe the presentation, radiologic findings, and treatment of a patient with a sinonasal cellular schwannoma and its aggressive clinical and histologic features. The patient presented with a poorly demarcated diffuse tumor infiltrating all of the ipsilateral paranasal sinuses with concomitant intracranial and orbital extension. Details of the initial presentation, diagnostic assessment, and treatment are highlighted here.

Case Report

A 62-year-old woman presented with a 6-month history of right orbital proptosis and right-sided headache. She denied diplopia or any change in vision. The headache was described as circumferential and constant, without any positional association or aura. Past medical history included bilateral retinitis pigmentosa, hip replacement, and tubal ligation. She was not taking any medications, and there was no history of smoking or relevant occupational exposures.

Clinical evaluation revealed a 5-mm proptosis and a 6-mm outward and a 2-mm inferior displacement of the right eye. There was slight under action with both upward and downward gaze. Visual acuity was recorded as 20/400 bilaterally, related to the retinitis pigmentosa history. The remainder of the cranial nerve examination was normal. Examination of the oral cavity and neck was unremarkable. Intranasally, anterior rhinoscopy revealed a fleshy mass originating from the right middle meatus surrounded by purulent secretions. This was substantiated with nasal endoscopy. Cranial computed tomography and magnetic resonance imaging demonstrated an invasive right paranasal sinus mass with intracranial and orbital extension (►Fig. 1). There was dural involvement.

An initial biopsy revealed a highly cellular spindle cell lesion, likely of peripheral nerve sheath origin, with normal mitotic rate and no vascular invasion. A definitive diagnosis could not be made, but suspicion for malignancy was relatively high based on the lack of encapsulation, infiltration of the sinonasal submucosa, and invasion into surrounding bone and tissue. A second biopsy revealed the same findings. At the first clinical visit, the patient was placed on a 14-day course of amoxicillin-clavulanic acid given the purulent discharge, which completely relieved the headache and resolved the purulent discharge.

The patient underwent an open skull base procedure, and the mass was resected. This involved a bifrontal craniotomy, bilateral tarsorrhaphy, right lateral rhinotomy, partial maxillectomy, sphenoidectomy, posterior nasal septectomy, right dacryocystorhinostomy, right orbital roof and anterior cranial floor resection followed by right orbital roof reconstruction with a split calvarial graft, and a bilateral pericranial flap for closure of the anterior cranial floor (►Fig. 2). The patient was discharged home 1 week following surgery without event. Following surgery, the patient received postoperative radiation therapy due to the extent of invasion and aggressive nature of the tumor, as well as close intracranial margins.

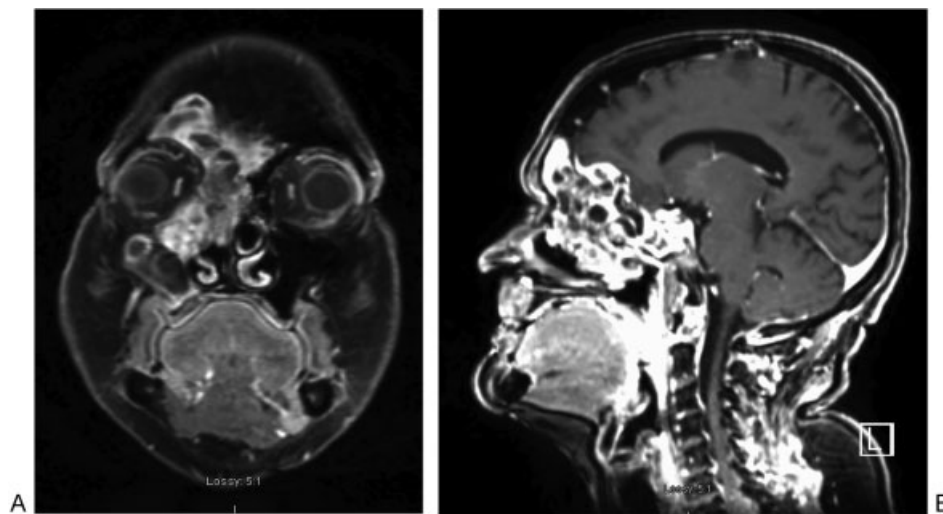


Fig. 1 Magnetic resonance imaging (MRI) demonstrating the extent of the paranasal sinus mass. (A) Coronal T1-weighted MRI with contrast demonstrating right orbital displacement without radiologic evidence of periorbital invasion. (B) Sagittal T1-weighted MRI with contrast demonstrating anterior-posterior tumor limits and intracranial extension.

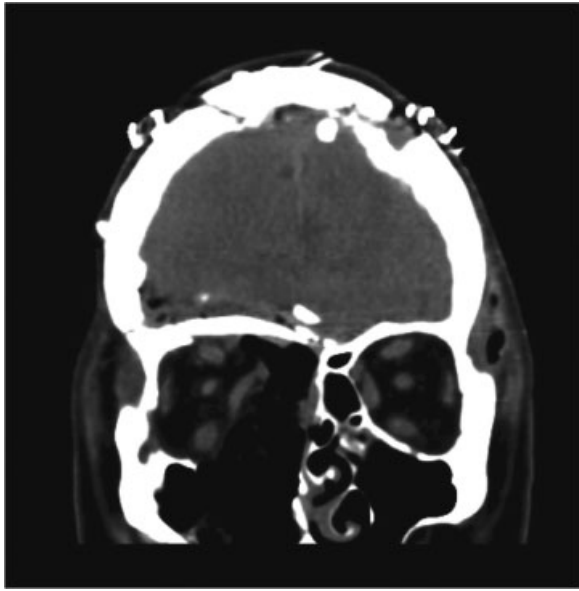


Fig. 2 Postoperative coronal computed tomography demonstrating resection of the tumor and orbital roof reconstruction.

There was no clinical or radiologic evidence of tumor recurrence 12 months postresection.

Final pathologic examination confirmed the diagnosis of cellular schwannoma. Microscopic examination revealed a nonencapsulated, highly cellular spindle cell lesion infiltrating the sinonasal submucosa and demonstrating bone invasion. No significant atypia was identified. There was no necrosis and the mitotic rate was quite low, in comparison

with the cellularity of the lesion, with approximately one to two mitoses per 10 high-power fields. The lesion stained strongly and diffusely positive for S100, and Ki-67 (proliferation index marker) highlighted 6.7% of tumor cells (1,000 cells counted) (►Fig. 3).

Discussion

Head and neck schwannomas rarely occur in the sinonasal cavities, and they are typically described as solitary encapsulated lesions confined to the nasoethmoid region.^{1-5,13-17} The less common cellular schwannoma has been described in the sinonasal tract, and the lack of encapsulation along with bony erosion of this variant was previously noted.^{11,16} Although schwannomas are uncommon in this location, and cellular schwannomas even more so, the recognition of this entity in this anatomical site is important.^{1-5,13-17}

The reported case reiterates the potential risk of rendering a malignant diagnosis on biopsy, particularly in light of the aggressive radiographic findings. Although bone erosion is well described in cellular schwannomas at a variety of anatomical locations, it appears that those arising in this anatomical site tend to have the additional aggressive feature of being nonencapsulated with an infiltrative growth pattern. These findings, along with the rarity of this lesion in this location, increase the likelihood that a malignant diagnosis may be rendered.

This case was a diagnostic challenge, which was demonstrated by a difference in initial biopsy pathology and post-excision pathology findings. Initial biopsies revealed a spindle

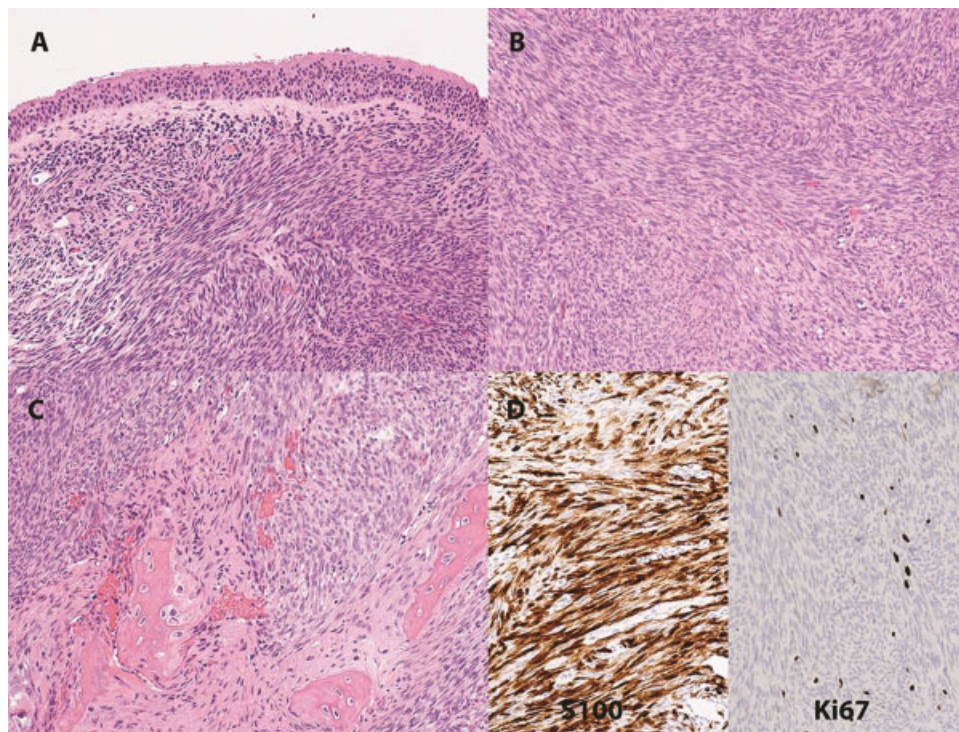


Fig. 3 (A) Low-power view ($\times 40$) of a highly cellular spindle cell lesion arranged in sweeping fascicles, infiltrating and expanding the submucosa with intact overlying sinonasal mucosa. (B) Medium-power view demonstrating long sweeping fascicles of uniform spindle cells. (C) Tumor with embedded bony spicules due to erosion/infiltration into adjacent bone. (D) (left) Tumor cells demonstrate strong, diffuse S100 immunoreactivity; (right) Ki-67 highlighted 6.7% of tumor cells (1,000 cells counted), and only one to two mitoses per 10 high-power fields were identified.

Table 1 Microscopic and immunohistochemical features to distinguish among conventional schwannoma, cellular schwannoma, and malignant peripheral nerve sheath tumor

	Conventional schwannoma	Cellular schwannoma	Conventional MPNST
Microscopic features	Antoni A (hypercellular) and Antoni B (loose/hypocellular) areas; thick-walled hyalinized blood vessels; no/rare mitoses	Mainly hypercellular Antoni A areas; cells may be hyperchromatic with or without pleomorphic; thick-walled hyalinized blood vessels; mitoses typically ≤ 4 per 10 high-power fields	Markedly hypercellular spindle cells in fascicular pattern, cells of uniform size and hyperchromatic; geographic necrosis and mitoses > 4 per 10 high-power fields; some may have epithelioid cells and some may have heterologous elements
S100 immunostaining	Strong diffuse staining	Strong diffuse staining	Scattered positive cells in ~ 50 – 70% of cases; can be strongly positive in epithelioid variant of MPNST

Abbreviation: MPNST, malignant peripheral nerve sheath tumor.

cell neoplasm that was suspicious for malignancy due to the lack of encapsulation, the infiltration of the sinonasal submucosa, and tumor invasion into bone. Although these findings were still present in the final postexcision specimen, the lack of malignant histologic features, such as necrosis, mitoses, and significant atypia, along with strong, diffuse S100 immunoreactivity confirmed the diagnosis of an unencapsulated cellular schwannoma.

Of particular significance in this case was the presence of bone infiltration and erosion with subsequent intracranial tumor extension. Bone erosion has been described in cellular schwannoma but typically as a pushing front into the adjacent bone.¹⁰ In this case, the tumor appeared to infiltrate the adjacent bone more aggressively, with bone spicules present within the tumor mass. There are very few reported cases of sinonasal cellular schwannoma with intracranial extension, and thus our case is an important addition to the literature.

Classic schwannomas typically contain a capsule believed to be derived from perineurium of the native nerve.³ Although there are many possibilities for the nerve origin for sinonasal schwannomas, this cellular schwannoma likely originated from sinonasal mucosal autonomic nerves because these nerves lack a perineurial layer.^{2,3,15} The nonencapsulated nature of this tumor accounted for its more infiltrative and aggressive nature.

Furthermore, given the rarity of this pathology, it is important to differentiate correctly between a conventional and a cellular schwannoma when reporting in the literature. This will allow us to better understand the clinical behavior and treatment results from the management of this pathology in the future. Histologic criteria to differentiate schwannoma, cellular schwannoma, and malignant peripheral nerve sheath tumor (MPNST) were previously described and are summarized in **Table 1**.¹¹ Using these criteria, we identified one publication with misclassified tumors.³ The histologic findings listed in Mey et al suggest that cases 1 to 4 are cellular schwannomas rather than conventional schwannomas based on the predominance of the Antoni A pattern, little to no Antoni B pattern, and mitoses and/or increased Ki-67.³ Case number 5 in the same article labeled as a malignant transfor-

mation of a schwannoma, is most likely an epithelioid MPNST, based on the decreased S100 staining (compared with cases 1–4) and atypical areas identified in **Fig. 2** in both the primary and the recurrence. The distinction between conventional schwannomas versus cellular schwannomas, which may have malignant clinical features, is more clearly distinguished in the pathology literature. For instance, cellular schwannoma involving the paranasal sinuses was actually first described in the publication by Hasegawa et al and not by al-Otieschan et al.^{11,16} Given the difference in clinical behavior between these two pathologies, close collaboration with a pathologist would be helpful for correctly reporting cases of schwannoma in the future.

The reported patient received postoperative external-beam radiation therapy given the close intracranial resection margins. The treatment of a conventional schwannoma is typically surgical excision.¹ However, given the aggressive and diffusely infiltrative nature of the cellular schwannoma reported in this case (which differentiates it from conventional schwannoma), adjuvant radiotherapy was administered. The use of adjuvant radiotherapy was reported in the literature for other patients with cellular schwannoma but with numbers too small to provide data on efficacy.¹⁰

Conclusion

This case of sinonasal cellular schwannoma is important because its clinicopathologic features are easily mistaken for a malignant entity. It should be emphasized that with this pathology, patient presentation may be variable and that biopsy and imaging alone do not fully outline the invasive potential of these tumors. Therefore, we suggest that sinonasal cellular schwannoma be considered in the differential diagnosis of a poorly defined invasive paranasal sinus mass.

Note

This material has never been published and is not currently under evaluation in any other peer-reviewed publication.

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