Olfactory Groove Schwannoma:
A Case Report and Literature Review

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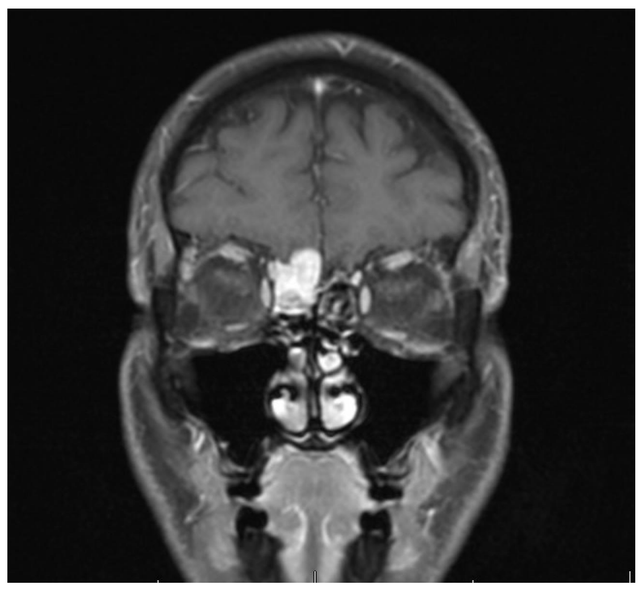
Introduction

Schwannomas are benign, slow-growing tumors which derive their name and origin from Schwann cells and occur most frequently in the cranial nerves. The **optic and olfactory nerves, however, lack Schwann cells and so CN I or CNII schwannomas are therefore not possibl**e. This case demonstrates **an intra-axial schwannoma** of the olfactory groove extending into the sinus cavity.

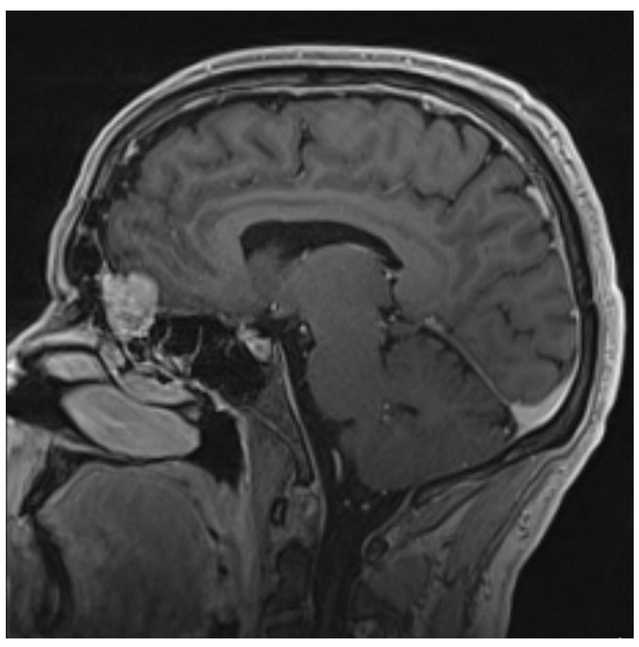
Clinical Case

A 56-year old male presented to our rhinology clinic for evaluation of a known intra-axial anterior skullbase mass that had been present for at least five years, previously managed with clinical monitoring and interval imaging at an outside hospital.

Surveillance MRI demonstrated an expansile mass approximately 2.1 x 1.4 cm in size centered over the cribriform plate with expansion into the ethmoid sinus.



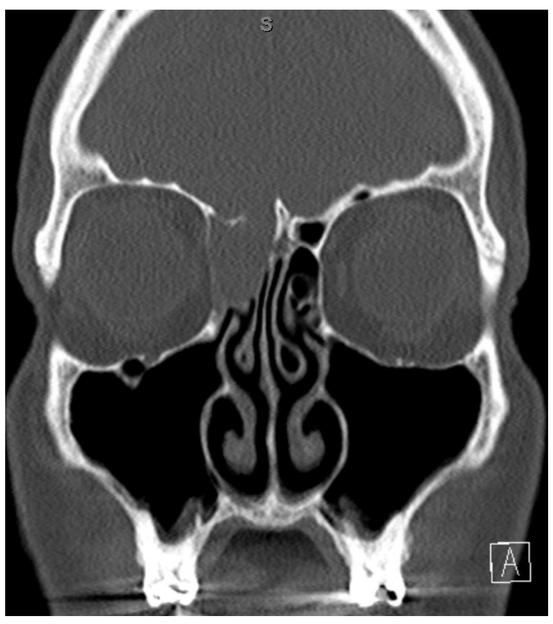
Preoperative surveillance MRI with contrast, coronal



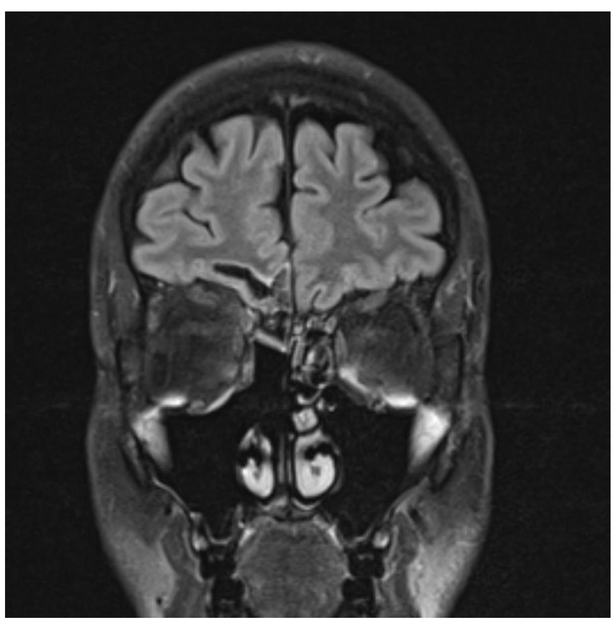
Preoperative surveillance MRI with contrast, sagittal

Given interval growth of the mass as compared to prior imaging, biopsy was performed in the operating room.

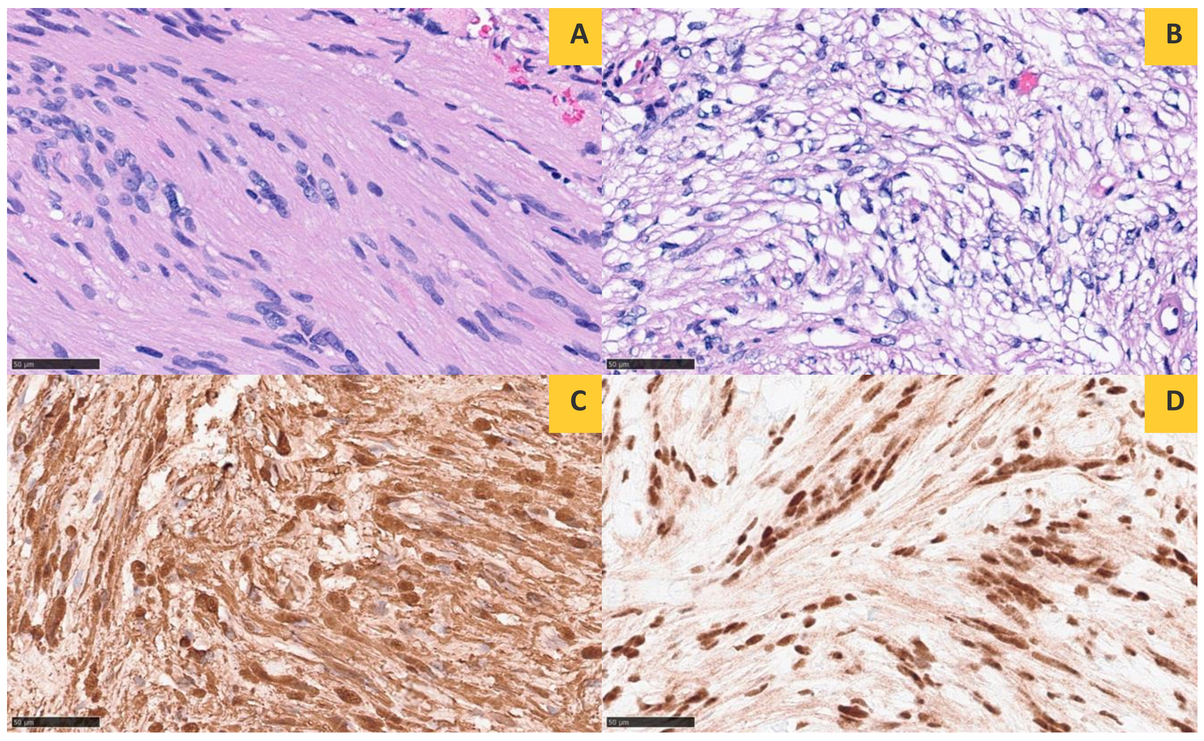
Histopathologic evaluation demonstrated biphasic tumoral proliferation. Cells are elongated & wavy, with interspersed collagen fibers. Immunohistochemistry stains showed that cells were diffusely positive for S100 while INI-1 is retained. According to these observations the mass was defined as a schwannoma, CNS WHO Grade I.



Preoperative CT without contrast, coronal



Postoperative surveillance MRI with contrast, coronal



A. Hypercellular area (Antoni A area) with elongated and wavy tumor cells with interspersed collagen fibers and Verocay bodies (H&E x400). B. Hypocellular and microcystic area (Antoni B area) with hyalinized blood vessels in upper left and lower right corners (H&E x400). C. Tumor cells are positive for S100 immunohistochemistry (×400). D. Tumor cells show retained INI-1 expression (×400).

Discussion

Schwannomas of the anterior cranial fossa, and specifically of the olfactory groove, are exceedingly rare tumors, **with less than 100 cases described in the literature**. The pathogenesis of olfactory groove schwannoma is not fully known, though it is generally agreed upon that given the lack of Schwann cells of CNI, the olfactory bulb is not the nerve of origin.

The most plausible sites of origin, rather, are the nearby structures whose myelin sheath is produced by Schwann cells. These include:

- The dural branches off of the first division of the trigeminal nerve
- The filia olfactoria (which connect olfactory bulb to olfactory epithelium and acquire Schwann cells 0.5 mm beyond the olfactory bulb)
- The nervus terminalis

Less obvious hypotheses based in developmental pathogenesis suggest the growth of schwannoma from **Schwann cells atypically present** in the central nervous system (CNS) due to aberrant differentiation of neural crest cells or multipotent mesenchymal cells. However, these theories are more likely to explain intraparenchymal than extraparenchymal schwannomas.

Definitive diagnosis consists of histopathologic evaluation revealing **spindle cells** in either Antoni A (palisading spindle cells with elongated nuclei and fibrillary cytoplasm) or Antoni B (loose myxoid stroma with small amount spindle cells) configuration; these cells stain strongly with S-100 on immunohistochemistry.

Treatment consists of complete surgical excision. This patient underwent an uninarial endoscopic endonasal trans-cribiform approach for resection with a fascia button graft and nasoseptal flap for closure. MRI completed 4 months postoperatively was unconcerning for residual or recurrent tumor.

Conclusions

Olfactory groove schwannomas are benign, rare tumors often presenting nonspecifically, making them difficult to diagnose without tissue sample. **They should be considered in the differential diagnosis of sinonasal masses with cribriform plate involvement.**

Contact

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