

Olfactory groove schwannoma masquerading as an orbital mass

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ABSTRACT

يعتبر الورم الشفاني بحز شمي ورم نادر جداً خلال قاعدة الجمجمة والحجاج. قد يمتد الورم أيضاً إلى منطقة تحت الفص الأمامي داخل الأم الجافية. أجريت عملية جراحية لحالة مريضة تبلغ من العمر ٣٥ عاماً مصابة بورم شفاني في المنطقة من الحز الشمي الأيسر مع بروز في الجانب الأيسر وازدواجية الرؤية ووجود كتلة فوق الحجاج وامتداد داخل الجمجمة. وجود قشرة العظام حسب ما تبين في التصوير الطبقي وغياب الذيل السحائي حسب ما تبين في التصوير بالرنين المغناطيسي هو في صالح التشخيص. كشف الفحص للأنسجة والتقييم الكيميائي النسيجي المناعي للبروتين إس-١٠٠ أن الورم هو ورم شفاني. يعتبر الاستئصال الكامل خلال ثقب الجمجمة من مقدمتي الجبهة وإعادة بناء قاعدة الجمجمة خطوات العلاج الرئيسية.

Olfactory groove schwannoma is a very rare tumor breaking through the skull base and orbit. The tumor may also extend to the subfrontal region intradurally. A case of olfactory groove schwannoma in a 35-year-old female, originating from the left olfactory groove with left proptosis, diplopia, supra-orbital mass, and gross intracranial extension is reported. The presence of bony scalloping on CT and absence of meningeal tail on contrast enhanced MRI are in favor of the diagnosis. Histological examination and immunohistochemical evaluation for S-100 protein, showed the tumor as a schwannoma. Total excision through a frontal craniotomy and skull base reconstruction are the main therapeutic steps.

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Intracranial schwannoma has a predilection for sensory nerves, particularly vestibular division of the eighth nerve, followed distantly by the fifth nerve and even more remotely by hypoglossal, olfactory, and facial nerves. Rare schwannomas arise from the parenchyma of the cerebrum especially the frontal lobe.¹ Intracranial schwannomas not arising from the main cranial nerves account for less than 1% of surgically treated schwannomas of the central and peripheral nervous system. Subfrontal schwannomas are rare and less than 20 cases have been reported in the literature. These tumors commonly take origin near the transition zone between the peripheral nervous system with its Schwann cell sheath, and central division with its oligodendroglial covering.² Here, we present a rare frontal lobe schwannoma, lying in the olfactory groove and left orbit, originating unusually from the left anterior ethmoidal nerve, in order to explain other possible pathogenetic mechanisms for the lesion.

Case Report. A 35-year-old lady presented due to seizures, diplopia, and headache within the past 5 months. She had an enlarging para-midline, supraorbital swelling for 2 years, followed by progressive left sided anosmia and diplopia. On examination, there was a soft medial canthal swelling below the superior orbital rim causing mild proptosis, shifting the left eye downward and forward. Fundoscopic examination showed moderate to severe bilateral papilledema. On bone window CT-scan, the mass had produced left medial orbit and olfactory groove scalloping without any apparent bone destruction, along with effacement of the posterior wall of the frontal sinus (Figure 1). On MRI, the mass was hyper intense heterogeneously on T2 and hypo intense on T1 weighted images, with fluffy non-homogenous enhancement and a non-enhanced center. There was no dural tail, and the mass was expanding from crista Gali to the tuberculum sella (Figure 2). The tumor separated the frontal lobes and filled the inter-hemispheric fissure displacing the corpus callosum without any peri-lesional edema or prominent feeding artery. Through a bicoronal scalp flap, bifrontal craniotomy was performed. The tumor had invaded the posterior wall of frontal sinus, coming down to the left supra-orbital rim and roof, piercing the dura at the cribriform plate. The tumor

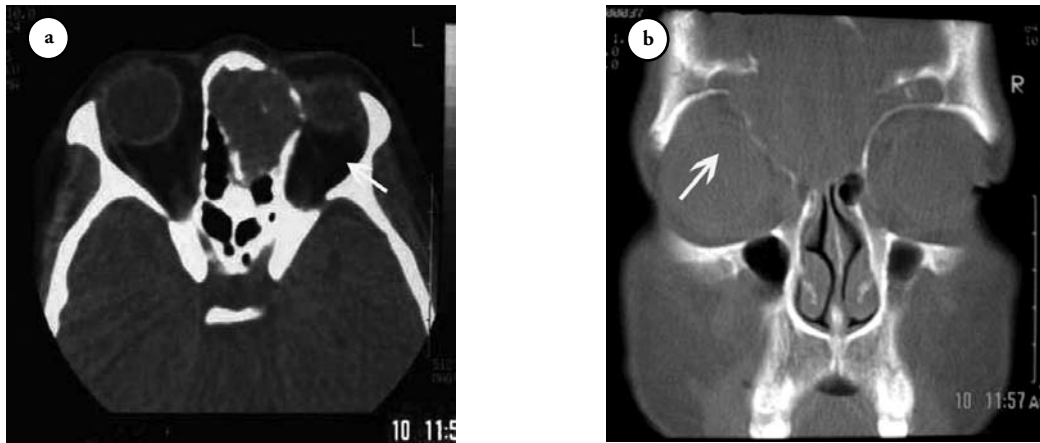


Figure 1 - Computed tomograms of the anterior skull base with bone window depicting the mass in the cribriform plate with prominent bone scalloping a) axial view, b) coronal view.

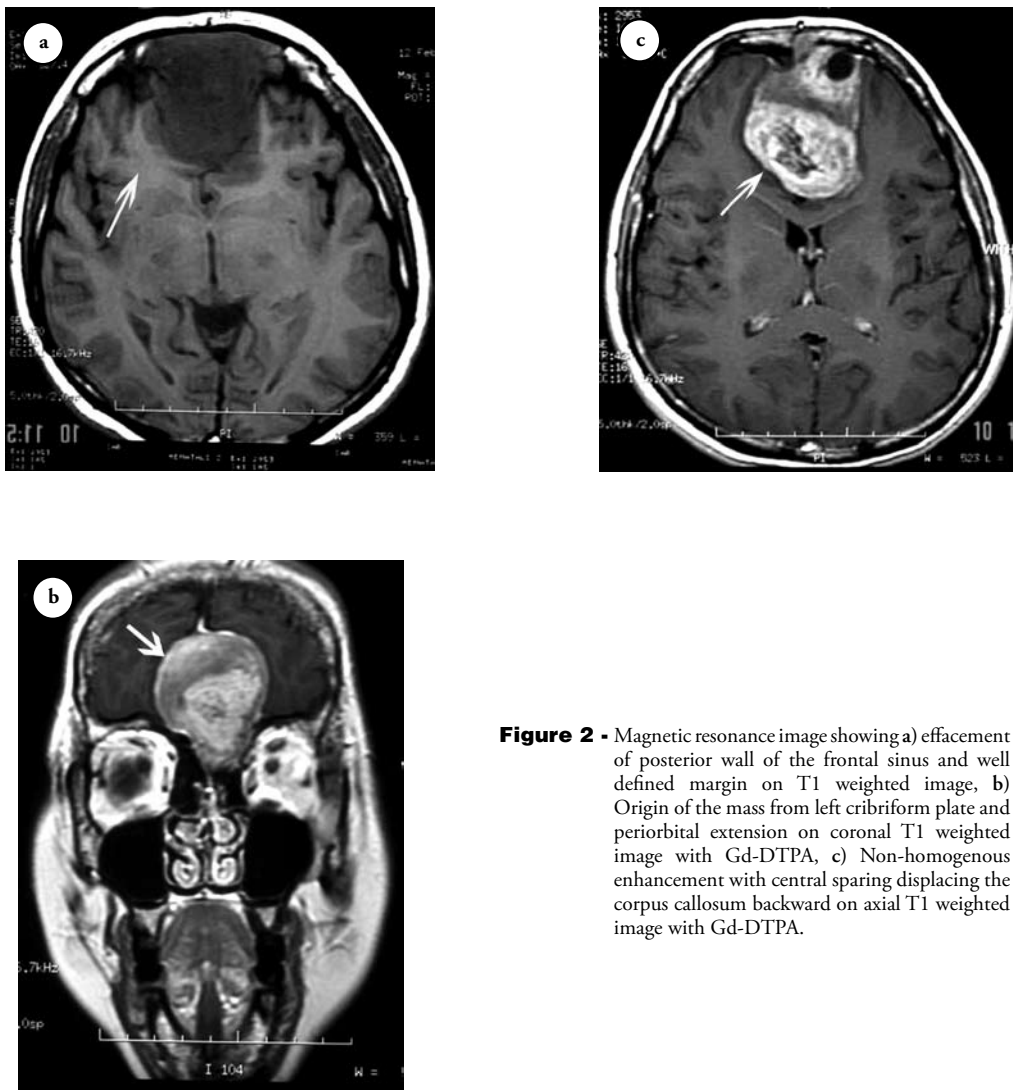


Figure 2 - Magnetic resonance image showing a) effacement of posterior wall of the frontal sinus and well defined margin on T1 weighted image, b) Origin of the mass from left cribriform plate and periorbital extension on coronal T1 weighted image with Gd-DTPA, c) Non-homogenous enhancement with central sparing displacing the corpus callosum backward on axial T1 weighted image with Gd-DTPA.



Figure 3 - Post-operative coronal enhanced MRI, revealing total removal and innocent faint pial enhancement.

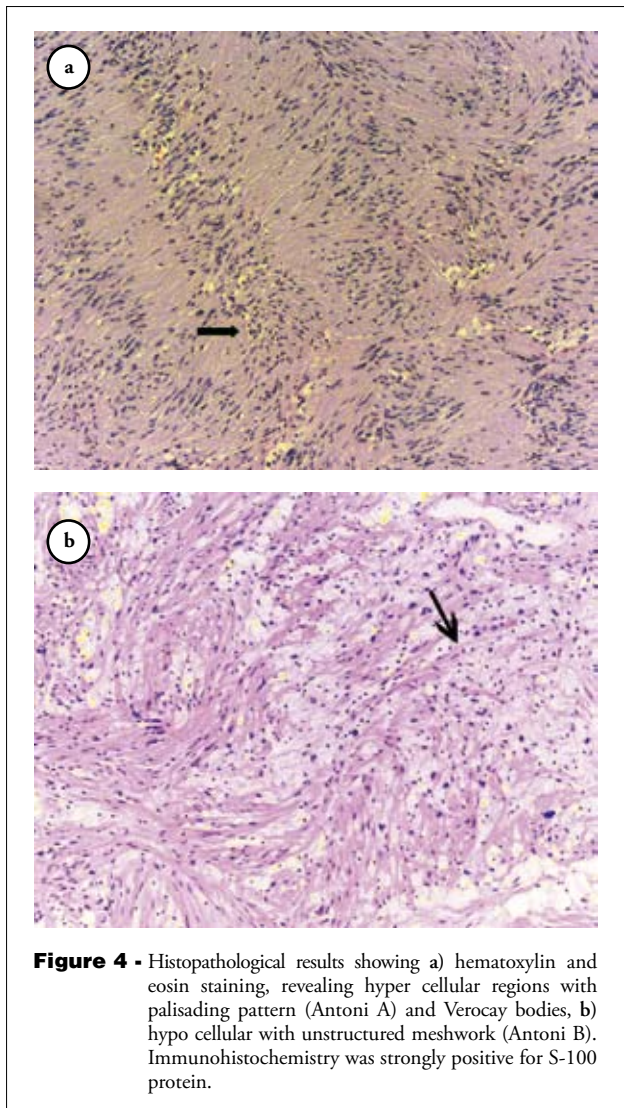


Figure 4 - Histopathological results showing a) hematoxylin and eosin staining, revealing hyper cellular regions with palisading pattern (Antoni A) and Verocay bodies, b) hypo cellular with unstructured meshwork (Antoni B). Immunohistochemistry was strongly positive for S-100 protein.

was thoroughly dissected from the corpus callosum in the inter-hemispheric region. Olfactory tracts were intermingled with the mass and not identifiable. Total removal was achieved, leaving a cavity at the midline replacing the cribriform plate. The resultant dural and bony defects were reconstructed by a peri-cranial sliding flap, and the bone harvested from the inner table of the frontal bone flap. The post-operative period was uneventful, except for transient rhinorrhea, which stopped with lumbar puncture and oral acetazolamide. In the contrast enhanced MRI carried out a month later, no residual tumor was observed (Figure 3). Histological examination showed highly cellular parts in the tumor consisting of spindle cells with elongated oval nuclei forming palisades (Antoni A) along with Verocay bodies and pauci-cellular portions containing unstructured meshwork of loosely disposed cells (Antoni B) (Figure 4). Immunohistochemical staining was positive for S-100 protein and negative for epithelial membrane antigen, characteristic of schwannoma. The patient is now doing well, but still traces of diplopia and anosmia are present.

Discussion. Schwannomas commonly originate from sensory nerves with a preponderance for the schwannoglial junction. Optic and olfactory nerves are devoid of schwann cells, therefore, schwannomas do not develop from these nerves theoretically.³ The pathogenesis of these tumors may be explained on the basis of developmental transformation of mesenchymal pial cells into ectodermal Schwann cells, and migration of the neural crest cells, or non-developmental tumorigenesis from normal Schwann cells in the perivascular plexus and meningeal branches of the anterior ethmoidal nerve.² In our patient, the origin of the tumor in the supero-medial aspect of the orbit strengthens the latter hypothesis. However, it has been speculated that some cells in the olfactory nerve may be phenotypically similar to Schwann cells, giving origin to tumor expressing histological characteristics of schwannoma.⁴ Although the classic histology of schwannoma is common in peripheral and intra-spinal tumors; intra-cranial schwannomas typically lack the prominent palisades and are composed of tissues of intermediate organization. As described before, our patient had a very typical schwannoma originating from left anterior ethmoidal nerve, which is a peripheral branch of the trigeminal nerve.

Some studies have reported these tumors to be heavily vascularized on angiogram or intra-operatively;¹ however in our case, the tumor was fairly vascularized. Presence of a prominent nutrient vessel on contrast enhanced studies may warrant pre-operative angiograms. Schwannosis in the olfactory region is a

hamartomatous lesion consisting of Schwann cells as a result, displaced neural crest being often associated with von Recklinghausen's disease or as a reaction to head injury.⁵ However, some authors have reported a history of cranial radiation in their patients, possibly contributing to the pathogenesis.⁶ Our patient did not have any stigmata of neurofibromatosis,¹ nor a history of a significant head injury or previous radiation.

The male to female ratio reported in different studies is approximately 3/1,² the average age at diagnosis has been approximately 30 years,³ our case was a 35-year-old female. Reported clinical features of subfrontal schwannoma include headache, seizure, anosmia, and increased intracranial pressure.⁵ Our patient also presented with these symptoms. She had total anosmia, while some studies have reported preservation of contra-lateral olfaction, probably with smaller tumors.² Mucocoeles of the frontal sinus give rise to proptosis and may extend intra-cranially in the epidural space. Symptoms of raised intra-cranial pressure and cranial nerve involvement are not common, in mucocoeles, while being present in our patient. Esthesioneuroblastoma and metastatic tumors usually show paranasal sinus invasion, and extensive bony destruction on CT scan,⁵ contrary to the findings of our patient. Total excision

through a bifrontal craniotomy, without any adjuvant therapy is the only plausible remedy. Unilateral and/or trans-nasal or trans-ethmoidal approaches have been associated with incomplete removal of the tumor.⁴

Olfactory groove schwannomas are extremely rare neoplasms of the midline anterior skull base and a high index of suspicion is required for a preoperative diagnosis and proper treatment.

References

1. Amador AR, Santonja C, Del Pozo JM, Ortiz L. Olfactory schwannoma. *Eur Radiol* 2002; 12: 742-744.
2. Shenoy SN, Raja A. Cystic olfactory groove schwannoma. *Neurol India* 2004; 52: 261-262.
3. Murakami M, Tsukahara T, Hatano T, Nakakuki T, Ogino E, Aoyama T. Olfactory groove schwannoma. *Neural Med Chir (Tokyo)* 2004; 44: 191-194.
4. de Souza HL, Ramos AM, Ramos CC, Melo SP, Pereira HS, Madureira JF, et al. Olfactory groove schwannoma. *Arg Neuropsiquiatr* 2003; 61: 125-128.
5. Prak JH, Kim TY, Park JT, Kim JM. Olfactory groove schwannoma. *J Korean Neurosurg Soc* 2006; 39: 156-158.
6. Sano H, Hayashi Y, Hasegawa M, Yamashita J. Subfrontal schwannoma without hyposmia. *Neurol Med Chir (Tokyo)* 2004; 44: 591-594.

CASE REPORTS

Case reports will only be considered for unusual topics that add something new to the literature. All Case Reports should include at least one figure. Written informed consent for publication must accompany any photograph in which the subject can be identified. Figures should be submitted with a 300 dpi resolution when submitting electronically or printed on high-contrast glossy paper when submitting print copies. The abstract should be unstructured, and the introductory section should always include the objective and reason why the author is presenting this particular case. References should be up to date, preferably not exceeding 15.