Skull Base Meningioma: Delayed Extracranial Presentation

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ABSTRACT

Meningiomas are common intracranial tumors that rarely extend to extracranial sites. When they occur extracranially near the skull base, their diagnosis and management represent tremendous challenges. Although the extracranial component tends to manifest at the same time as the intracranial component, this is not always the case. In the patient reported in this article, 13 years separated the diagnosis of the intracranial tumor from the appearance of an extracranial component. The clinical presentation, radiologic findings, and management of this large skull base meningioma are discussed with reference to the literature. A high index of suspicion and clinical acumen are needed to ensure that the extracranial component of these frequent tumors is discovered, especially when it presents later than the intracranial component.

KEYWORDS: Meningioma, multidisciplinary, extracranial

Meningiomas are common intracranial tumors that rarely extend to extracranial sites. The prevalence of extracranial extension in patients presenting with intracranial meningiomas has been reported by Shuangshoti¹ and Farr et al.² to be 9% and 20%, respectively. The most frequent sites of an external mass are the orbit, the outer table and scalp, the paranasal sinuses and nasal cavity, and the parotid and parapharyngeal region.² When they occur in the head and neck, their diagnosis

and management often present many challenges. For example, the inconclusive pathology of small biopsy specimens from anomalous sites creates confusion with other more common tumors of the head and neck.² A high degree of clinical suspicion is therefore necessary to diagnose extracranial meningiomas correctly. Most often, the extracranial component is detected during the initial evaluation of the intracranial lesion. We report our experience with a large olfactory groove meningioma

Skull Base, volume 11, number 2, 2001. Reprint requests: Anthony G. Zeitouni, M.D., McGill University Health Center, 687 Pine Avenue West, Suite E4-46, Montreal (QC) Canada, H3A 1A1. E-mail: azeitouni@hotmail.com. Departments of ¹Otolaryngology and ²Neurosurgery and ³Division of Plastic and Reconstructive Surgery, McGill University Health Center, Montreal, Quebec, Canada. Copyright © 2001 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2001,11,02,105,108,ftx,en;sbs00211x.

with delayed extracranial extension. The clinical presentation, evolution, and management of this tumor are discussed with reference to the literature. Craniofacial reconstruction details are also included.

CASE REPORT

A 49-year-old man presented with new-onset frontal headaches. Computed tomography (CT) demonstrated a mass filling the right anterior cranial fossa that was resected through a right frontal craniotomy (Fig. 1). It recurred 10 years later, and the patient underwent two more resections. Pathology continued to show a benign meningioma. There were no en plaque features. Radiology showed no involvement of the sinuses, even at the time of the patient's surgeries. He was followed clinically at regular intervals and follow-up CT scans were obtained.

The patient was referred to our institution 13 years after his initial presentation with proptosis of the left eye and diplopia. The investigation included three-dimensional (3D) reformatted CT

Figure 2 MRI demonstrating extracranial recurrence in the frontal sinus.

and magnetic resonance imaging (MRI). Intracranially, residual meningioma was present along the dura. However, most of the meningioma was located extracranially. Extensive bilateral involvement of his sinuses and left orbit by the meningioma was demonstrated (Figs. 2, 3). Surgical access was achieved by a bicoronal incision combined with a lateral rhinotomy incision. A sub-

Figure 1 CT scan demonstrating large meningioma in anterior cranial fossa, but not in frontal sinus.

Figure 3 The tumor involved the ethmoid, frontal, and maxillary sinuses.

frontal approach was selected. The tumor had destroyed the walls of the frontal sinuses and the region of the glabella down to the nasal bones. The cribriform plate region was deemed free of gross disease. Although good exposure was obtained with the lateral rhinotomy and subfrontal approaches, rigid sinus endoscopes were used at the end of the case to ensure that all traces of the disease had been removed. Craniofacial reconstruction involved a rectus abdominus free flap to fill the defect and a large parietal cranial bone graft to reconstruct the entire frontal bar in one piece. Hydroxyapatite cement (Bonesource, Leibinger, Richardson, TX) was added to the adjacent smaller bony defects. Good postoperative contour was achieved (Fig. 4). After surgery, the patient underwent radiotherapy.

DISCUSSION

Prolonged follow-up management and clinical acumen are necessary to diagnosis a recurrent meningioma, especially if it occurs extracranially in a delayed fashion. This case demonstrates that meningiomas not only occur in extracranial sites, but that they can do so in a delayed fashion, years after resection of the intracranial component.

Meningiomas account for 13 to 18% of all brain tumors³ and are believed to arise from meningothelial or arachnoid cells. As a result, the most common sites of meningiomas are intracranial, particularly in the parasagittal meninges, falx cerebri, and sphenoid ridge.^{2,4} Extracranial meningiomas usually develop as an extension of an intracranial meningioma (secondary extracranial meningioma). The tumor can spread through the foramina of the skull, as Farr and others have described,^{2,3,5,6} entering the orbit through the supra-orbital fissure, the nasopharynx and nasal cavity through the cribriform plate, and the paranasal sinuses and pterygoid region through the floor of the middle cranial fossa and suture lines of the skull.

Figure 4 Despite extensive resection of the frontal bone and glabella, the reconstruction with a free flap, cortical bone, and hydroxyapatite cement resulted in good contour.

Alternatively and less commonly, meningiomas arise from an extracranial site with no evidence of intracranial involvement (primary extracranial meningioma).⁷ The pathologic features of extracranial meningiomas are identical to those of more frequent intracranial lesions.² Meningiomas may be divided grossly into globular and en plaque tumors. The globular type is more common and usually evokes symptoms of increased intracranial pressure.⁸ The en plaque type produces symptoms by encasing nerves and vessels as it grows along the skull base.⁸ It tends to invade underlying bone^{8,9} and is more difficult to eradicate. This case presentation shows, however, that globular tumors can recur at extracranial sites.

Although benign, meningiomas of the skull base are locally invasive and have a high recurrence rate.⁸ If the tumors are allowed to grow, they progressively erode the skull base and produce significant neurological morbidity. Three factors influence the recurrence rate: extent of surgical removal, tumor pathology, and location. Total surgical removal at an early stage is the recommended therapy for most patients^{5,7,10,11} but is not always feasible because of vital structures located at the skull base.

Meningiomas at the skull base have the highest recurrence rate of all intracranial meningiomas, probably because their poorly accessible location and their proximity to vital structures make total surgical excision difficult and often impossible.^{10,11} As a result, the recurrence/progression rate probability in one series of skull base meningiomas at 5 and 10 years was 34% and 54%, respectively.¹¹ No noted relationship to age, sex, or duration of symptoms predicts tumor recurrence or the progression rate.¹²

COMMENTS FOR PUBLICATION

In this report, Al-Sebeih and coworkers present the unusual case of a recurrent meningioma primarily located extracranially. The authors review the literature on extracranial meningiomas and emphasize the unusual finding of a primarily extracranial extension with delayed presentation. They state that a high index of suspicion is needed to ensure that the extracranial component of these tumors is discovered.

The concept of extracranial extension of meningiomas is well known. Likewise, the tendency for meningiomas to recur after a delay is also well described. This case report represents a combination of these two features of meningiomas in the same patient. The extracranial recurrence in this case was not clinically "occult." However, it is

conceivable that periodic routine imaging might identify an extracranial recurrence before patients become symptomatic. From this perspective, the report supports the role of periodic imaging to follow patients who have had a meningioma resected, a practice that we employ at our institution.

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