Cases

Large oculomotor schwannoma presenting as a parasellar mass: A case report and literature review

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Abstract

Background: Large schwannomas arising from the oculomotor nerve are very rare. The common site of tumor occurrence in this nerve is the segment within the interpeduncular cistern and the cavernous sinus.

Case description: We report a case of a large left-sided oculomotor nerve schwannoma with minimal clinical signs and symptoms of oculomotor nerve involvement resembling a large parasellar mass. The radiological features of the mass were more consistent with a medial sphenoid wing meningioma causing brain stem compression. Complete resection of the tumor was achieved via a left pterional approach. The patient developed complete third nerve palsy postoperatively.

Conclusion: The management of these large benign tumors with brain stem compression includes surgical resection. Intraoperative anatomical preservation of the third nerve was impossible given its course in the tumor. We discuss the pertinent literature and management of large oculomotor schwannomas.

Key Words: Meningioma, oculomotor schwannoma, skull base

INTRODUCTION

Schwannomas constitute about 7% of all intracranial tumors and commonly arise from the vestibulocochlear and trigeminal nerves. Motor nerve schwannomas arising from the oculomotor nerve are very rare. Approximately 30 cases of oculomotor nerve schwannomas have been described in the literature, of which 11 cases were described as large (≥2.5 cm). The management of these large tumors is especially challenging given the relationship of these tumors to the cranial nerves and the brainstem. Here we report a large oculomotor schwannoma and review the management of these uncommon tumors.

Case description

A 38-year-old female developed symptoms of headache, dizziness, occasional diplopia and drooping of the left eyelid. Imaging confirmed a large left suprasellar mass (3.5 cm in diameter) with midbrain compression, suggesting a meningioma [Figures 1, 2 and 3]. A biopsy via a right pterional approach at an outside institution showed a benign spindle-cell neoplasm. The material was too small to definitively distinguish between meningioma and schwannoma. No tumor was resected at this operation. At MDACC her examination confirmed a bi-temporal hemianopsia related to previous surgery. There were no clinical signs of oculomotor palsy. The preoperative diagnosis in our patient was a meningioma,
given the paucity of clinical findings. A left pterional craniotomy, drilling of the sphenoid wing and wide splitting of the Sylvian fissure for tumor access was carried out. The tumor was well encapsulated. After substantial debulking of the mass, the third nerve appeared from the interpeduncular cistern as a bundle and ran straight into the base of the mass. No other significant attachments of the tumor were recognized. Postoperatively, the patient developed complete third nerve paralysis on the left side. At 6-month follow-up, she continued to have complete third nerve palsy with no evidence of residual tumor [Figures 1, 2 and 3].

Pathologic description: Multiple rubbery fragments of tumor aggregated to a mass about 3.5 cm in diameter. Multiple histologic sections showed typical appearance of a benign schwannoma with short-spindled cells and random nuclear pleomorphism [Figure 4a]. Most of the tissue was the denser, more organized, Antoni A type, with very few Antoni B areas. No Verocay bodies were seen. Focal perivascular hemorrhage was identified. Occasional histiocytes and small aggregates of mature lymphocytes were sometimes present around blood vessels [Figure 4b]. A few blood vessels showed mural hyalinization. Immunohistochemistry for S-100 protein showed diffuse strong nuclear and cytoplasmic reactivity in the schwannoma [Figure 4c]. Because of the consideration of meningioma, an immunohistochemical stain for epithelial membrane antigen was performed but was entirely nonreactive [Figure 4d].

DISCUSSION

Schwannomas arising from the ocular nerves are extremely uncommon, unless associated with neurofibromatosis. Kovacs,6 who in 1927 described an isolated oculomotor nerve schwannoma observed during an autopsy, was probably the first to report such a tumor. There are approximately 32 cases of well-documented oculomotor nerve schwannomas15 in the literature, of which 11 cases described were large (2.5 cm or larger in diameter). These large tumors, because of their location, present unique diagnostic and surgical challenges to the neurosurgeon and will be the focus of this discussion. These large tumors typically present as a parasellar or suprasellar mass and can mimic a medial sphenoid wing or a posterior clinoidal meningioma. There are no radiological findings in these large parasellar tumors that can reliably distinguish schwannomas from meningiomas. Generally these large tumors are misdiagnosed as meningiomas preoperatively, given the paucity of clinical signs and symptoms of third cranial nerve involvement, as seen in our case [Table 1].

Of the 12 patients (including our patient), there were 9 females and 3 males with a median age of 41.5 years. Celli et al.31 divided oculomotor cranial nerve schwannomas
into three groups, i.e., 1) cisternal, 2) cisternocavernous and 3) cavernous lesions. This classification was based on the preferred extension of these tumors. All the cases described except 2 had a large cisternal extension based on imaging and operative findings. The explanation for this could be that the ventral cistern of the brainstem is a potential space where these slow-growing tumors expand and can also cause brainstem compression. Five (42%) patients presented with signs of hemiparesis as a result of brainstem compression. The cases described by Oakamoto et al. and Barat et al. had intraorbital extension along with a large parasellar component. In both these cases, exophthalmos was the presenting symptom. Ten patients presented with some ocular signs or symptoms, with 4 (33%) out of 12 patients presenting with a mild ptosis. The ocular findings surprisingly were minimal, given the large size of the tumors [Table 1]. Multiple cranial nerve deficits were seen in only 1 case described by Barat et al. involving the fourth, fifth and sixth nerves. This tumor, however, had a large intracavernous component.

Seven (58%) out of 12 patients underwent a complete resection, 4, a partial or subtotal resection, and in 1 patient, the resection status was not specified. Of the 7 patients who underwent a complete resection, 6 (86%) patients had complete third nerve palsy following surgery. Niazi and Boggan described a case where the tumor was resected completely after the second operation (4 years apart). The patient, however, only experienced incomplete third nerve palsy.

Hiscott and Symon describe a tumor with a large cisternal component, and at the end of the resection, describe the third nerve as “a stout bundle as it appeared from the interpeduncular cistern and ran straight into the mass, spread out and disappeared.” We
Table 1: Summary of histologically verified large (≥ 2.5 cm) oculomotor schwannomas as described in the literature

| Author, Year       | Age (yrs), Sex | Preoperative symptoms/ signs                                    | Tumor size (cm) | Extent of resection | Location along third nerve | Postoperative third nerve deficits |
|--------------------|---------------|---------------------------------------------------------------|-----------------|---------------------|----------------------------|----------------------------------|
| Broggi and Franzini, 1981 | 45, M         | Hemiparesis, central seventh nerve palsy                      | 3a              | NS                  | NS                         | NS                               |
| Hiscott and Symon, 1982  | 58, F         | Headaches, drowsiness, hemiparesis, minimal ptosis            | 4a              | Total               | Cisternal                  | Complete third nerve palsy       |
| Luenda et al., 1982    | 11, M          | Headaches, hemiparesis, impaired upward gaze                  | 5.5a            | Total               | NS                         | Third nerve palsy                |
| Oakamoto et al., 1985  | 52, F          | Exophthalmos, convulsions, third nerve findings                | 4a              | Subtotal            | Parasellar, intraorbital    | Unchanged                        |
| Lunardi et al., 1989   | 60, F          | Headache, diplopia, hemiparesis                               | 3.5a            | Total               | Cisternal                  | Complete third nerve palsy       |
| Mehta et al., 1990     | 19, F          | Cerebellar signs, minimal ptosis, anisocoria                  | 5a              | Subtotal            | Parasellar, posterior fossa | Complete third nerve palsy       |
| Takano et al., 1990    | 65, F          | Diplopia, ptosis                                              | 2.5             | Partial             | Parasellar, middle fossa    | Complete third nerve palsy       |
| Barat et al., 1992     | 27, F          | Exophthalmos, visual impairment, cranial nerves IV, V, VI paresis | 4a              | Subtotal            | Intracavemosus ophthalmic canal | Complete third nerve palsy     |
| Niazi and Boggan, 1994 | 13, F          | Ptosis, anisocoria, hemiparesis                               | 3a              | Two operations: 1985 (subtotal), 1989 (complete) | Parasellar, cavernous, cisternal | Incomplete third nerve palsy     |
| Kachara et al., 1998   | 61, M          | Headaches, diplopia, minimal ptosis, trochlear nerve paresis   | 5               | Total               | Parasaellar, suprasellar    | Complete third and fourth nerve palsy |
| Netuka and Benes, 2003 | 12, F          | Headaches                                                    | 2.8             | Total               | Parasaellar, suprasellar, preptone | Complete third nerve palsy improved at 12 months |
| Prabhu et al., 2009    | 38, F          | Headaches, diplopia                                          | 3.5             | Total               | Parasaellar, suprasellar, preptone | Complete third nerve palsy     |

*Estimated based on findings from computed tomography or magnetic resonance imaging in the literature*

came across a finding similar to this in our patient, and third nerve could not be preserved and was sacrificed. Of the 4 patients who had an incomplete resection, 3 (75%) had a complete palsy of the third cranial nerve. Preserving function of the third nerve in these large tumors can be a challenge, as confirmed in this report, where 9 (75%) of the 12 patients operated (whether complete or partial resections) upon had complete third nerve palsy. In these large tumors, the distortion of the normal anatomy and intimate involvement of the nerve with the tumor make it difficult to preserve oculomotor nerve function after surgery. This would be an important message to be conveyed to patients who harbor these large tumors.

Oculomotor nerve reconstruction may be important for cosmetic reasons (to avoid ptosis) and in cases involving blindness in the other eye. Nerve grafting, tried after resection of smaller tumors, produces only partial recovery. None of the patients described in this report underwent any type of reconstructive procedure.

**CONCLUSION**

Large schwannomas of the third cranial nerve are a rare entity and are frequently misdiagnosed as meningiomas on preoperative imaging because of the paucity of clinical signs and symptoms of cranial neuromatoses. Although a significant number of these patients are left with complete third nerve palsy following surgical intervention, gross total resection of these tumors is recommended for treatment or prevention of brainstem compression. The patients need to be adequately counseled regarding the high morbidity from third nerve palsy associated with resection of these tumors.

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