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MAJOR REVIEW

Primary Optic Nerve Sheath Meningioma in Children

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Abstract. Primary optic nerve sheath meningioma represents a proliferation of meningothelial cap cells of the arachnoid villi within the optic nerve sheath. Patients younger than 20 years of age make up less than 5% of all cases of pediatric primary optic nerve meningiomas. Histopathologically, the most common subtypes in children are transitional (54%) and meningotheliomatous (38%). This tumor has been called aggressive in the pediatric population, with surgical excision recommended. However, the tumor may spread intraorbitally, intracranially, or intraocularly after subtotal surgical resection. Recent studies examined the use of fractionated, stereotactic radiation in children with this tumor; however, follow-up is limited. Neurofibromatosis type 2 is concomitantly diagnosed in 28% of patients with pediatric primary optic nerve sheath meningioma. There have been no known deaths attributed primarily to this tumor. (**Surv Ophthalmol 53**:543–558, 2008. © 2008 Elsevier Inc. All rights reserved.)

Key words. children • meningioma • optic nerve • orbit • pediatric • sheath • tumor

I. General Considerations

Tumors of the optic nerve are uncommon. They affect either the optic nerve sheath (typically, a meningioma) or the optic nerve itself (typically, a glioma). Optic nerve sheath meningioma is a proliferation of meningothelial cells within the nerve sheath of the orbital or intracanalicular portion of the optic nerve. This type of proliferation is found more frequently in adults than in children.[17]

Orbital meningioma may be either a primary tumor of the optic nerve arachnoid or a secondary tumor due to extension of a meningioma into the orbit from a primary intracranial meningioma. Only 10% of orbital meningiomas are primary to the optic nerve (92% intraorbital optic nerve; 8% intracanalicular optic nerve).[17] Intracranial meningiomas may arise from the cavernous sinus, falciform ligament, clinoid, sphenoid wing, pituitary fossa, planum sphenoidale, frontal-parietal area, or olfactory groove.[40,59] Rare forms of meningioma may develop apart from the optic nerve and intracranial extension from ectopic orbital arachnoid, other intraorbital nerve sheaths, or orbital mesenchymal cells.

Previously reported cases of primary pediatric nerve sheath meningioma (PPONSM) in persons younger than 20 years of age have addressed two clinical features: the relative rarity of this type of neoplasm and its aggressive behavior in this age group. A retrospective review of the Mayo Clinic patient database, searched using diagnostic codes for the period between 1960 and 2005, identified seven patients with PPONSM. Seven additional patients with PPONSM were identified by our colleagues at Universita' Degli Studi in Naples, Italy, and at Plastic Eye Surgery Associates in Houston, Texas. Cumulatively, the 14 cases we report, along

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with the 39 previously reported cases (total 53 patients), underscore both the uncommon nature of this condition and the loss of ocular function caused by tumor progression or treatment.

A. HISTORICAL PERSPECTIVES

PPONSM (see Table 1 for summary) was first studied in detail by Walsh,[61] who reported in 1970 on seven patients whose clinical course appeared to be more rapid and aggressive than that of older patients. Two patients in the series also had neurofibromatosis type 2 (NF-2).

In 1971, Lloyd[37] presented the case of a 10-year-old boy with proptosis and blindness in the affected eye. Plain film radiographs showed hyperostosis of the orbit and calcification of the optic nerve sheath. Pathologic studies confirmed an intracanalicular nerve sheath meningioma.

In 1974, Karp et al[32] studied 25 cases of ONSM accessioned at the Armed Forces Institute of Pathology between 1925 and 1968; 10 (40%) occurred in patients younger than 20 years of age. Data were compiled on the basis of histopathologic studies, predating the advanced radiographic studies available today. Seven years later, Alper[2] reviewed these 10 cases and 5 of his own, and found that 15 of 55 PPONSM patients (27%) were younger than 20 years of age. Both series advocated surgical resection for PPONSM because of its aggressive course in young persons.

In 1979, Cooling and Wright[13] described an 8-year-old girl who initially was diagnosed with an optic nerve meningioma after excision of the intracranial portion of the optic nerve. The diagnosis was changed to an optic nerve glioma after a second operation wherein the entire orbital portion of the optic nerve was removed and examined histologically. Arachnoid proliferation, a reactive response in the meninges, not meningioma, was found in the glioma. This finding indicates the difficulty in diagnosing the tumor even with tissue sampling. The authors questioned whether the large number of cases previously reported could be explained by misdiagnoses, since only 1 in 20 patients (5%) in their own study was younger than age 20.[13] Their findings demonstrated the difficulty in diagnosing PPONSM, particularly without advanced neuroimaging techniques, and the difficulty in identifying its accurate incidence.

In 1984, Sibony et al[56] reported one case of PPONSM in an 18-year-old female patient with bilateral optic nerve sheath meningiomas that severely affected visual acuity. This report was followed by one by Cibis et al[12] that documented the only case to date of intraocular extension of an optic nerve sheath meningioma in a child. They attributed the intraocular invasion to the aggressive nature of this tumor in children.

In 1989, Wright et al[63] summarized their experience with 50 cases of optic nerve sheath meningioma treated at Moorfields Hospital, London. Six of the 50 patients (12%) were 20 years old or younger, including a 10-year-old boy who underwent exenteration with craniotomy to remove the invasive tumor and who was alive and tumor free at 8-year follow-up.

In 1990, Kuroda et al[34] reported two cases of PPONSM. One patient was a 6-year-old boy who was previously misdiagnosed with an optic nerve glioma on the basis of the radiographic appearance of "kinking" in the optic nerve. Both patients underwent fine-needle biopsy for diagnosis and ultimately had subtotal transcranial excision. Of note, the second patient, an 11-year-old boy, underwent biopsy, orbitotomy, and transcranial resection without any visual symptoms preoperatively.

Two more cases of PPONSM were reported in 1991. A 4-year-old boy and an 11-year-old boy presented with decreased vision, proptosis, and extraocular motility deficits. Both patients underwent transfrontal craniotomies, and one underwent exenteration.

In the largest review of optic nerve sheath meningiomas in all ages, Dutton[17] surveyed all the medical literature up to 1991. In this large series of 256 optic nerve meningiomas, the mean age at presentation was 40.8 years (range, 2.5–78 years). Only 4% of these patients were younger than age 20.

In 2002, Pitz et al[47] reported the first case of PPONSM treated with stereotactic fractionated irradiation. The patient, a 13-year-old boy, was diagnosed 5 months before treatment with visual acuity at the counting fingers level in the affected eye. The tumor was confined to the orbit and vision improved to 20/200 at 37 months follow-up. In 2004, Baumert et al[4] treated a 9-year-old with stereotactic fractionated radiation therapy. Before these two cases, the standard of care was surgical resection by an anterior orbital approach or transfrontal craniotomy.

There has been no report to date on the use of observation alone in PPONSM.

B. DEMOGRAPHICS

1. Epidemiology

Demographic information about children with optic nerve sheath meningioma is difficult to obtain because of the rarity of this disorder. There is also controversy in the medical literature about potentially misdiagnosed cases.

The incidence of intracranial meningioma for all age groups is 2 per 100,000 per year,[49] or 18% of all

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