Pediatric Meningiomas- A Case Report and Review of the Literature

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Abstract

Background: Pediatric meningiomas are relatively rare accounting for only 3% of intracranial neoplasms. As such most of the literature on pediatric meningiomas exists in the form of isolated case reports or as a case series with small patient numbers. Despite this scarcity of literature several important features distinguish pediatric meningiomas from those that occur in adults. These include unique risk factors; a male predominance; larger tumor sizes at presentation; unusual sites of occurrence including intraventricular and parenchymal; higher World Health Organization histological grades; more aggressive clinical behavior and an increased tendency for recurrence. We present a 12-year-old male patient whom presented to our unit with a left fronto-parietal convexity meningioma of an alarming size.

Methods: A 12-year-old male patient presented to our unit with an eight-month history of a progressive headache complicated by a two-week history of vomiting. He also complained of progressive weakness of the right side of his body which had worsened to a point that he was no longer independently ambulant. General examination revealed craniomegaly with tortuous scalp veins. Neurological examination revealed a right upper motor neuron facial nerve palsy and a right hemiplegia of 2/5. Radiological investigation revealed an extremely large left fronto-parietal dural based space-occupying lesion which crossed the midline. Due to the adolescent’s progressive neurology he was taken to the operating room for emergency resection of the lesion. Gross total resection was achieved, and histopathological analysis confirmed the lesion to be a fibroblastic meningioma.

Result: Post-operatively the patient demonstrated a complete resolution of his hemiplegia and at three weeks post operatively he was already independently ambulant with power 4/5 on the previously hemiplegic side.

Conclusion: Despite the notorious features that characterize pediatric meningiomas, as well as an often-intimidating radiological appearance, adherence to standard Neurosurgical operating principles has the best chance of ensuring a successful outcome.

Keywords: Pediatric meningioma; Benign meningioma pediatrics; Unique features of pediatric meningiomas

Introduction

Meningiomas arise from arachnoid cap cells and in adults comprise 20-30% of primary intracranial neoplasms (Figure 1). In the pediatric population they are however relatively rare and comprise between 1 and 3% of primary intracranial neoplasms [1,2]. While having a female predominance in the adult population in the pediatric population several studies note these tumors to occur more commonly in males [3,4]. The male to female ratio in the pediatric population is described to be as high as 2-3:1 [5,6].

Unique risk factors exist for the development of meningiomas in the pediatric population and the strongest association is in patients with neurofibromatosis type II where a finding of multiple intracranial meningiomas is not uncommon. In fact, between 6-39% of pediatric meningiomas are reported to occur in children with neurofibromatosis type II [7]. Another study reported meningiomas to occur in 53% of neurofibromatosis type II patients and in the most severe phenotype called the Wishart variant meningiomas occur in up to 83% of patients [8]. The next strongest
association regarding pediatric meningiomas is in children whom have had previous ionizing radiation exposure [9,10]. Here the latency period is described to be on average 25 years in a study that evaluated the meningioma incidence in patients irradiated for childhood leukemia [11]. This specific subset of radiation induced meningiomas behave quite differently from sporadic meningiomas and are more often multifocal, more aggressive and exhibit a higher recurrence rate [12]. Besides the two established risk factors for pediatric meningiomas mentioned above several others are noted, although not specific for pediatric meningiomas but rather meningiomas in general, and include a family history of meningiomas, hormonal factors, obesity and cigarette smoking [13-16].

At a histological level the most common types of pediatric meningiomas are, like in adults, the meningothelial and fibroblastic subtypes [17]. When considering pediatric meningiomas overall several studies do however note a proportionately higher percentage of WHO grade II and III tumors (Figure 2). In pediatric meningiomas the mitotic rates as well as their proliferation indices are commonly higher, and they exhibit an increased rate of brain invasion as compared to the typical meningiomas that occur in adults [12,18]. They are also reported in several studies to exhibit aggressive behavior and to have an increased recurrence rate [19-21].

The unusual topographical distribution of pediatric meningiomas also sets them apart from their adult counterparts. Like adults the cerebral convexities have been reported to be the most common location for pediatric meningiomas in some series (Figure 3). Other studies note an increased occurrence of pediatric meningiomas in unusual locations such as intraventricular, parenchymal and in the cerebellopontine angle [22-25]. Other sites noted to predominate in the pediatric population are the orbit and optic nerve sheath and in contrast to adults' certain sites are extremely rare in children which include the suprasellar and parasellar regions, the cavernous sinus and around the foramen magnum [26].

Methods

A 12-year-old male patient presented to our unit with an eight-month history of a progressive headache complicated by a two-week history of blurring of vision and vomiting (Figure 4). He also
complained of progressive weakness of the right side of his body of 3 months duration which had recently worsened to a point that he was no longer independently ambulant. General examination revealed craniomegaly with an occipito-frontal circumference of 45cm and tortuous scalp veins. Neurological examination revealed the patient to be fully conscious and orientated to time person and place. Cranial nerve examination revealed a right upper motor neuron facial nerve palsy and motor examination revealed a right hemiplegia of 2/5 in all myotomes (Figure 5).

A computed tomographic brain scan was performed which revealed an isodense well-defined dural based left fronto-parietal space occupying lesion that on the coronal view demonstrated subfalcine herniation (Figure 6). The lesion measured 87x81x90mm and entrapment hydrocephalus of the right lateral ventricle was present. With the administration of contrast the lesion showed avid heterogeneous enhancement. An MRI showed the lesion to be well demarcated and hypo intense on T1 weighted imaging and hyper intense on T2 weighted imaging (Figure 7). With the administration of gadolinium, the lesion showed heterogeneous enhancement. MR spectroscopy was performed which noted a distinctive choline peak.

Due to the adolescent’s progressive neurology he was taken to the operating room for emergency micro-surgical resection of the lesion (Figure 8). A left fronto-parietal craniotomy was performed, and the lesion was noted to be dural based with no obvious overlying bony involvement. The tumor consistency was firm and after devascularising the tumor by excising its dural base demonstrated moderate bleeding while being internally debulked. The tumor rim separated easily from the surrounding brain tissue and was removed (Figure 9). A duroplasty was performed using a synthetic dural substitute and the bone replaced and reattached with mini-plates and screws. The scalp was closed in layers and the patient was transferred to the Intensive Care Unit (Figure 10).
Result

The child was extubated on day two post-operatively and a post-operative MRI confirmed that gross total resection had been achieved. Histopathological analysis confirmed the lesion to be a fibroblastic meningioma. Over the subsequent two weeks the child demonstrated a gradual resolution of his right hemiplegia. At three weeks post-operatively he was already independently ambulant with power 4/5 on the previously hemiplegic side. He was discharged into the care of his parents and booked for out-patient review in six months. No adjuvant radiotherapy was planned.

Discussion

Our patient had no family history of Neurofibromatosis nor previous exposure to ionizing radiation and hence this pediatric meningioma would be regarded as sporadic. The craniomegaly suggests a slow growing tumor which over time was partially accommodated by a disproportionately enlarging calvarium. The slow growing nature of this meningioma is also supported by the fact that despite its enormous size the child was fully conscious due to the brain having time to compensate for the slowly enlarging mass. This is further supported by the histopathological result of a fibroblastic meningioma which is a WHO grade I variant and considered benign (Figure 11).
Regarding prognosis a large meta-analysis recently published noted a 12% overall mortality for pediatric meningiomas of which 25% are secondary to intra-operative complications [26]. Another study noted an intra-operative complication rate of 46% in pediatric patients undergoing meningioma resection procedures, with the result that these patients often have long hospital stays [27]. Deaths related to post-operative adjuvant radiotherapy are also recognized and add to the mortality rate [26].

Due to our patient having experienced a successful gross total resection confirmed by post-operative MRI as well as the benign histopathological result, adjuvant radiotherapy and the potential complications of this were avoided. Re-operation rather than radiation would be considered as the first line therapy for recurrence in our particular patient.

In conclusion pediatric meningiomas are a specific neurosurgical consideration and should not be seen as on a par with the adult form of the disease. Besides the pediatric meningioma itself special surgical considerations related to the pediatric patient undergoing cranial surgery must also be considered, and planned for, to ensure a successful operative outcome [28]. These tumors require an interdisciplinary team for a successful outcome and as our case demonstrates this can be achieved.

**Conflict of Interests**

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**References**
