Anaplastic meningioma in an adolescent: Case Report and brief literature review

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ABSTRACT

Anaplastic meningioma is an uncommon neoplasm in childhood and adolescence. In contrast to the case in adults, pediatrics have a predominance of males over females. We report a case of a 17 years old with progressive scalp swelling on the right side. Patient underwent subtotal tumor resection at previous hospital three months ago. The operation was stopped due to extensive bleeding. Radiological investigations revealed a dura-based tumor with a cystic component. Following preoperative embolization, a craniotomy was done, and the tumor was almost completely removed. The tumor's histopathological analysis revealed characteristics of an anaplastic meningioma. The aggressive behavior of this tumor necessitates meticulous monitoring of therapeutic measures in such patients.

Keywords: anaplastic meningioma; craniotomy

INTRODUCTION

Childhood or adolescent meningiomas are a rare entity in this age group.1–3 There are various distinctive feature compared with adult cases, meningiomas in pediatric usually correlates with higher grade tumors (WHO grade II and III) in frequency, histological type that are more aggressive, higher recurrence rates, atypical sites predilection (spinal cord in particular), brain invasion frequency, and mitotic as well of proliferation indices. In contrast to the case in adults, pediatrics have a predominance of males over females.1,4

In this article we present the case of a 17 year old presenting to our institution with recurrent tumor at his right frontotemporal region diagnosed as anaplastic meningioma done at another institution. As a rare form of neoplasm in adolescent, this case is discussed along with a short literature review.

CASE REPORT

A 17-year-old boy presented to our neurosurgery clinic with progressive swelling on his right fronto-temporal...
scalp. The patient had a history of two previous craniotomies at another institution three months ago. (Figure 1) The pathology result was anaplastic meningioma. The bone flap was removed due to intraoperative swelling and bleeding. Based on information from the patient and his family, the operation was performed twice with an interval of 1 month, but the tumor could not be completely removed due to bleeding during surgery. The patient was referred for radiotherapy. 

Figure 1. Clinical photograph of the patient with a large swelling at his previous craniotomy site

The patient had a history of generalized tonic-clonic seizures four months ago, after which he was diagnosed and recommended for surgery. At the time of presentation, he was not symptomatic. He has had a large swelling on his right side at the craniotomy site and a new lump on his forehead since three weeks ago. A review of the systems revealed no evidence of recent changes during the previous two months, like headaches, weariness, or behavioral changes. He had no illnesses, and up until his presentation, he had been in generally good health. Upon physical examination, he displayed intact extraocular muscle movements, was alert and oriented, and had a bilateral and symmetric pupillary response. The rest of the cranial nerves remained unharmed. His motor, sensory, and cerebellar functions were examined, but the results were unremarkable and positive. Brain Magnetic Resonance Imaging (MRI) shows a mixed-intensity extra-axial mass with an inhomogeneous picture on contrast admission at the fronto-temporo-parietal area and a perifocal edema effect around it, as shown in Figure 2.

The patient was considered for reoperation as the lesion was too large and brain compression was prominent. Before the surgery, the patient underwent embolization for three days before undergoing reduced vascularization. A hypervascular lesion fed by the branches of the right external carotid was embolized using the super selective
cannulation co-axial technique into the feeding artery, followed by embolization using PVA 500–710 m until stasis was achieved. After embolization, vascularization to the lesion was significantly reduced, as shown in Figure 3.

![Figure 2](image1.png)

**Figure 2.** T1-weighted image with gadolinium contrast on Magnetic resonance Imaging (MRI) A. Axial Section B. Coronal Section C. Sagital Section

![Figure 3](image2.png)

**Figure 3.** Digital Subtraction Angiography (DSA) before and after embolization

Dexamethasone was administered postoperatively on the day of surgery to reduce cerebral edema. A myocutaneous flap was mirrored by a curvilinear pterional incision that continued through the earlier incision made short of the zygomatic arch. The adhesions from his previous operation were opened, and the dura was incised in cruciate fashion. The huge tumor mass was seen as multilobulated, gross, and firm, with immense infiltration into the cerebral cortex. The tumor was debulked using microsurgical techniques with coagulation and division of the direct feeding pial vasculature, which the tumor had parasitized from the surrounding brain. The cystic component had yellowish fluid, which was also sent to pathology but revealed no tumor. (Figure 4)
The tumor was firm, rubbery and also hypervascular during the procedure (A).

The patient is positioned before surgery for extended pterional craniotomy over the previous incision (B). Tumor excised and debulked sent for pathological examination (C).

Next, the deeper portion of the tumor was removed in a piecemeal fashion. This was of different characteristics, not as calcified but extremely hard, rubbery, and fibrous. The tumor was attached to the temporal bone. A near total excision was achieved; there was residual tumor adherent to the branches of the MCA. The dura was reconstructed using artificial duramater and fascia lata. The bone flap was not reconstructed at this time.

Postoperatively, the patient had no evidence of deficits in speech, personality changes, or motor or sensory deficits. Postoperative CT scan imaging showed no evidence of residual tumor, hemorrhage, or infarction. An evaluation MRI was performed on the patient 1
month after the surgery. There was a postoperative right temporal lobe defect with hemorrhagic and pneumoencephalic foci around the postoperative defect. The post-contrast, intensely heterogeneous lesion appeared in the right temporal lobe (peripheral postoperative defect), measuring 2.7 x 2.6 cm, with perifocal edema. The intensified lesion was also seen in the subcutis of the scalp in the right frontal region, with a diameter of 1–1.9 cm. A defect in the right temporal bone has developed after surgery. No midline deviation was seen, as shown in Figure 5.

Pathological examination results showed a hypercellular tumor with meningo-thelial differentiation. The tumor mass consists of round, oval, or spindle-shaped cells that grow in sheet-like clusters, partly arranged in the fasciculus. The nucleus cells were highly pleomorphic and hyperchromatic. Mitoses with 21 per 10 high-power fields (HPF) were found, along with areas of necrosis and bleeding. Eccentric nuclei in tumor cells with abundant cytoplasm and an eosinophilic appearance suggest gemistocytic or rhabdoid disease. Immunohistochemistry revealed that vimentin and Ki67 were positive in more than 20% of tumor cells, while EMA, GFAP, and AE1/3 were negative, as shown in Figure 6. Based on the histologic features and immunohistochemistry examinations, the diagnosis was an anaplastic meningioma (WHO grade III).

Figure 5. T1-weighted image with gadolinium contrast on Magnetic resonance Imaging (MRI) evaluation 1 month after surgery. A. Axial Section, B. Coronal Section, C. Sagital Section.
Figure 6. Histological features of the spindle-shaped cells tumor, pleomorphic, hyperchromatic, clear nuclei, eosinophilic cytoplasm, high mitotic along with necrosis and bleeding (Hematoxylin an eosin, original magnification A1. 10x, A2. 100x).

Immunohistochemistry results were positive on vimentin (B) and Ki67 (C) while AE1/3 (D), EMA (E), GFAP (F) were negative.

**DISCUSSION**

Meningiomas make up between 0.4 to 4.6 percent of all primary central nervous system (CNS) tumors in adults, compared to about 30% in children. Only 1.5–2.0 percent of meningiomas affect children. With a small male predominance in children, they are twice as prevalent in women as in men. Pediatric meningioma
incidence increases with age, and the average age at diagnosis is higher than that of other childhood CNS malignancies.³ When it occurs in youngsters, it is more likely to be associated with neurofibromatosis Type 2 (NF2) or past neoplasia radiation therapy. Meningioma was the first solid tumor in adults to be linked to the cytogenetic abnormality monosomy 22. Loss of NF2 gene expression was found to be an early tumorigenic event in subsequent studies. One of the most common genetic alterations is the loss of heterozygosity on chromosome 22q, which is associated with the bi-allelic inactivation of the NF2 tumor suppressor gene. Merlin is the protein product of NF2 gene expression and is thought to regulate cell growth, cell motility, binding of several transmembrane signaling proteins, and interaction with important proteins.³ Spontaneous meningiomas, on the other hand, are less common in children and can have uncommon imaging findings because they might originate in odd sites (spinal meningioma) and appear to be associated with a higher clinical grade (WHO grade II/III).²⁴⁻⁵ Genetic examination was not available in this case. The symptoms of meningioma in children are not unique to childhood. Intracranial hypertension symptoms (headache, vomiting, and optic nerve swelling) predominate. Epilepsy (32.9%), visual impairment (21.5%), motor impairment (17.1%), and cranial nerve dysfunction are among the other symptoms (14%).⁶ Our patient initial symptoms were seizures that occurred only one time before seeking medical attention. In contrast to adults (2–4%), imaging modalities (CT scan and MRI) demonstrate a high frequency of cystic transformation (15%) in childhood meningiomas which were also found in our patient. Another typical occurrence in this age group is a lack of dural attachment, which occurs in 28.5 percent of children, but it is uncommon in adults.⁵ The gold standard treatment for any meningioma is surgical resection, particularly gross total excision, which results in complete long-term remission.⁷ The risk of significant intraoperative hemorrhage and post-operative neurological morbidity may be limited by the size, vascularity, and location of the tumor in children. Pre-operative embolization may be beneficial in giant vascular tumor followed by multi-stage operation of the tumor.³ We achieved subtotal resection with a small residue that was adherent with MCA branches. Regardless of the malignancy grade of the meningioma, recurrence-free and overall
survival rates are higher after total resection. Subtotal resection is associated with lower recurrence-free survival and appears to be dependent on malignancy grade. Age has no bearing on recurrence-free survival. Children aged 3 to 12 years have a higher overall survival rate than children aged under 3 years or older 12 years. Any brain tumor diagnosed before the age of three years has a poor prognosis. This is likely due to the fact that children in this group are more likely to have congenital tumors with a more aggressive biological course. Hormones' oncogenic role is thought to be the primary cause of poor prognosis in children over the age of 12. Survival rates for patients with anaplastic meningioma have been shown to improve when surgical resection and adjuvant radiation therapy are combined, compared to surgery alone. A 3D conformal planning was used to deliver radiotherapy with a minimum dose of 50.4 Gy (median 55.80 Gy) for low-grade tumors and a higher dose for grade II and grade III tumors. Chemotherapy has been shown to be ineffective in the treatment of high grade meningiomas. Subtotal resection surgery was conducted in our patient after the embolization with scheduled adjuvant radiation therapy as the following post-operative management to reduce the chances of residual growth.

CONCLUSION

Anaplastic variations are extremely rare, and meningiomas are infrequent primary brain tumors in children. Although the clinical behavior of this tumor in young patients is unpredictable with distinct features when compared to adult cases, the treatment goal in adolescent meningioma is not that different from adult cases. Long-term monitoring is necessary due to the high likelihood of recurrence. Due to the rarity of these cases, guidelines for the treatment of meningiomas in children have not been established and need further research.

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REFERENCES


