Congenital Orbitocranial Meningioma

Presenting in a Neonate

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Abstract

Purpose: To report an unusual case of orbitocranial meningioma presenting in a neonate.

Methods: Case report

Results: A 1-day-old neonate was presented with severe proptosis of the left eye. CT scan and MRI showed an extensive mass with involvement of left orbit and brain. Discrete calcification could be seen on CT scan. The patient did not have any signs of neurofibromatosis and family history was negative. Histopathologic evaluation of orbital biopsy was compatible with meningioma.

Conclusion: This is an unusual presentation of meningioma in a neonate and to our knowledge this is the second report of such a case in the literature.

Keywords: Orbitocranial Meningioma, Orbital Tumor, Neurofibromatosis


Introduction

Neonatal brain tumors constitute 0.5-1.9% of all brain tumors in childhood.1 Intracranial meningiomas that occur within the first month of life are rare. Congenital tumors define as tumors that present or produce symptoms within the first month of life.2 To date only nine cases of such tumors have been previously reported in the literature.1,3-9 We report a rare case of menigioma in a 1-day-old neonate that presented with severe proptosis and involvement of orbit and temporal lobe-a unique report of a congenital menigioma invading orbit.

Case Report

A 1-day-old female newborn was referred to Oculoplastic and Orbital Service, Farabi Eye hospital, Tehran, Iran, in September 2006 with severe proptosis of the left eye from birth. She was a full term baby, 2300 grams weight. He had been the product of full-term, normal delivery without prenatal or intrapartum complications.

On physical examination severe proptosis of the left eye, chemosis, and conjunctival injection of the left eye was visible (Figure 1). Slit lamp examination showed only mild exposure keratopathy of the left cornea. Other globe examinations (anterior and posterior segments) of the both globe were unremarkable.
Systemic evaluation did not reveal any abnormal finding. The patient had no history of seizure, no neurologic deficit. Developmental exams were within normal range. The patient had not any sign of neurofibromatosis. He had not any remarkable familial history, and his mother did not take any suspected medication or trauma during pregnancy.

CT scan showed an extensive ill-defined inhomogeneous nearly high-density mass with involvement of left middle and anterior fossae with extension to the left orbit and caused severe proptosis (Figure 2).

Optic canal was markedly enlarged. Discrete amorphous calcification could be seen on CT scan within the tumor. This lesion could be seen on T1- and T2-weighted MRI as hyperintense mass (Figure 3).

The patient underwent incisional biopsy through lateral orbitotomy. Histopathologic evaluation of the biopsy specimen showed moderately cellular tissue, partially necrotic with calcification and composed of fibro-connective background, infiltrated by ill-demarcated spindle cells, forming whorl formations. The cells of the tumor had homogenous eosinophilic cytoplasm and round oval shaped vesicular nuclei, which altogether were compatible with meningioma with necrosis and calcification (Figure 4).
After 3 weeks the patient was presented with a large corneal ulcer impending to perforation. Smear and culture from the corneal ulcer revealed Staphylococcus aureus. The patient was treated with drop vancomycin and amikacin fort. However, after 3 days cornea was perforated and we encountered with an eviscerated eye so that evisceration was completed in operating room. 2 weeks later the patient was admitted in a pediatric hospital and craniotomy and radical excision of the tumor and exenteration was performed by a neurosurgeon. Postoperatively the patient had CSF leakage from orbit that controlled after 3 weeks. After recovery the patient was referred for radiotherapy, but her physician did not accept her radiotherapy due to fear of side effects. Fortunately, after 3 months the patient is well developed and she is going to undergo subperiosteal orbital implant to help orbital growth.

Figure 5. Postoperative axial orbital CT scan showing temporal and orbital areas that has been cleared from the tumor

Discussion

Infant brain tumors have some distinctive characteristics compared to those seen in older children. Most of these tumors are of neuroectodermal origin. Pattern of infantile intracranial meningioma is usually akin to that of adults and located supra-tentorially. In contrast, most brain tumors in children are infratentorial. Histological appearance of infantile meningioma is generally fibroblastic.

Congenital meningiomas are extremely rare. To our knowledge, 27 meningiomas in infancy, but only 9 congenital meningiomas, presenting within the 1st month of life, have been reported. Review of clinical and pathological finding of the 10 congenital meningiomas, including our case revealed that five cases were male and five others were female; 6 cases were “definitely congenital”, presenting with signs of increased intracranial pressure or with enlargement of the head or proptosis (our case) at birth; 3 cases were “probably congenital”, presenting with vomiting, exophthalmos, seizures within the 1st week of life; Only one case was “possibly congenital”, which was presented or produced symptoms within the 1st month of life. Tumors were located mainly in the supratentorial region. Dural attachment and cyst formation were noticed in six and three of the cases, respectively. Histopathologically, seven were fibroblastic, two were angioablastic, and one was angiomatous.

In our case, a supratentorial meningioma invaded the temporal lobe and orbit, and caused severe exophthalmos. There has been only one case of a congenital meningioma presenting as exophthalmos in the literature. Meningioma with extracranial extension is uncommon. Niida et al reported only 5 cases of 192 meningiomas over the 12 years that had been extended extracranially. Some meningiomas seem to extend through bone at places of decreased resistance such as sutures and foramina. In our case we did not find any extracranial extension of the tumor.

It is generally accepted that meningiomas in children tend to grow rapidly in spite of benign histological appearance. There is generally agreement that complete surgical resection is the therapeutic procedure of choice in cases with meningioma. If a radical resection cannot be performed, additional radiation therapy should be down because of the possibility of the rapid growth. Because cranial radiotherapy to young children has not been established due of possible damage to immature brain, some suggest reducing radiation dose, when the tumor is located in the deep region to avoid cerebral damage.

Conclusion

This is an unusual presentation of meningioma in a neonate and to our knowledge this is the second report of such a case in the literature.
References