CASE STUDY

GROSS TOTAL RESECTION IN A RARE CASE OF OPTIC NERVE ASTROCYTOMA: A CASE REPORT

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ABSTRACT

The authors present a 3-year-old female with increasing proptosis and absent vision in the right eye. Chemotherapy had done for 3months. But her ailments lingered. The right eye exhibited severe proptosis and poor vision, whereas the left eye was normal with 20/20 vision. Preoperative MRI revealed a dumbbell-shaped tumor in the intra-orbital and intra-cranial section of the right optic nerve. A lateral supra-orbital approach was used to dissect the dumbbell-shaped tumor and the right optic nerve. No remnant of the tumor was discovered during a follow-up examination.

The case study demonstrates how to identify and treat ONA surgically. However, we need further research on optic nerve PA to gain a better understanding of their behavior. While gross total resection (GTR) is usually curative, tumors in deep locations may be unresectable and require alternative therapeutic procedures. Additionally, the case study emphasizes the importance of additional research on early detection and prevention.

KEY WORDS: glioma, pilocytic astrocytoma, optic nerve glioma, extent of resection

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INTRODUCTION

Optic nerve astrocytoma is a rare and slow-growing glioma classified as pilocytic astrocytoma (PA). It is more prevalent in children and teenagers. PA make up 1.5–3.5% of orbital tumors and 66% of optic nerve tumors. Seventy-five percent of optic pathway pilocytic astrocytomas occur in children less than 12 years old. The most prevalent locations are in children's optic nerves and young adults' optic chiasms. Pilocytic astrocytoma and neurofibromatosis (NF) I have been connected. NFI individuals are more likely to develop pilocytic astrocytoma, a tumor of the optic nerve or chiasm. NFI patients with pilocytic astrocytoma make about 15%–21% of all cases. Astrocytoma without NFI on the optical pathway is very unusual [1]. Primary brain tumors in children (0–19) and in adolescents (0–14) account for 15.4% and 17.6%, respectively, of the total. According to other studies, it occurs at a rate of 4.8 per million people per year. PA, on the other hand, may strike anybody at any time and is becoming more uncommon over time. In the cerebellum (42%), supratentorial compartment (36%), optic pathway and hypothalamus (9%) and brainstem and spinal cord (9%), PA may develop anywhere in the CNS (2%) [2,3]. Tumors of this kind are slow-growing, well-circumscribed, and do not infiltrate nearby tissues or progress to more malignant categories unless they are treated aggressively[2]. This study presents a case of 3-year-old female patient with the diagnosis of PA of optic nerve.

CASE REPORT

After a 6-month history of developing proptosis and deteriorating vision, a 3-year-old child presented to our hospital-regional center for neurology and neurosurgery in Uzhhorod with absent vision in her right eye. Her first visit to the other institution was nine months before, when an MRI revealed a tumor the shape of a dumbbell in the intra-orbital portion of the right optic nerve that extended into the intracranial. At that time, decision was to perform chemotherapy after a MRI diagnosis as an optic glioma. She had ten rounds of chemotherapy (carboplatin, vincristine) for three months but no decrease in size of tumor. The right eye was found to be severely proptosis whereas the left eye was found to be normal, with 20/20 vision. She didn't seem to be doing anything else. Before admitting to our hospital, we again perform MRI of the brain, which showed the mass extends about 5mm from optic chiasma (Figure 1).

So we have recommended surgical intervention to remove the mass to prevent its extension into optic chaisma and the hypothalamus.

A lateral supra-orbital approach used to accomplish gross complete resection. During surgery, a Mayfield head holder was used after the patient was anesthetized supine. To stimulate frontal lobe retraction, the heads were stretched to a maximum of 20 degrees and rotated 25 degrees toward the contralateral side. From the supraciliary arch, 3-4cm of skin incision was done to

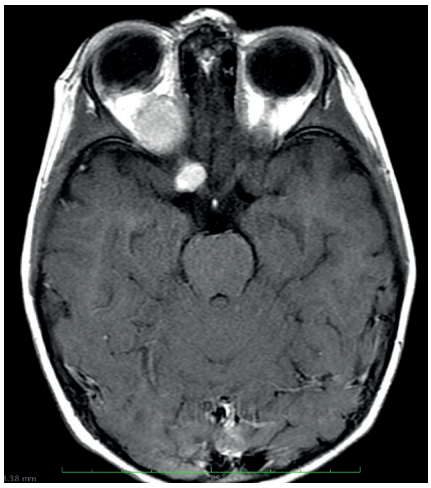

Fig. 1. Axial T1-Weighted Contrast of MRI of the brain shows dumbbelled shape of tumor extends intra-orbital and intra-cranial

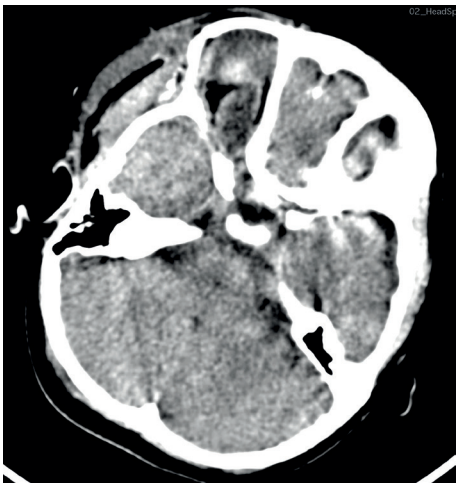

Fig. 2. Post-operation CT scan of Brain shows no hematoma

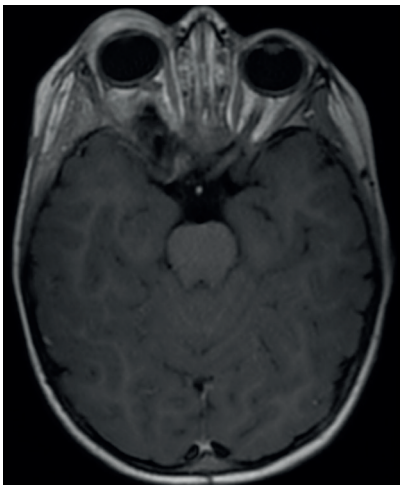

Fig. 3. First Follow up after 3month of T1-weighted MRI of the brain (axial) showed gross total resection of the tumor and no residue of tumor

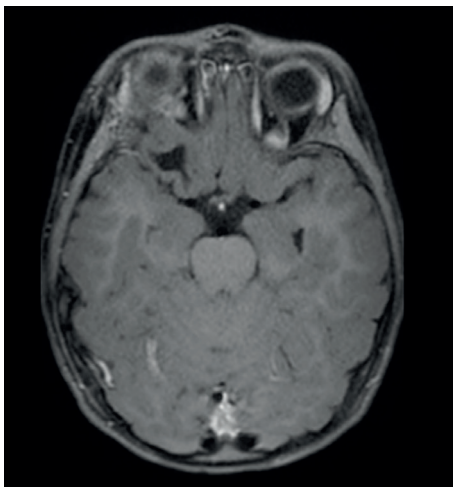

Fig. 4. One year follow up of T1-weighted contrast of MRI of the brain (axial) showed no sign of remnant of tumor

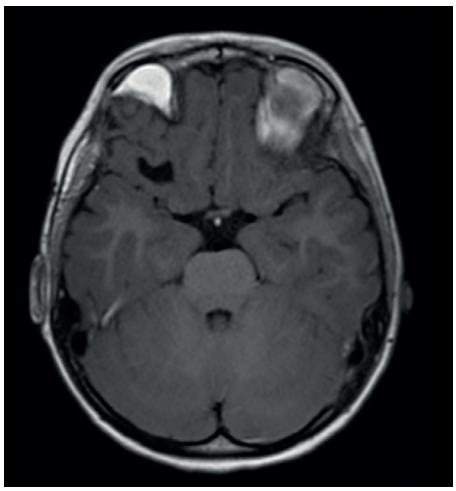

Fig. 5. The second-year followed up of T1-weighted contrast of MRI of the brain (axial)

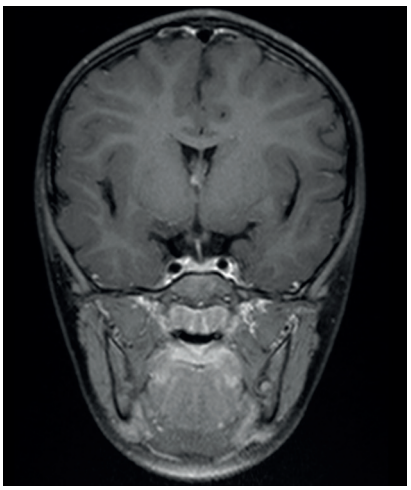

Fig. 6. The second-year followed up of T1-weighted contrast of MRI of the brain (coronal)

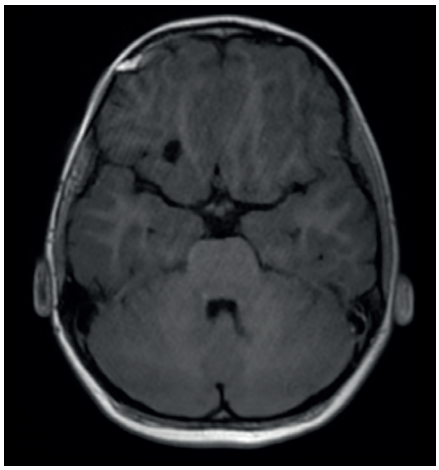

Fig. 7. Third-year followed up T1-weighted contrast of MRI of the brain (axial) shows no sign of remnant of tumor

the zygomaticofrontal suture. The orbicular muscle was pulled lower to disclose the anterior temporal line. A burr hole was drilled above the temporal line, posterior to the frontozygomatic suture. Drilling the inner border of the orbital rim near the optic nerve increased the surgical corridor and working angle. In the optic foramen of the lesser sphenoid wing. The anterior clinoid process was reached by lifting the dura immediately lateral to the optic canal. Following tumor exposure, the intracranial and intraorbital tumors were grossly resected, including the affected right optic nerve.Then step by step, the closure of skin was performed. Patient was discharged from hospital after a one week with the recommendation of a constant follow up with MRI. After 24 hours of surgery, the CT scan was performed (Figure 2) and it showed no evidence of any post-operative hematoma.

Histopathology revealed a pilocytic astrocytoma grade I. The tumor has a biphasic structure under the microscope,

with more compact sections composed of bipolar zones and highly eosinophilic Rosenthal fibers alternated with looser, spongier portions containing prominent microcysts on the microscopic level. Eosinophilic granular entities may be found in both the packed and loose zones. For the first time, the first follow-up was done after 3month, and MRI of the brain was done where no residue of tumor was seen (Figure 3).

Follow-up was made every year simultaneously to understand the status of patient, but after 3 years of follow-up, we found that there is no any sign of remnant of tumor. (Figure 4, Figure 5, Figure 6, Figure 7) and patient condition was very excellent.

Optic nerve astrocytomas are low-grade tumors that have a very diverse and surprising clinical history. They are most often seen in the optic nerve. Gliomas of the optic nerve are very uncommon and are only sometimes observed in routine clinical practice. The pilocytic astrocytoma (PA) is a benign brain tumor that often develops in infancy or early adolescence and is considered noninvasive.

These tumors usually impact the anterior optic system, causing symptoms including unilateral vision loss and proptosis. Optic nerve pilocytic astrocytomas are sporadic, like other PA. They are more common in persons with neurofibromatosis type 1. Pilocytic astrocytomas are generally indolent, regressing spontaneously after gross total resection or even without surgery.There are a variety of aggressive pilocytic astrocytomas that may extend to the brain and spinal cord, such as the pilo-myxoid type, which can return more often [3].

The tumor's topography seems to influence the disease's ocular progression. Acuity loss is more common in post-chiasmatic and optic pathway gliomas (62%) than intraorbital or pre-chiasmatic lesions (32 %). Proptosis is rare in posterior optic pathway gliomas. Absence of proptosis does not exclude saving intraorbital portions of the optic nerve. Early puberty affects 12-40% of children with chiasmatic optic pathway glioma.Lesions near the hypothalamus are thought to influence the hypothalamo-hypophyseal-gonadal axis, hence initiating puberty [4,5].

Radiologically, On CT scans, pilocytic astrocytomas often present as well-defined round/oval lesions that are iso- or slightly hypodense and significantly enhanced with contrast medium. On T1 sequences, PAs are often hypo- or iso-intense; on T2-weighted or FLAIR images, they are hyperintense [6, 7]. Vision testing, field of view measurement, ophthalmoscopic examination, and visual evoked potentials (VEP). The most reliable method is visual acuity [5].

It has the same histological and immunohistochemical characteristics as intracranial pilocytic astrocytoma. A biphasic shape, Rosenthal fibers, eosinophilic granule masses, and atypical mitotic patterns characterize both optic nerve and intracranial pilocytic astrocytomas.

Optic nerve pilocytic astrocytoma immunohistochemically positive for GFAP, OLIG2, and synaptophysin but negative for Neu-N [8,9].

Optic nerve pilocytic astrocytoma commonly appears as a cystic tumor with a mural nodule. Anatomically, a tumor is biphasic, with dense portions made up of bipolar zones and highly eosinophilic Rosenthal fibers and spongier regions with microcysts. Affected regions have eosinophilic granular bodies [2]. Consistent with the biology of pilocytic astrocytoma in all sites, the chance of recurrence is related to the surgical resection extent An great prognosis is achieved when PAs are entirely removed during surgical care [9-12].

Deep astrocytomas of the optic nerve (chiasma and brainstem) are treated with surgery, chemotherapy, and radiation. Chemotherapy should be given first in all patients with radiological or clinical progression.The treatment's side effects must constantly be handled. While multifractionated conventional radiotherapy is the most extensively used treatment, other methods of focusing radiation may be more beneficial [13, 14]. In symptomatic patients with optic pathway gliomas, imaging and follow-up visits are recommended [1].

In our case, a 3-year-old female with increasing proptosis and absent vision in the right eye was admitted. She had taken 3 months of chemo (carboplatin, vincristine) before us. But her ailments lingered. Preoperative MRI revealed a dumbbell-shaped tumor in the intra-orbital section of right optic nerve and intra-cranially extended before the optic chiasma. So, the decision was made to perform gross total resection through a lateral supra-orbital approach to dissect the dumbbell-shaped tumor and the right optic nerve to prevent the extension of the tumor to the optic chiasma so that both eye's vision would be compromised. No remnant of tumor was discovered during a follow-up on magnetic resonance imaging.

CONCLUSIONS

These uncommon pilocytic astrocytomas closely associate with neurofibromatosis type I. In our case, there was no indication of neurofibromatosis type I. Optic nerve astrocytomas grow slowly and may be managed conservatively. Optic nerve astrocytomas have a poor prognosis without therapy. Thus, surgery is only needed in cases of blindness, extreme pain, or severe proptosis. Nevertheless, all the patients should go for radiology for the evidence of extension posteriorly. When the chiasm is endangered, gross total or subtotal resection through craniotomy is essential to avoid hypothalamus or third ventricle involvement. Intracranial and intra-orbital optic nerve astrocytomas have a favorable prognosis with gross total resection and a slightly poorer after irradiation. A thorough workup, quick management, and imaging with frequent checks minimize vision loss in instances with optic nerve pilocytic astrocytoma.

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