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Case Report

Primary Optic Nerve Meningioma: A Unilateral Case Report in a Child

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Abstract: Primary optic nerve sheath meningioma is rare in children. It may be unilateral or bilateral. It is a sight threatening tumor due to it's location; one of the clinical features which draw attention is exophtalmos. We present a unilateral case with loss of vision in a female child.
Keywords: optic nerve, meningioma, exophtalmos

INTRODUCTION

Optic nerve sheath meningioma may be primary or secondary. Secondary optic nerve sheath meningioma arise intracranially from dura on or near the planum sphenoidale and spread anteriorly within the confines of the optic nerve sheath through the optic canal to surround the orbital portion of the nerve, whereas primary nerve sheath meningioma arise from arachnoid cap cells within the dural sheath surrounding the orbital or, less commonly, the canalicular portion of the optic nerve [1]. Primary optic nerve sheath meningiomas are the most common primary tumours of the optic nerve sheath [2] however they are extremely rare among children [3].Optic nerve sheath meningioma occurs more commonly in middleaged women. Primary Optic nerve sheath meningioma account for approximately one third of primary optic nerve tumors and 5% to 10% of orbital tumors [4].

Clinical manifestations include ipsilateral visual loss, color vision disturbance, visual field defect, proptosis, optic disc oedema and motility disturbance. Main lesions included in the differential diagnosis are optic glioma, orbital pseudotumor, lymphoma. Optic gliomas most commonly occur in children up to 10 years of age[5]. The patient presented here had proptosis and loss of vision.

CASE REPORT

A 2-year-old female child was brought to our office for left eye proptosis. According to her mother, the onset was gradual 2 years ago and the eye protruded progressively. No pathologic history was found, the pregnancy was normal as well as the delivery. Ophthalmic examination found in the left eye: a limitation of ocular motility in all directions, a relative afferent pupilary defect. The visual acuity was light perception.The fundoscopy was normal. Examination was othewise normal. We performed a Computed Tomography (CT) which showed a fusiform mass arround the optic nerve. Then we performed a biopsy under general anesthesia and sent it to the pathologist who concluded to an optic nerve sheath meningioma.

Fig-1: Anterior view of the child showing left eye proptosis

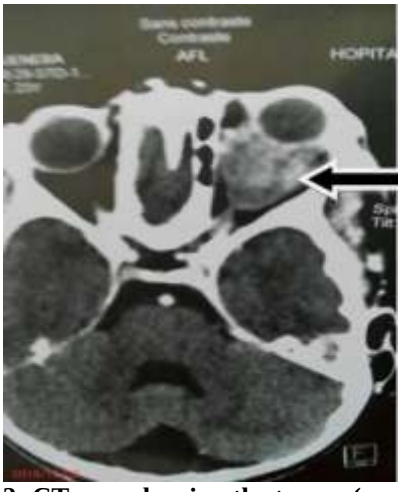

Fig-2: CT scan showing the tumor (arrow)

DISCUSSION

Optic nerve sheath meningioma is a rare benign tumors of the optic nerve. 60-70% of cases occur in middle age females, and is more common in older adults (mean age 44.7 years). It is also seen in children, but this is rare. It is typically a slow growing tumor and has never been reported to cause death. However, there is concern that the tumor can grow into the brain and cause other types of neurological damage [6]. Approximately 4% to 7% of Optic nerve sheath meningiomas occur in childhood . Optic nerve sheath meningiomas in children often behave in a more aggressive fashion characterised by faster growth, and more frequent intracranial and bilateral involvementthan occurs in adults [7]. Some Optic nerve sheath meningiomas remain localised to a small segment of the optic nerve, whereas others spread to surround the entire length of the orbital and canalicular portions of the nerve. Rarely, the tumour infiltrates the dura and adjacent orbital structures, including fat, extraocular muscles, and bone. When the tumour spreads to adjacent bone, it may enter the Haversian canal system, inciting hyperostosis and bone proliferation spreads beyond the confines of the nerve to infiltrate [2]. In our patient, the tumor had infiltrated the surrounding tissue of the optic nerve.

Primary and secondary orbital meningiomas present similar clinical presentation; exophthalmos and unilateral visual loss are the most common features described in the literature [8]. The diagnosis of optic nerve sheath meningioma relies heavily on imaging findings. Growth pattern can be either tubular, lobular, fusiform or focal. Tubular patterns marked by widening along the length of the nerve sheath are further subdivided into diffuse expansion, apical expansion towards the orbital apex, or anterior expansion towards the globe [9]. Treatment options vary; surgical options are limited and biopsies are becoming more uncommon in part because of associated morbidity. ‘‘En bloc resection’’ may be considered in cases of poor vision. Finally, radiation treatment is considered when there is evidence of progressive optic nerve compromise or definite tumor enlargement in non-diabetic patient [10].

CONCLUSION

Optic nerve sheath meningioma is not common in children. Because of its relationship with the optic nerve, it may end up with blindness. Early diagnosis and subsequent treatment are of the utmost importance for saving the sight and sometimes the life of the patients.

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