Case Report

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Proptosis Due to Isolated Intra-Orbital Meningocele- A Case Report

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ABSTRACT

Congenital orbital wall defects are very rare bone abnormalities that can cause protrusion of intracranial contents into the orbit. The protrusion of the meninges, which encompasses cerebrospinal fluid (CSF), result in a condition known as the orbital meningocele, a rare cause of pulsatile proptosis. Here, we present a 4-year-old female with left eye proptosis referred to our clinic. On magnetic resonance imaging (MRI), a cystic structure was present. The patient underwent surgery for cyst removal. CSF leakage was noted during surgery. Computed tomography (CT) scan revealed a bony defect in the sphenoid greater wing and the resulting meningocele. Craniotomy surgery was performed to close and fill the defect.

Introduction

Meningocele refers to herniation of the meninges through a bony defect in the skull or spinal canal which is accompanied by accumulation of the CSF. Orbital meningocele is a very rare abnormality in which the meninges herniate into the orbit through a defect in the orbit wall. In this condition, the meninges also enclose the CSF. The defect may be caused by trauma or congenital anomalies. The orbit wall defect may be in the roof, floor, medial or lateral wall of the orbit. Herniation can also occur through natural openings of the orbit [1]. Orbital meningoceles account for 1% to 1.5% of all meningoceles [2]. Here we presented a 4-year-old female with left eye proptosis due to isolated intra-orbital meningocele. We also described diagnostic and treatment modalities.

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Case Presentation

A 4-year-old female with left eye proptosis without any relevant medical history was referred to our clinic (Figure.1). She had no history of trauma, head injury, co-morbidity, previous illness, or congenital anomalies. According to her parents, the protrusion of the left eye gradually developed from early infancy. On many occasions, she complained of double vision on lateral gaze. It was not associated with any other symptoms such as headache, blurred vision, or trouble with eye movement. The patient's visual acuity was 20/20 in both eyes. Intraocular pressures were normal in both eyes, and anterior and posterior segment examination was unremarkable. The patient had proptosis with inward depression of the left eyeball, and Hertel exophthalmometry was 21 millimeters. There was no previous significant ocular history or family history. She had full range of eye movements in both eyes. Double vision was noted in lateral gaze of the left eye. Magnetic resonance imaging (MRI) of the brain and orbit was performed and revealed a cyst in the left orbit causing extrinsic pressure and subsequent proptosis of the eyeball (Figure 2). MRI did not elucidate any abnormal enhancement. There was no evidence of any intracranial mass. The brain and cerebellum appeared normal.

This patient, with diagnosis of a simple epithelial cyst, underwent transconjunctival anterior orbitotomy. During surgery, a large cerebrospinal fluid (CSF) leak into the cyst was noted. Fluid analysis was performed. A computed tomography (CT) scan was performed for diagnosis of any defect that might result in CSF leakage. The result of the fluid analysis was compatible with CSF (Table 1). The CT scan revealed that there was a defect in the sphenoid greater wing and orbit floor causing a protrusion of the meninges to the left orbit and subsequent proptosis of the eyeball (Figure 3). With this information, a diagnosis of isolated intra-orbital meningocele was made. A neurosurgical consult was obtained and left frontal craniotomy surgery was conducted. The greater wing of the sphenoid and dura defect was closed and filled with tissue replacement. At her six-month follow-up, her proptosis had resolved and the patient had no complaints.

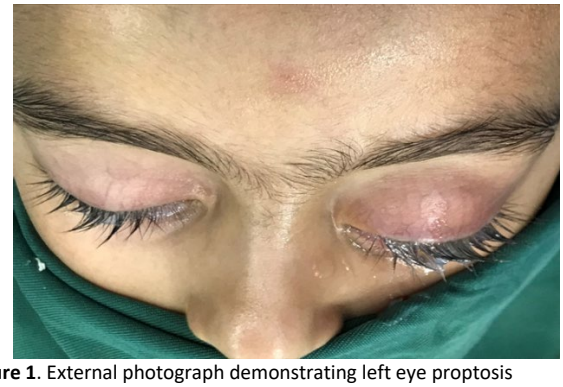

Figure 1. External photograph demonstrating left eye proptosis

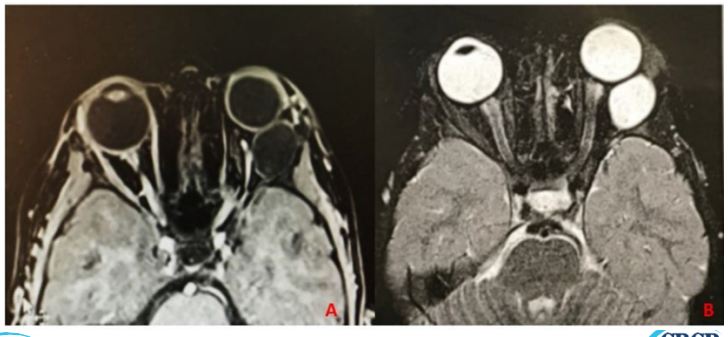

Figure 2. A: Postcontrast axial T1weighted fat-suppressed MRI of the orbits showing hypointense lesion with no contrast enhancement. B: Postcontrast axial T2weighted fat-suppressed MRI of the orbits showing hyperintense lesion.

Discussion

Differential diagnosis of proptosis in the 1-5 years age group includes dermoid, metastatic neuroblastoma, rhabdomyosarcoma, epithelial cyst, glioma of the optic nerve, sphenoid wing meningioma and, hemangioma [3]. Orbital meningocele is a very rare cause of proptosis. The rarity of this condition can be recognized from scarcity of literature on this topic. Most of the orbital meningocele cases are primarily due to congenital abnormalities but rarely etiology is trauma [4, 5]. This herniation is usually through a bony defect in wall of the orbit or very uncommonly through the natural structural openings in the orbit wall such as optic foramen or sphenoidal fissure. The protrusion site of the meningocele demonstrates some variations. In the anterior type, which is more common, the herniation occurs between the junction of the frontal and lacrimal bones, cribriform plate and nasal process of the maxilla. The posterior variety is much less common and usually occurs through the sphenoid fissure, optic foramen or posterior ethmoidal foramen. In majority of cases, the brain tissue may also be enclosed, in which case it is usually atrophic and edematous, and it is called orbital encephalocele; if the cerebral ventricle is included in the tumor, it is called hydro-encephalocele [6]. As a result, if the herniation of the meninge was suspected, herniation of the brain parenchyma must also be considered. Small defects are rarely symptomatic but may present with proptosis, recurrent meningitis, vertigo, seizures, and focal neurologic deficits. If symptomatic, clinical presentations begin to reveal at birth or early infancy and is rarely delayed until adolescence [1].

A correct diagnosis is a substantial step for management. CT scan is usually sufficient to establish the diagnosis and planning of surgery [7-9]. Bony defect opposite the cyst may be present in a CT scan. A significant challenge in making an accurate diagnosis of intra-orbital meningocele is to identify the bony defect in the orbit wall, which may be very small and not apparent on CT scan or MRI or may vary from case to case [1]. CT scan is of substantial importance for the diagnosis of bone defects rather than for characterization of the lesion itself, in which MRI is more useful. Meningocele demonstrated characteristics of CSF in MRI: T1 hypointense and T2 hyperintense. On the other hand, Encephalocele could appear as a protrusion of the brain parenchyma through a bony defect into the orbit. Encephalocele enhances the same as normal brain parenchyma, whereas meningocele will demonstrate no enhancement [1]. If clinical suspicion for meningocele is high and no bone defect is evident, analysis of the cystic fluid, which is in concordance with CSF fluid, will confirm the diagnosis [10]. A meningocele can easily be misdiagnosed as a simple epithelial cyst confined to the orbit. However, some of these cases never develop symptoms and live without the condition being diagnosed [10].

Treatment of congenital meningoceles consists of excision of the cyst and closure of the bone defect [4, 11]. The bone defect is closed and filled with granulation tissue and in large bony defects either bone graft, gel film or synthetic material like titanium are used. With early surgical intervention, majority of cases go through life without any further problems [9]. A meningocele protruding through the orbit may be misdiagnosed due to rarity and lack of awareness of this entity. CT scan is usually sufficient to establish the diagnosis and planning of surgery. Treatment of congenital meningoceles consists of excision of the cyst and closure of the bone defect.

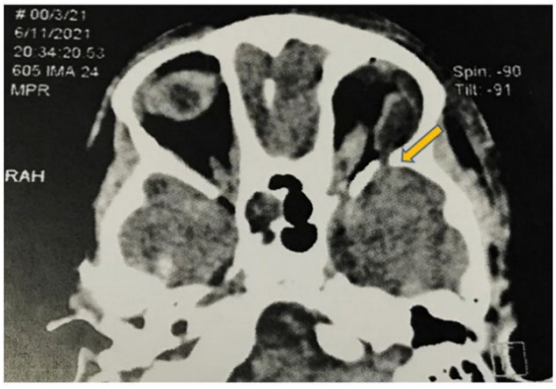

Figure 3: Axial computed tomography image showing a defect in sphenoid greater wing (arrow)

Table 1: The results of cerebrospinal fluid analysis

	VALUE
APPEARANCE	Clear, colorless
PROTEIN (MG/DL)	31
GLUCOSE (MMOL/L)	59
GLUCOSE CSF: SERUM RATIO	0.6
WHITE CELL COUNT	< 5 Cell Count
GRAM STAIN	Negative

Ethical Considerations

Compliance with ethical guidelines

All activities elucidated in the current investigation were carried out in adherence to ethical principles. The patient provided informed consent for the disclosure of their anonymous data in this manuscript.

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Conflict of interest

The authors declared no conflict of interest.

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