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Grade III Ptosis Revealing Congenital Orbital Encephalocele

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

This article aims to provide an overview of the clinical, diagnostic, therapeutic and prognostic aspects of orbital encephalocele. A patient with age of 48, operated on for thyroid goitre and treated for acute lymphocytic leukemia for 5 years, presented for one month with rapidly worsening ptosis of the right eye, with limited elevation and no other ophthalmoplegia or associated signs, A cranio-orbital MRI was ordered, revealing an intraorbital encephalocele exerting a mass effect on the upper eyelid levator muscle and the homolateral upper rectus. Orbital encephalocele usually occurs due to a failure in neural tube closure during embryonic development. This malformation manifests itself as a herniation of cerebral structures through a bony opening in the base of the skull. Encephaloceles can vary in type, with different contents, including the cerebral cortex or meningeal structures. Long- term follow-up is crucial to assess neurological development and quality of life. Ongoing research in this field is essential to improve clinical outcomes and develop more effective treatment strategies.

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Keywords: Encephalocele; neurological development; eyelid levator muscle; goitre.

1. INTRODUCTION

Orbital encephalocele is a rare congenital malformation characterized by the protrusion of brain tissue and meningeal membranes through an abnormal opening in the skull, typically located near the orbit. The encephalocele may arise through natural foramina such as the superior orbital fissure or the optic foramen (i.e. posterior orbital encephalocele) or bony defects in the orbital roof (i.e. anterior orbital encephalocele) [1,2]. This condition can lead to a variety of clinical complications, including visual disturbances and facial abnormalities [3,4]. The brain lacks part or the entire cerebrum which is an area of the brain responsible for cognitive functions like thinking, vision, hearing, touch, and some movement. There is no bony structure covering over the back of the head and there may also be missing bones around the frontal and lateral parts the head [5,6]. This article aims to provide an overview of the clinical, diagnostic, therapeutic and prognostic aspects of orbital encephalocele.

2. CASE PRESENTATION

A patient aged 48, operated on for thyroid goitre and treated for acute lymphocytic leukemia for 5 years, presented for one month with rapidly worsening ptosis of the right eye, with limited elevation and no other ophthalmoplegia or associated signs, A cranio-orbital MRI was ordered, revealing an intraorbital encephalocele exerting a mass effect on the upper eyelid levator muscle and the homolateral upper rectus. The patient was referred to the neurosurgeon for tumour exseresis.

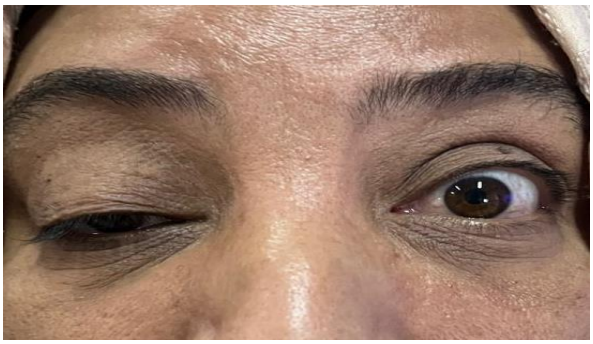

Fig. 1. Grade III ptosis of the right eye for 1 month

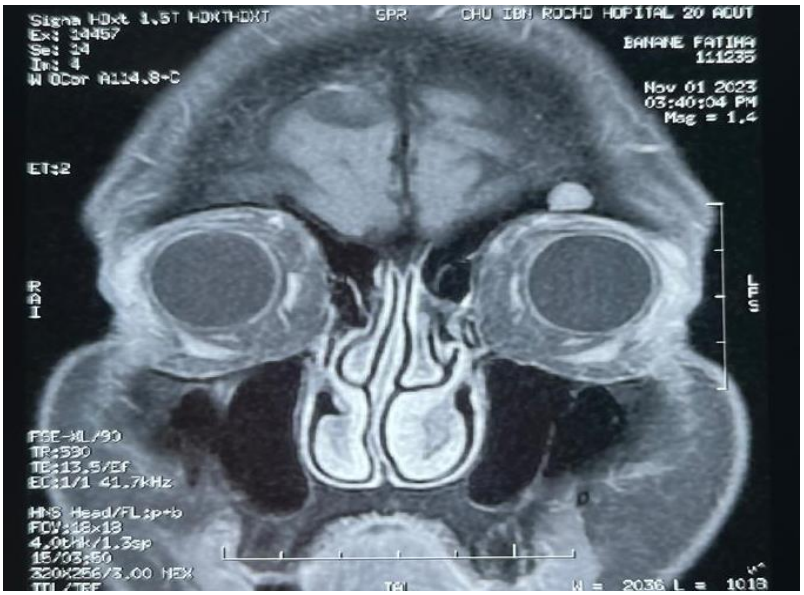

Fig. 2. An intraorbital encephalocele exerting a mass effect on the upper eyelid levator muscle and the homolateral superior rectus

2.1 Epidemiology

Orbital encephalocele is a rare malformation, with an estimated prevalence of around 1/5,000 live births. Although its incidence is relatively low, it is often associated with other congenital anomalies, such as neural tube defects. Risk factors include family history, chromosomal abnormalities and environmental exposures during pregnancy.

2.2 Anatomy and Pathophysiology

Orbital encephalocele usually occurs due to a failure in neural tube closure during embryonic development. This malformation manifests itself as a herniation of cerebral structures through a bony opening in the base of the skull. Encephaloceles can vary in type, with different contents, including the cerebral cortex or meningeal structures.

2.3 Clinical Manifestations

Clinical signs may include:

- Visible protrusion of a mass near the orbit.
- Facial abnormalities (asymmetry, deformity).
- Visual disturbances (strabismus, amblyopia).
- Neurodevelopmental delay in more severe cases.

2.4 Diagnosis

Diagnosis of orbital encephalocele relies primarily on imaging techniques, such as:

- Magnetic resonance imaging (MRI): to assess brain structures and determine the extent of herniation.
- Computed tomography (CT): useful for visualizing bony structures and skull anatomy. A full neurological assessment is also essential for accurate diagnosis and treatment planning.

2.5 Treatment

Treatment of orbital encephalocele is surgical, aimed at correcting the cranial opening and reintegrating brain tissue into the skull. The main stages of the procedure include:

- Resection of the herniated mass.
- Reconstruction of the skull base to avoid future complications, such as infection or cerebrospinal fluid leakage.

Post-operative management includes neurological follow-up, monitoring of complications and rehabilitation, if necessary.

3. DISCUSSION

The results of clinical studies show that prognosis depends on several factors, including the size of the encephalocele, age at surgery and the existence of associated malformations. Patients who undergo early surgery generally have better functional outcomes. Occipital encephalocele are the most common form of this congenital disorder and are manifested as a swelling of different sizes over the occipital bone in the midline. Proper diagnosis and treatment is highly important in the management of this congenital malformation of brain [7,8,9].

4. CONCLUSION

Orbital encephalocele is a complex congenital malformation requiring a multidisciplinary approach to diagnosis and treatment. Long-term follow-up is crucial to assess neurological development and quality of life. Ongoing research in this field is essential to improve clinical outcomes and develop more effective treatment strategies.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of this manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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