

## Case Report

# Intraorbital Encephalocele Presenting with Exophthalmos and Orbital Dystopia : CT and MRI Findings

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A 15-year-old female patient with progressive pulsatile exophthalmos caused by intraorbital encephalocele was evaluated with computed tomography (CT) and magnetic resonance imaging (MRI) in our clinic. She had no history of trauma or reconstructive surgery. When she was a little girl, she had undergone surgery for congenital glaucoma on the right eye. On the three-dimensional image of CT, a hypoplastic bone defect was observed in the greater wing of the right sphenoid bone. MRI and CT scan showed herniation through this defect of the arachnoid membrane and protruded cerebral tissue into the right orbita. Intraorbital encephalocele is an important entity that can cause pulsatile exophthalmos and blindness.

**Key Words :** Intraorbital encephalocele · 3-dimensional CT · MRI.

## INTRODUCTION

Different to a meningocele, the bulging portion of which contains only cerebrospinal fluid and overlaying dura, an encephalocele is a cystic malformation of the brain tissue herniating through bone<sup>6</sup>. Encephaloceles occur at a rate of one per 4000 live births worldwide<sup>1</sup>. Encephaloceles are classified according to their localizations in the cranium. They are often located in the occipital region; basal encephaloceles are relatively rare, constituting only 1.5% of all cases<sup>7</sup>. Encephaloceles are classified according to 5 anatomic sites, namely the speno-ethmoidal, trans-sphenoidal, speno-orbital, transethmoidal and spenomaxillary<sup>6</sup>. Congenital absence of portions of the wings of the sphenoid leads to speno-orbital encephalocele, a rare entity in the literature<sup>2</sup>. We wanted to share this case who had not seen any treatment until puberty and being purely congenital as being not related to trauma or neurocutaneous syndrome and also in the literature review there isn't any case like our case.

## CASE REPORT

Our case was a 15-year-old female patient, who had progressive pulsatile exophthalmus and dystopia in the right eye, beginning

at early age (Fig. 1). The orbital pathology was evaluated with CT scanning and MRI. At age 4, she had undergone trabeculectomy on the right eye for congenital glaucoma. She had no history of chronic disease or trauma. No congenital or acquired patch or rash was detected on her skin. There was no genetic disease in her family history. On the three-dimensional (3D) image of CT, a



**Fig. 1.** Skin reformatted three-dimensional CT scan of the patient showing exophthalmos and dystopia in the right eye and increased distance between the eyeballs due to dystopia.

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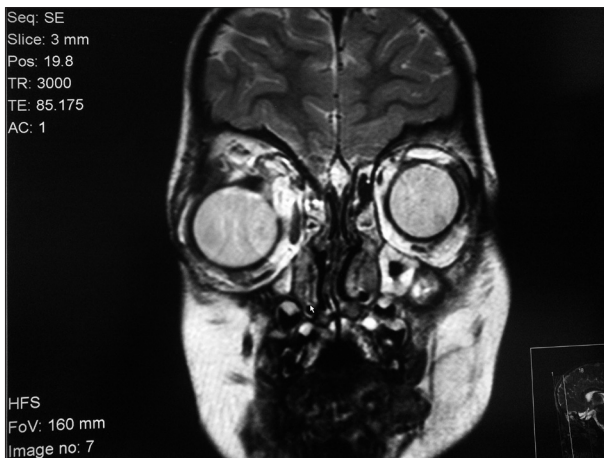
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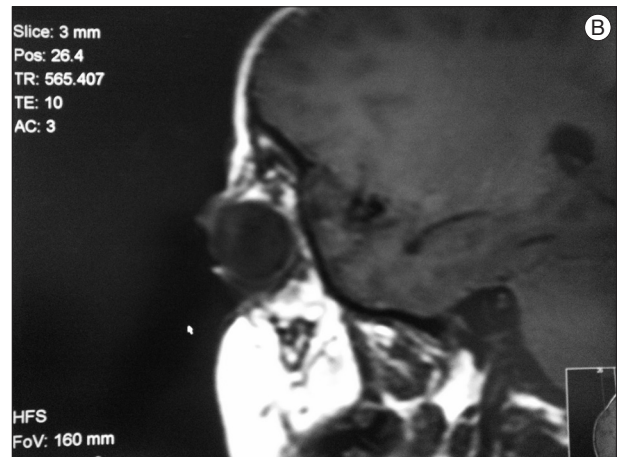
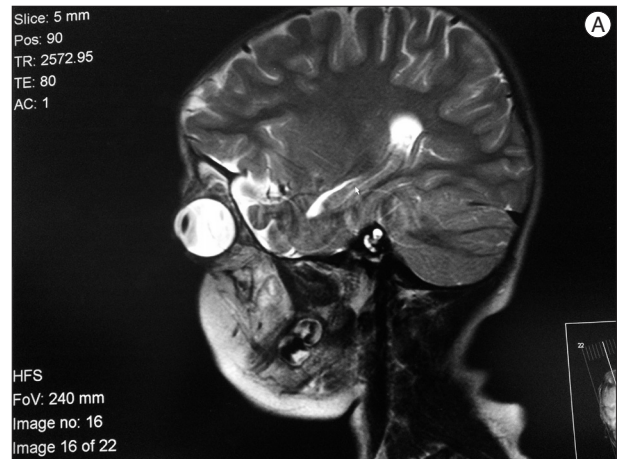


**Fig. 2.** Bone-reformatted three-dimensional CT scan of the patient showing enlargement of the right orbital cavity and large bone defect in the superior orbital fissure secondary to hypoplasia. The bone defect allows the petrous part of the temporal bone to be viewed in the posterior plain (arrows). The normal superior orbital fissure in the left orbita is shown with arrows.



**Fig. 3.** T2-weighted coronal MRI showing the inferio-lateral displacement of the right bulbus oculi. The right bulbus oculi is markedly larger than the left bulbus oculi.

large hypoplastic bone defect of 20.6×30.7 mm (35.8 cm<sup>2</sup>) was observed in the superior orbital fissure of the greater wing of the right sphenoid bone (Fig. 2). On MRI, the anterior-posterior diameter of the right bulbus oculi, which measured 27.9 mm, was relatively larger than the anterior-posterior diameter of the left bulbus oculi, which measured 22 mm; the distance between the two medial canthi, which measured 33.1 mm, was increased, and the right bulbus oculi was seen to have been displaced to the inferior by 5.9 mm (Fig. 3). The right orbita was markedly exophthalmic. The T1- and T2-weighted images obtained on MRI showed the left temporal lobe, with the subarachnoid membrane extending into the orbita through the defect (Fig. 4). In studies investigating the possible presence of accompanying neurofibromatosis, no optic glioma was found. Lish nodules were not observed on ophthalmological examination. Following completion of all the examinations, surgery was recommended to the



**Fig. 4.** A : The T1-weighted sagittal MRI without contrast demonstrating a large bone defect and cerebral tissue within the defect protruding into the orbita. B : The T2-weighted sagittal MRI demonstrating no apparent dysplasia in cerebral tissue, but an increase in the levels of peripheral cerebrospinal fluid and marked exophthalmos and displacement toward the anterior of the right bulbus oculi.

patient. Her parents were informed about the planned surgical intervention, but they didn't accept risks of the surgery, so surgery did not performed and the patient was followed up.

## DISCUSSION

Encephaloceles are seen in the rate of one in every 4000 live births and defined as the protrusion of the intracranial structures<sup>1,3</sup>. Most of them occur sporadically and do not accompany to hereditary syndromes<sup>6</sup>. Encephaloceles are classified according to their intracranial localizations and basal encephaloceles are classified under 5 main headings according to their localizations including spheno-ethmoidal, trans-sphenoidal, spheno-orbital, transethmoidal and spheno-maxillary<sup>6,7</sup>. Absence of the sphenoidal wings leads to spheno-orbital encephaloceles and brain tissue and meninges protrude to orbital cavity through superior orbital fissure<sup>2,6</sup>. Increased herniation due to the dural defects and the absence of the limited effect of the bone during physiologic growing of the brain and the cranium, continuous pulsation

of the brain are the accepted mechanisms of the enlargement of the bone defect<sup>7</sup>. Impairment of dura is important for encephalocoeles and cerebrospinal fluid (CSF) pulsation erodes the bone gradually<sup>7</sup>. The pulsation of the brain in the intraorbital field leads to the pulsation of the globe through the defect<sup>4</sup>. Although encephalocoeles are one of the important causes of pulsatile exophthalmus accompanying to dystopia, they are rare in this localization and usually accompany with neurofibromatosis type 1<sup>8</sup>. It is quite rare without trauma and NF 1, as in our case.

Definite diagnosis of encephalocoele is made through imaging techniques<sup>5</sup>. 3D CT imaging is the first step recommended for detection of the orbital bone defect<sup>7</sup>. The greater wing of the left sphenoidal bone was hypoplastic and the left superior orbital fissure was enlarged on 3D CT examination in our case, consistently with the congenital orbital encephalocoeles which does not accompany with a few syndromes in literature, although it is quite frequent. As in our case, there was the right sphenoidal wing dysplasia on 3D CT in a 22-year-old patient described by Sharma et al.<sup>8</sup> and the patient had progressive exophthalmus since 7 years of age and the right temporal lobe was herniated to orbita<sup>1,3</sup>. MRI provides definite data about the localization of the bone defect, the content of the encephalocoele and surrounding tissues (5). Continuity of the herniated tissue with brain is shown with MRI examination. In our case, the encephalocoele sac contained CSF and brain tissue of the left temporal lobe on MRI. The herniated brain tissue may vary depending on the encephalocoele localization. While the orbital encephalocoele defined by Sharma et al.<sup>8</sup> contained the brain tissue of the left temporal lobe, the orbital encephalocoele defined by Morihara et al.<sup>7</sup> contained the encephalomalasic brain tissue of the right frontal lobe. The 25-year-old male patient had orbital roof defect due to a motorbike accident experienced when 10-year-old<sup>7</sup>. MRI examination is superior for detection of any accompanying anomalies and the association of the encephalocoele with the vascular structures and determination of the major vascularization supplying blood to this field<sup>1</sup>.

The definite treatment method is surgical intervention after determination of encephalocoeles and assessment of their structure<sup>5</sup>. The whole encephalocoele sac should be repaired in a multidisciplinary way. The whole gliotic structures in the orbital cavity should be removed and the dura defect should be closed watertight and reconstructive cranioplasty should be performed. Temporal muscle fascia and watertight repair is sufficient for preventing CSF pulsation if the orbital defect area is below 2.5×2.5 cm<sup>2</sup>. Autogenous bones and artificial materials should be used for prevention of CSF pulsation if the defect area is above 3×3 cm<sup>2</sup>. Retraction of foramen rotundum and foramen ovale and crani-

al nerve, cavernous sinus, sagittal sinus injuries, intraorbital traumas and hemorrhages should be avoided during surgery. The artificial materials used during cranioplasty should be fixed so as not to allow dead space. Tight bandage and dressing should be applied in postoperative period and subcutaneous hydrops should be prevented. The most frequent complication is CSF leakage<sup>4,7</sup>.

Most authors recommend surgery within the first weeks. Early surgery enables better fixation of sutures and complete closure of dura. CSF leakage-related complications are seen more frequently in delayed cases<sup>8</sup>.

For sphenoidal-orbital encephalocoeles, the following can be concluded : The important clinical findings are : pulsating exophthalmos induced by pulsating brain and orbital dystopia; 3D CT scanning and MRI are important modalities in the preoperative diagnosis, and the only effective treatment is surgery.

## CONCLUSION

For sphenoidal-orbital encephalocoeles, the following can be concluded : the important clinical findings are : pulsating exophthalmos induced by pulsating brain and orbital dystopia; 3D CT scanning and MRI are important modalities in the preoperative diagnosis, and the only effective treatment is surgery.

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