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CASE REPORT

# A torcular encephalocele with proatlas defect and os-terminale

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## ABSTRACT

Encephalocele means if meninges and brain tissue protrude out of the cranium. There are different types of encephalocele. The occipital encephaloceles are the most common type. Craniocervical junction and upper cervical spine abnormalities can rarely be associated with occipital encephalocele. We discuss this case because there is rare association between torcular encephalocele and proatlas anomalies.

**Key words:** Cranio-cervical-junction anomaly, os terminale, proatlas anomaly, torcular encephalocele

## Introduction

Occipital encephalocoeles are more common than anterior encephalocoeles. In the Western hemisphere, occipital encephalocoeles constitute 80 to 90% of all encephalocoeles.[1-3] Abnormalities of craniocervical junction include: Basilar impression, atlanto-occipital dislocation, atlantoaxial dislocation, and occipitalization of the atlas or thin or deficient posterior arch of atlas.[4]

Os-odontoideum is a separate bony ossicle of variable size, with smooth cortical borders, separated from a foreshortened odontoid peg, occasionally may fuse with the clivus.

Os-odontoideum mimics type 1 or 2 odontoid fracture. Etiology is debated. Diagnosis and treatment do not depend on which etiologic theory is correct. This is a developmental anomaly, nonunion of dens to body of axis. However, it does not follow known ossification centers, and has been demonstrated in nine patients with previously normal odontoid processes. Acquired ones are postulated to represent an old nonunion fracture or injury to vascular supply of developing odontoid.[5,6]

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In ossiculum terminale, there is failure of fusion of odontoid process. We report a case of torcular encephalocele with OS-terminale and split atlas.

## Case Report

An 18-year-female patient, admitted in our unit, with complaints of a swelling at the back of head since birth. The swelling was increasing in size slowly, almost rounded in shape 8 cm in diameter and 21 cm in circumference. Consistency of swelling was soft, non pulsatile. Over laying skin was normal. Headache was present at the back of head for the last two years. Patient was born at a hospital by normal vaginal delivery; her mother died just following her birth, and perinatal period was uneventful. No history of unconsciousness, visual disturbance, seizure, limb weakness, dysphasia, nasal regurgitation, bladder and bowel dysfunctions. Her developmental mile stones were normal. Pupil was bilaterally normal, equal in size and reacting to light. The visual acuity and field of vision and fundi were normal. Other cranial nerves were normal. There was no sensory and motor deficit. Clinical and radiological diagnosis was Torcular encephalocele with pro-atlas defect and os-terminale. Magnetic resonance imaging of brain showed encephalocele and brain tissue herniated into the sac [Figure 1].

### Surgical procedure

Under general anesthesia with prone position excision and repair of sac was done [Figures 2 and 3].

### Comment during discharge

Patient prognosis appeared good in view of minimal gliosed parenchyma present in sac. Patient should continue to wear hard cervical collar. Patient will need posterior fixation for C1 abnormalities. Hence, she must be on regular follow-up in Neurosurgery Out Patient Department.

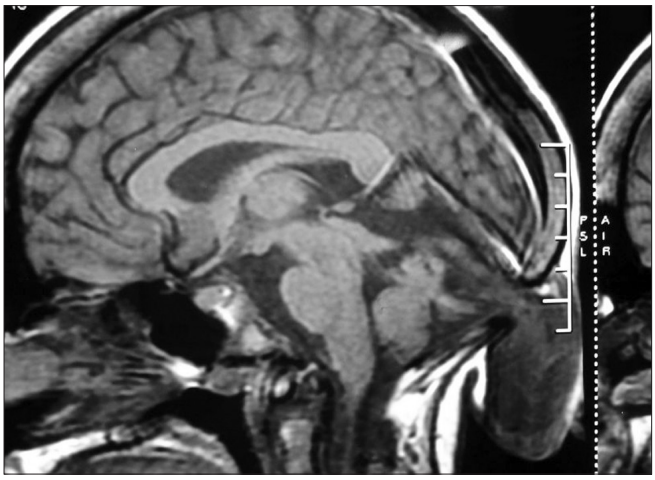

**Figure 1:** Sagittal MRI shows torcular encephalocele containing brain tissue

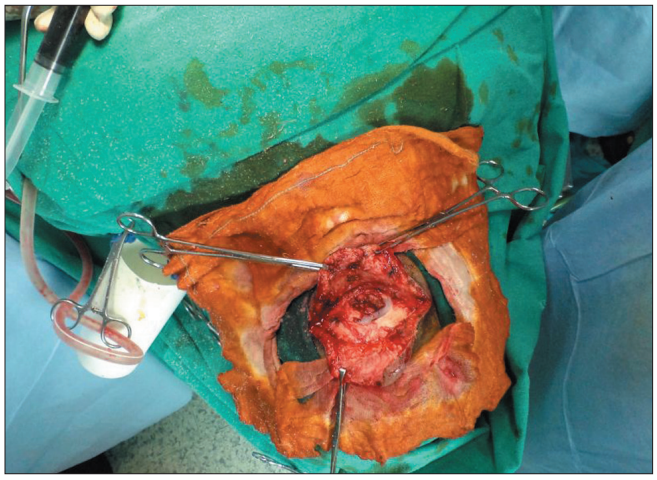

**Figure 2:** Intraoperative photo shows encephalocele sac

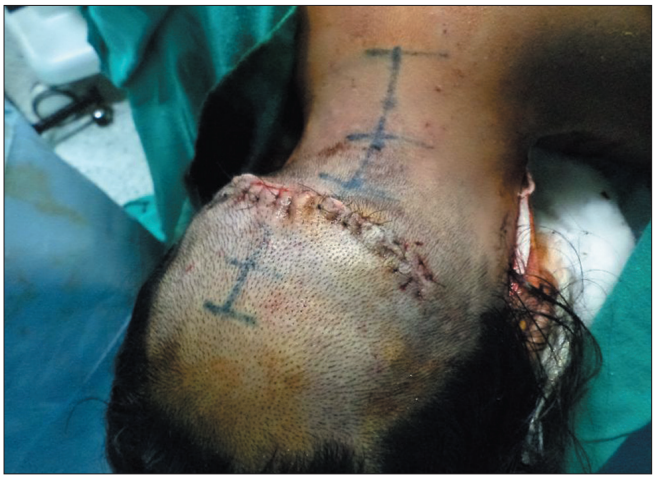

**Figure 3:** Intraoperative photo at the end of surgery showing sutured wound

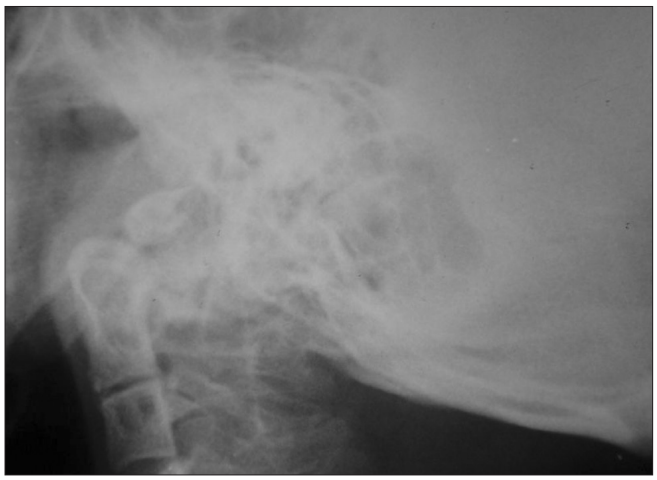

**Figure 4:** Lateral X'Ray CVJ showing OS Terminale

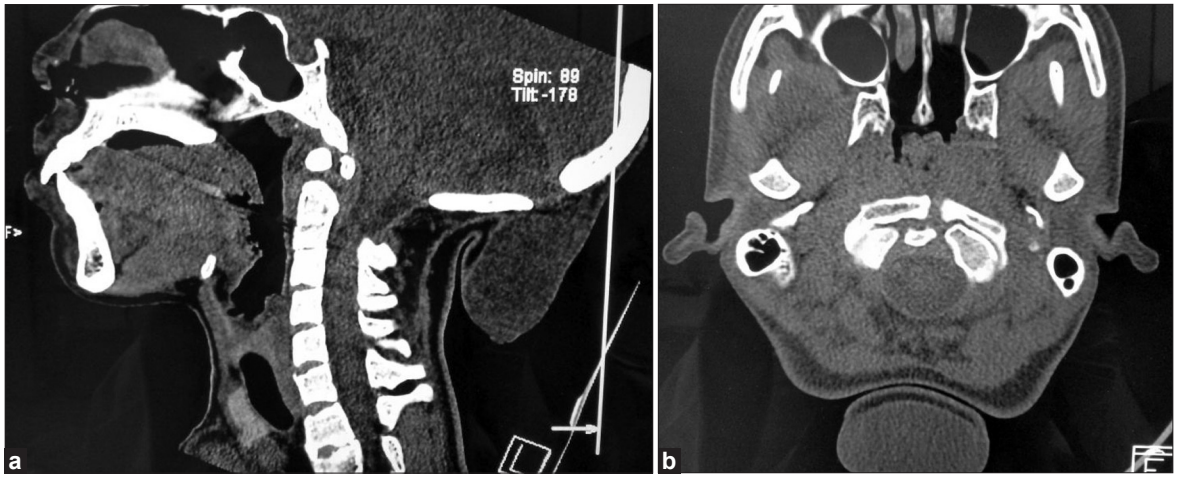


**Figure 5:** (a) Sagittal and (b) axial view CT CV junction show OS terminale and encephalocele with bone defect

## Discussion

True os-odontoideum is rare. Ossiculum terminale is nonunion of the apex at the secondary ossification center is much more common. Two anatomic types are orthotopic: Ossicle moves with the anterior arch of C1 and dystopic: Ossicle is functionally fused to the basion, and may subluxate anterior to the C1 arch.

Presentation of os-odontoideum included occipitocervical/neck pain, myelopathy and intracranial signs and symptoms.[7]

Most patients are neurologically intact and present with atlantoaxial instability which may be discovered incidentally. Many symptomatic and asymptomatic patients have been reported with no new problems over many years of follow-up.[8] Conversely, cases of precipitous spinal cord injury after seemingly minor trauma have been reported.[9]

Generally, patients with an occipital encephalocele are operated in the prone position with controlled ventilation and close temperature monitoring. Aspiration of the cerebrospinal fluid prior to incision in patients with large encephalocele helps in dissection of the sac. For a circular encephalocele with a small occipital bone defect, a transverse incision is ideal. The sac is separated from the flap. Patients in whom the encephalocele extends above and below the posterior fossa need a vertical incision. Sometimes, the brainstem and occipital lobe are present in the sac. Care must be taken to identify the contents of the sac. Rarely, the sagittal sinus torcular and the transverse sinus are in the vicinity of the sac. It is preferable to preserve the neural tissue. The dura is repaired meticulously to get a water tight closure. The dural defect can be repaired by using the pericranium as a graft. In neonates and infants, no attempt should be made to cover the bone defect by a bone graft.[10,11]

A large number of factors influence the outcome in patients with occipital encephaloceles. These are the site, the size, the amount of brain herniated into the sac, the presence of brainstem or occipital lobe with or without the dural sinuses in the sac and the presence of hydrocephalus.[12-15]

Previous authors described a 4-day-old boy was admitted with a large posterior fontanelle encephalocele. The baby had a small head with a circumference of 30 cm only and encephalocele with a circumference of 37 cm. Excision and repair of encephalocele was done without neurological postoperative complications or neurological deficit.[16] In our case, simultaneously presented with torcular encephalocele with os-teminale and there was only neck pain and no neurological deficit.

There was no hydrocephalus. X-ray of the cervical region showed os-terminale [Figure 4], split atlas, both anterior and posterior arch defect [Figure 5a and b].

The patient was operated in a prone position, and immobilized neck by hard cervical collar during surgery. At operation, there was gliosed brain inside the sac, which was excised. Bone defect was 3 cm in diameter. Dura was closed using 5-0 vicryl and wound was closed in multiple layers. Patient had an uneventful recovery was discharged on 8$^{th}$ postoperative day. She was followed-up after 15 days, when she had no problem.

We advised use a cervical collar and regular monthly follow-up at neurosurgery outpatient department, for os-terminale, which may need a surgery, if required.

## Conclusion

There is a rare association between torcular encephalocele and pro-atlas defect with Os-terminale. So this will bring attention to the world neuro scientist. Neural tube defect sometime presented with different type of congenital anomaly.

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