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- A** Study Design
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- C** Statistical Analysis
- D** Data Interpretation
- E** Manuscript Preparation
- F** Literature Search
- G** Funds Collection

## Giant Atretic Occipital Lipoencephalocele in an Adult with Bony Outgrowth

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### Summary

**Background:**

We present unique case of a giant extracranial atretic occipital lipoencephalocele in an adult patient with new bone formation within it which was not associated with any developmental malformation of brain. Resection of the lipoencephalocele was performed for esthetic reasons.

**Case Report:**

18 year old female patient presented to the surgery OPD with complains of a large mass in the occipital region present since birth. It was of size of a betel nut at the time of birth and gradually increased in size over a long period of time. It was painless and not associated with any other constitutional symptoms. On examination the rounded fluctuant mass was present in the midline in occipital region covered with alopecic skin with dimpling in the overlying skin. On MRI there was mass showing both T1 and T2 hyperintense signal area suggestive of fat component. Herniation of meninges and atretic brain parenchyma was also seen through a defect in the occipital bone in the midline. There was a Y shaped bony outgrowth seen arising from occipital bone into the mass which was quite unusual in association with an atretic lipoencephalocele.

**Conclusions:**

A large lipoencephalocele with bony outgrowth in an adult patient is a rare presentation of atretic occipital encephalocele.

**MeSH Keywords:**

Encephalocele • Meningocele • Occipital Bone

**PDF file:**

<http://www.polradiol.com/abstract/index/idArt/895453>

### Background

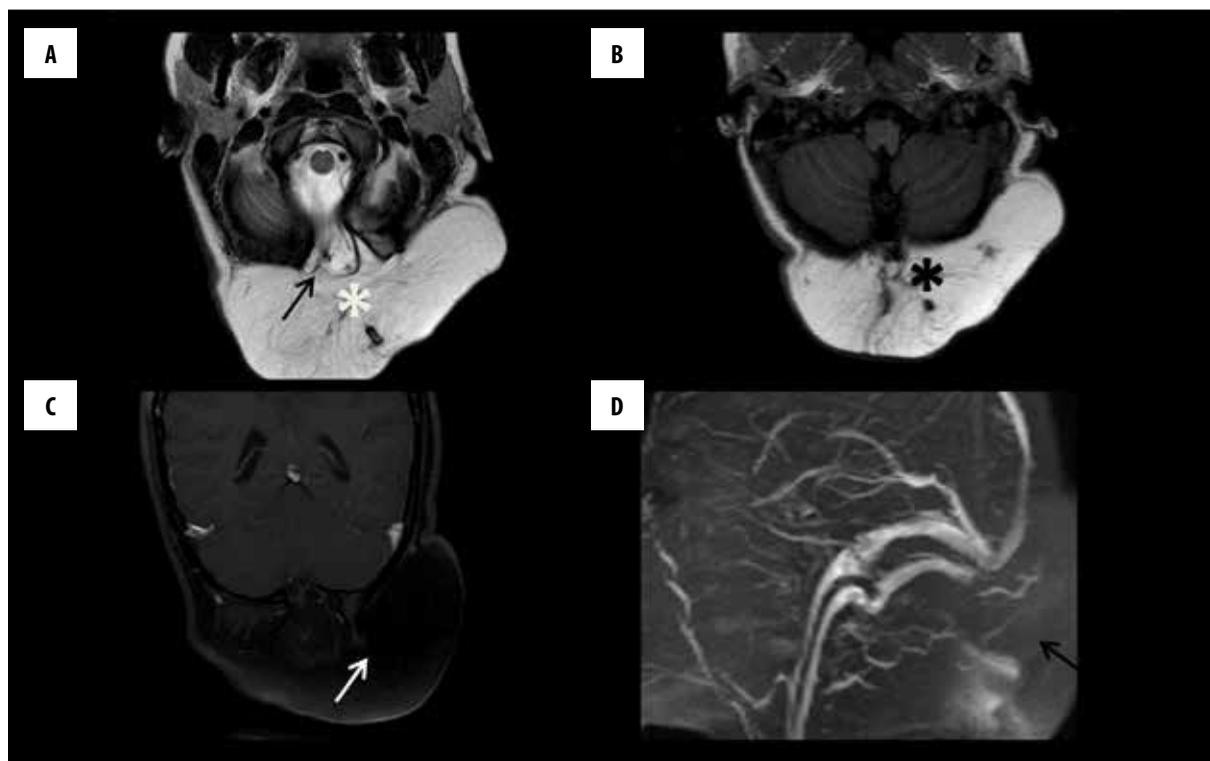
We present a unique case of a giant extracranial atretic occipital lipoencephalocele in an adult patient with new bone formation within it which was not associated with any developmental malformation of the brain. Resection of the lipoencephalocele was performed for esthetic reasons.

### Case Report

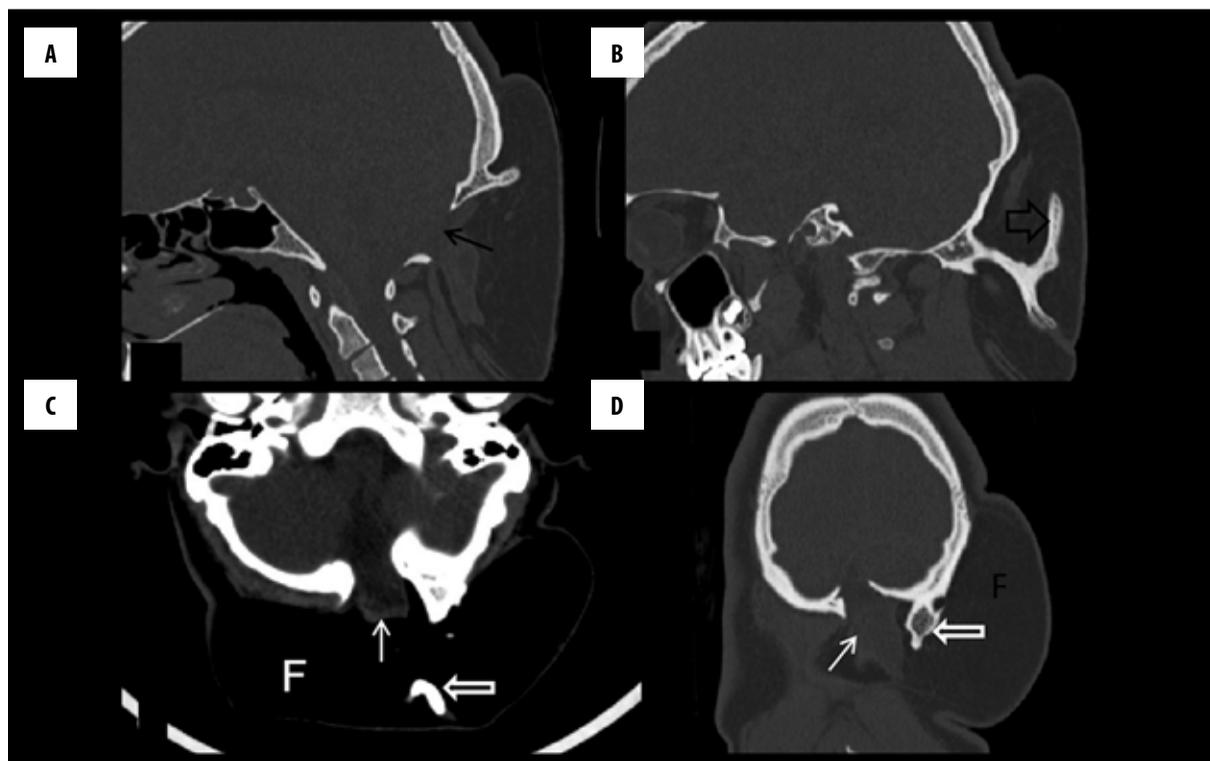
An 18-year-old female patient presented to surgery OPD with complains of a large mass in the occipital region present since birth. It was of size of a betel nut at the time of birth and gradually increased in size over a long period of time. It was painless and not associated with any other constitutional symptoms. There was a rapid increase in size of the mass in the last 2 months for which the patient consulted a neurosurgeon. On examination the rounded

fluctuant mass was present in the midline in the occipital region covered with alopecic skin with a dimpling in the overlying skin. CT and MRI of the brain were done for characterization of the mass. There was a large mass showing both T1- and T2-hyperintense signal area suggestive of fat component (Figure 1). Herniation of the meninges and atretic brain parenchyma were also seen through a defect in the occipital bone in the midline. There was a Y-shaped bony outgrowth seen arising from the occipital bone into the mass (Figure 2) which was quite unusual in association with an atretic lipoencephalocele. There was no communication of the mass with the cerebral venous system. The patient underwent resection of the atretic cephalocele (Figure 3) and had uneventful post-surgical recovery.

Atretic cephaloceles are small, skin-covered, subscalp lesions that contain meninges and neural and glial rests. Atretic cephaloceles probably represent a manifestation of



**Figure 1.** Atretic occipital lipoencephalocele in an 18-year-old female. (A, B) Axial T2WI and T1WI showing both T1- and T2-hyperintense mass (\*) in the occipital region with herniation of CSF, meninges and atretic brain parenchyma (black arrow) through a defect in the occipital bone. (C) Sagittal T1WI fat-saturated post-gadolinium image showing suppression of fat component of the mass (white arrow). (D) is a 3D TOF SPGR image in the venous phase showing no communication of the mass with the cerebral venous system.



**Figure 2.** Atretic occipital lipoencephalocele in an 18-year-old female. (A) Sagittal reformatted CT image in a bone window showing a defect in the occipital bone (black arrow) and (B) showing Y-shaped bony outgrowth from the occipital bone (hollow arrow). (C) Axial image in a modified window showing a fat-density mass (F), encephalocele (white arrow) and bony component (hollow arrow). (D) is a coronal reformatted bone window image showing a fat-density mass (F), encephalocele (white arrow) and bony component (hollow arrow).



**Figure 3.** Photograph of the resected specimen showing fatty tissue (A) and bony outgrowth (B).

defects in closure of a part of the neural tube [1]. The etiology of encephaloceles is controversial, with some groups supporting the hypothesis that these malformations result from the failure of neural tube closure [2]. Other groups suggest that the meninges, membranous skull, and blood vessels result from a common mesenchymal stem cell that rests between the neural tube and superficial ectoderm on day 9 of fetal life [3]. Insults during differentiation of these stem cells may result in a defect in the formation of the meninges and skull, and lead to the herniation of the intracranial contents through the defect [4,5]. Lipoencephalocele associated with bony outgrowth as seen in our case has not been previously reported as per our knowledge. Probable explanation of bony outgrowth is the origin from mesenchymal stem rest cells between the neural tube and superficial ectoderm cells on day 9 of fetal life.

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### Conclusions

A large lipoencephalocele with bony outgrowth in an adult patient is a rare presentation of atretic occipital encephalocele.