

A Rare Case of Right Frontal and Ethmoidal Bony Deformity With Schizencephaly

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Summary of Case

A seven month old male infant presented with boggy swelling over forehead since birth (Figure 1). The swelling measured 13 cms horizontally and 6 cms vertically. The bony defect could be felt along the medial margin of the swelling. His head circumference was 45 cms. Anterior fontanelle measured 2x2 cms. The findings of systemic examination were normal. He had uneventful antenatal and birth history. His growth and development were normal. His x ray skull lateral view showed defect in the frontal bone (Figure 2). CT Brain revealed frontal and right medial ethmoidal focal calvarial deformity, right frontal lobe atrophy and extra dural focal CSF collection with schizencephaly (Figure 3).

Agensis of skull bones, especially if associated with brain anomalies are incompatible with life. These defects are commonly located over parietal bones and the vertex between anterior and posterior fontanelles. Possible etiologies for skull bone defects are genetic,

chromosomal, defective midline ectodermal closure, placental infarcts and amniotic adhesions. The growth of cranial vault depends on the growth of underlying neural structures. In this case frontal lobe atrophy is noted on right side. Available treatment options are surgical and conservative treatment such as preventing infections and injuries. Long term neurodevelopment follow-up is required.



Fig 1: Photograph showing boggy swelling over forehead



Fig 2: Lateral skull radiograph showing bony defect in frontal area.



Fig 3: CT brain showing right frontal and right medial ethmoidal focal calvarial deformity, right frontal lobe atrophy and extra dural focal CSF collection with schizencephaly.

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