

Enlarged parietal foramina presenting as scalp swelling in an infant

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SUMMARY

“Enlarged parietal foramina” is a congenital malformation with autosomal dominant inheritance. The condition is usually self-limiting and doesn't require any treatment. However, it may also be associated with encephalocele, vascular anomalies or may be a part of syndrome. We present a case of enlarged parietal foramina in a child and discuss its imaging findings and the associated intracranial vascular malformations.

KEY WORDS:

Enlarged parietal foramina, persistent falcine sinus, encephalocele

INTRODUCTION

Enlarged parietal foramina occur due to a defect in membranous ossification of the skull and considered as a spectrum of cranium bifidum.¹ Recently mutations in MSX2 and ALX4 genes responsible for this condition have been identified.² Radiology plays an important role not only in confirming the condition but also to identify other associated vascular and intracranial abnormalities.¹ Treatment is usually not necessary unless associated malformations are present.² We discuss the imaging findings and associated abnormalities in an asymptomatic child who was noticed to have incidental parietal swelling.

CASE REPORT

A 2-month-old female child was noticed to have small symmetrical soft swelling over bilateral parietal regions. The swelling was noted incidentally by the parents at 2 months of age. The swelling was around 2 cm on either side and the overlying skin was intact. The head shape was unremarkable and the head circumference was normal at 37 cm. Anterior fontanelle was open and posterior fontanelle was closed. No dysmorphic facial features or other congenital abnormalities were identified. The child was otherwise clinically normal. The swelling was not demonstrated on the antenatal ultrasound screening and brain was reported to be normal. The child was delivered at 39 weeks by normal vaginal delivery without any use of instrumentation and birth history was unremarkable. Her birth weight and length at birth were 3.1 kg and 46 cm respectively. There was no family history of skull or bone abnormalities.

Radiograph of the skull (Figure 1) showed bilateral symmetrical radiolucencies in the parietal bones with mild adjacent soft tissue swelling. Further evaluation with Magnetic resonance imaging (MRI) of the brain was performed to rule out encephalocele and to identify intracranial abnormalities. MRI of the brain demonstrated the bilateral calvarial defects in the posterior parietal regions, with the surface of the brain attuning to the contour abnormality of the overlying skull (Figure 1). There was persistent primitive falcine sinus which drained into the superior sagittal sinus at the level of the skull defect and was associated with absence of the straight sinus (Figure 2). No encephalocele was identified. Diagnosis of bilateral enlarged parietal foramina was made based on MRI findings. The family was offered genetic counselling as the condition had autosomal dominant inheritance, but lost follow up.

DISCUSSION

Enlarged parietal foramina are usually bilateral and symmetrical which may persist into adult life. It is a disorder of membranous ossification of the skull bone, which leaves unossified portions in bilateral parietal regions. The brain is covered by dura mater and the overlying scalp is intact. It is considered a spectrum of cranium bifidum which includes more severe malformations such as encephalocele and meningocele.¹

Most of the cases go unnoticed and the documented prevalence of the condition varies from 1 in 15,000 to 1 in 50,000. The condition is shown to have autosomal dominant inheritance with high penetrance. Mutations in MSX2 and ALX4 genes are reported to be associated with enlarged bilateral parietal foramina.^{2,3} These genes encode for the homeodomain proteins required for multiple developmental process such as skull, face, hair follicle, genitals etc. Skull is more sensitive to the mutation in these genes. It is usually isolated abnormality, but it may be also associated with syndrome conditions like Proximal 11p deletion or Potocki-Shaffer syndrome, ALX4-related frontonasal dysplasia, Saethre-Chotzen syndrome, Cleidocranial dysplasia, Craniofacial dysplasia with genitourinary and skin abnormalities and Acromelic frontonasal dysostosis.^{2,4}

Antenatal ultrasound may detect the calvarial defect; however there is no herniation of dura mater or brain through the defect.¹ The bilateral symmetric appearance of

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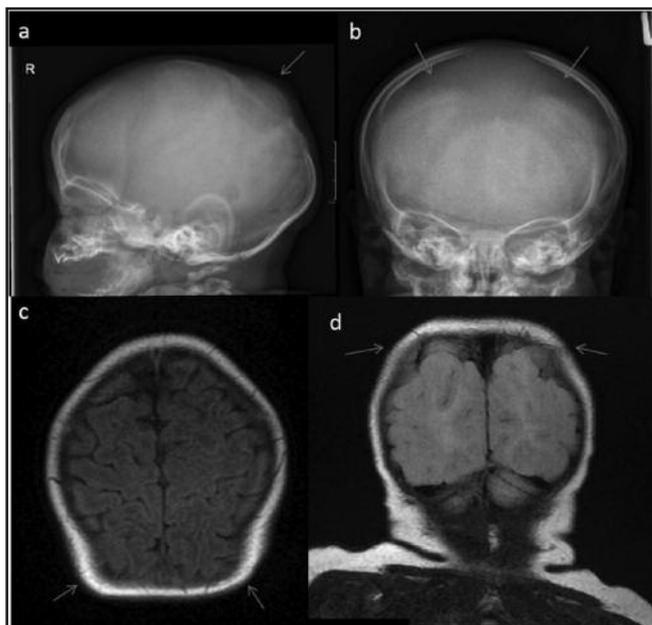


Fig. 1: X-ray skull lateral (a) and anteroposterior (b) views demonstrate the bilateral symmetrical radiolucencies in parietal regions (arrows in b) with mild adjacent soft tissue swelling (arrow in a). Axial T1 weighted image through high parietal region (c), coronal T2 weighted FLAIR image with posterior parietal bones (d) also demonstrate bilateral posterior parietal defects (arrows in c, d) with the brain parenchyma conforming to the contour abnormality.

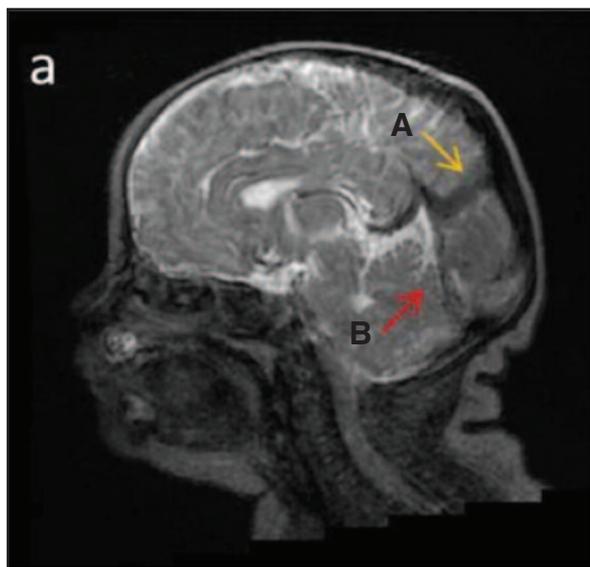


Fig. 2: Sagittal T2-weighted image demonstrates the persistent falcine sinus ('A') draining into the superior sagittal sinus. Also note the absence of straight sinus ('B').

enlarged parietal foramina is a main diagnostic indicator as opposed to the common unilateral causes of scalp swelling such as encephalocele or meningocele.³ Recognising this abnormality may be essential to avoid birth injury.²

The condition is usually asymptomatic except for the mild soft tissue swelling. It may also manifest as epilepsy because of associated meningeal or vascular abnormality.²

On radiograph, it appears as bilateral symmetric radiolucencies in the parietal regions; however accurate depiction may be hindered by overlapping bones.⁵ The condition is usually benign; however some of them have associated intracranial abnormalities. The common associated abnormalities include persistent falcine sinus with hypoplasia or atresia of the straight sinus as noted in our case. The cause of the venous anomalies is believed to be due to interruption in the normal development of the venous drainage system. Other intracranial abnormality which has been reported includes polymicrogyria.¹

There has been no definite association between the size of the defects and associated intracranial abnormalities. MRI is usually performed to rule out associated encephalocele and other intracranial abnormalities.¹ Treatment is usually conservative as the cranial defects usually become smaller with increasing age.^{1,2}

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