During embryonal development the human skeleton undergoes 2 types of ossification. Long bones mainly undergo endochondral ossification; other bones such as frontal, parietal, and squamous parts of temporal bones of the cranium undergo intramembranous ossification. During endochondral ossification a cartilage model of bone temporarily forms. In contrast intramembranous ossification undergoes direct ossification inside the vascularized membrane. Cranial bones grow and migrate to the midline during the development to completely cover the brain. Physiological openings persisting between these cranial bones at birth are known as fontanelles. There are 2 superolateral and posterior major fontanelles and 4 small fontanelles that exist during the newborn period.

The appearance of cranium bifidum, also known as cleft skull or enlarged parietal foramina, is characterized by the unsuccessful midline migration of the cranial vault. Most previous cases describe enlarged parietal foramina; there are only 4 previously reported cases in which the patients presented with a complete cranium bifidum.

Although the unsuccessful cranial development anomalies occur more frequently in eastern populations (< 3%) than in European and African populations (< 1%), reported cases are very rare.

Case Report

This infant boy was born at full term to a 24-year-old mother following an uneventful pregnancy. The baby’s Apgar score was 10. The boy had a normal appearance and did not have a skin or other developmental abnormality. The pediatrician present during the labor period noted the patient’s skull abnormality. Cranium bifidum was diagnosed based on physical examination and imaging findings. This was the first baby of the family, and there were no hereditary disease, intelligence, or medical problems in either parents or their families. The parents were nonconsanguineous. Blood tests and imaging studies of the abdomen and thorax did not reveal any other abnormalities.

At the physical examination no parietal bone was palpable, and both temporal and frontal bones were partly formed. No parietal bone was seen on radiographs or CT scans, and cranial magnetic resonance imaging did not reveal a parenchymal abnormality (Fig. 1). Punctuating ossification lines and ossification progression were observed on reconstructed 3D CT scanning (Fig. 2). The patient had an uncomplicated newborn period. The patient is still undergoing observation; the follow-up images have shown no successful ossification of the calvaria after 2.5 years (Fig. 3). To date, the patient has not undergone surgery. We expect to close the calvaria after brain growth is completed.

Discussion

Although there are some reports on parietal foramina in the literature, this condition is exceptionally rare and usually genetic. The autosomal-dominant hereditary transition has been emphasized in recent publications. Parietal foramina are bilaterally oval and symmetrically parietal bone defects. This defect normally closes during the 5th month of

Abbreviation used in this paper: CT = computed tomography.
gestation. At birth or later an accompanying scalp defect is often found. Enlarged parietal foramina are usually considered benign in nature and are generally asymptomatic in most instances, but they may be present either as an isolated disease or as a part of a syndrome.\textsuperscript{12,14–16} Often there are some scalp abnormalities associated with the clinical diagnosis.\textsuperscript{7,13} There are 2 types of parietal foramina. Both have been found to be caused by mutations in the \textit{MSX2} and \textit{ALX4} genes located on chromosomes 5 and 11.\textsuperscript{1} The authors of 1 report have claimed that parietal foramina and cranium bifidum are the same clinical entity.\textsuperscript{10} To our knowledge, however, the extremely rare occurrence of cranium bifidum may not support such a hypothesis. There seems to be familial inheritance in the majority of reported cases of parietal foramina; however, we were unable to find any reports of hereditary transition or familial occurrence for cranium bifidum.

In 1892 Greig\textsuperscript{4} first reported on 2 brothers with parietal foramina and normal levels of intelligence. In 1922 Goldsmith\textsuperscript{3} reported on a family of 56 members living in the US, 16 of whom had large parietal fontanelles. This is the largest series to have been described in the literature, and all family members were reported to have normal intelligence levels and no other abnormalities. Symmetric foramina have also been reported by Cohn\textsuperscript{2} (in 1924), LeLong and Bouquet\textsuperscript{9} (in 1932), Pendergrass and Pepper\textsuperscript{12} (in 1939), Laurenzen\textsuperscript{8} (in 1942), Terrafranca and Zellis\textsuperscript{16} (in 1953), Kite\textsuperscript{7} (in 1961), and Hollender\textsuperscript{5} (in 1967).

Murphy and Gooding\textsuperscript{11} first reported on a patient who exhibited apparent cranium bifidum progressing to parietal foramina that continued into adulthood. Although these authors could not show a genetic inheritance, they thought that cranium bifidum may later progress to parietal foramina during adolescence and into adulthood.\textsuperscript{10}

The underlying pathophysiology of cranium bifidum could not be explained in these rare cases, but there is a general opinion that it is identical to that of spina bifida.\textsuperscript{15} The interruption of development of calvarial bone formation

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure1.jpg}
\caption{Axial T2-weighted magnetic resonance image showing a normal appearance of the parenchymal region.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure2.jpg}
\caption{Reconstructed CT scan of the calvaria showing a mid-sagittal bone defect after birth.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure3.jpg}
\caption{Reconstructed CT scan showing persistent midsagittal bridging defect 2.5 years after diagnosis.}
\end{figure}
during the embryonic stage may result in an abnormality such as cranium bifidum. We conclude that such rare cases may be a result of coincidental mutations during the embryonic period.

References

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