Türk Kadın Sağlığı ve Neonatoloji Dergisi

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Case Report

Isolated Lacunar Skull Defect in a Newborn

Yenidoğanda İzole Laküner Kafa Deformitesi

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Abstract

The lacunar skull is a fetal ossification disorder first defined in the early 19th century, commonly associated with spina bifida and meningocele, and rarely seen as an isolated defect. It is characterized by defects on the inner table of the skull, in the form of shallow depressions or deep cavitations causes the skull to appear fenestrated because of mesenchymal dysplasia during intramembranous ossification. These bone defects may be present in any part of the calvarium but most frequently occur in the parietal and frontal bones. Clinically, lacunar skull may be suspected, but the diagnosis usually depends on the roentgenographic appearance. We report a newborn diagnosed with severe lacunar skull defect without any congenital malformations however accompanying severe vitamin D deficiency of maternal origin.

Keywords: Lacunar skull; newborn; congenital malformation

Öz

Laküner kafatası, ilk olarak 19. yüzyılın başlarında tanımlanan, genellikle spina bifida ve meningosel ile ilişkili, nadiren izole bir defekt olarak görülen bir fetal kemikleşme bozukluğudur. Kafatasının iç tabulasındaki yassı kemiklerde, intramembranöz kemikleşme sırasında gelişen mezenkimal displazisi nedeni ile oluşan, sığ veya derin boşluklarla karakterize bir kemikleşme kusurudur. Lezyonlar kafatasının herhangi bir bölümünde görülebilir ancak frontal, parietal ve üst oksipital kemiklerde daha belirgindir. Tanıda kafa grafisi bulguları önemlidir. Bu yazıda herhangi bir doğumsal malformasyonu olmayan, kafatasında büyük laküner defektleri olan ve ağır D vitamini eksikliği eşlik eden bir yenidoğan olgu sunulmuştur.

Anahtar Kelimeler: Laküner kafatası; yenidoğan; doğumsal anomali

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1. Introduction

The lacunar skull is mesenchymal dysplasia of calvarial ossification in which the fetal skull appears fenestrated. This ossification defect, also known as Luckenschadel, is characterized by shallow or deep lacunas in the membranous bones of the skull. Lacunar skull is generally associated with underlying pathological conditions, most common neural tube defects (1-3). However, the lacunar skull was reported as an incidental finding in normal newborns as early as 1924, but after then it was rarely defined as an isolated defect (3). Here in we report a newborn with large lacunar defects in the skull without any congenital malformations.

2. Case

A male infant was born by normal vaginal delivery to a 17-year-old mother at 37 weeks of gestation. The mother was not regularly followed up during the antenatal period.

There was no history of consanguinity or family history of malformations. Apgar scores were 8 and 9 at one and five minutes, respectively. The skull was palpated so soft at first physical examination and he was admitted to the neonatal intensive care unit for detailed examination.

Birth weight was 2700 g (10^{th}-50^{th} percentile), length was 46 cm (10^{th}-50^{th} percentile), and head circumference was 33 cm (10^{th}-50^{th} percentile). Overall the skull was very soft on palpation, the anterior fontanelle was large, soft and it was connected with the posterior fontanelle. The sagittal suture was wide on the parietal bone and extended into the occipital bone. Bone was not palpated in the temporal region. Otherwise there was no remarkable finding on physical examination.

On blood biochemistry analysis serum calcium (9.8 mg/dl, normal range: 8–9.6), phosphorus (4.7 mg/dl, normal range: 2.5–5) and alkaline phosphate (286 U/L, normal range:75-316) levels were normal. The endocrine evaluation revealed normal serum parathyroid level (58 ng/L, normal 10 to 65 ng/L), but a very low 25-hydroxyvitamin D level (3.88 ng/mL, normal 20–80 ng/mL). Then the mother was evaluated for vitamin D deficiency. Maternal vitamin D level was 1.09 mg/dL, which shows a severe deficiency. The bone mineral density (BMD), assessed by dual-energy X-ray absorptiometry (DXA), revealed Z-score - 2.3, which showed a peak bone mass is below average. Serological tests for TORCH and syphilis were all negative.

Skeletal roentgenograms showed normal radiological findings. The skull X-ray revealed multiple large oval lucent areas (Figure 1). Cranial computed tomography scan demonstrated general bone defects (Figure 2), without any evidence of hydrocephalus. Three-dimensional computed tomography scan revealed multiple oval and round lacunae at the parietal and frontal areas which were in the tendency to join (Figure 3a,3b). Cranial and abdominal ultrasonography revealed normal findings. Echocardiography revealed patent foramen ovale. Ophthalmological and audiometric examinations were normal.

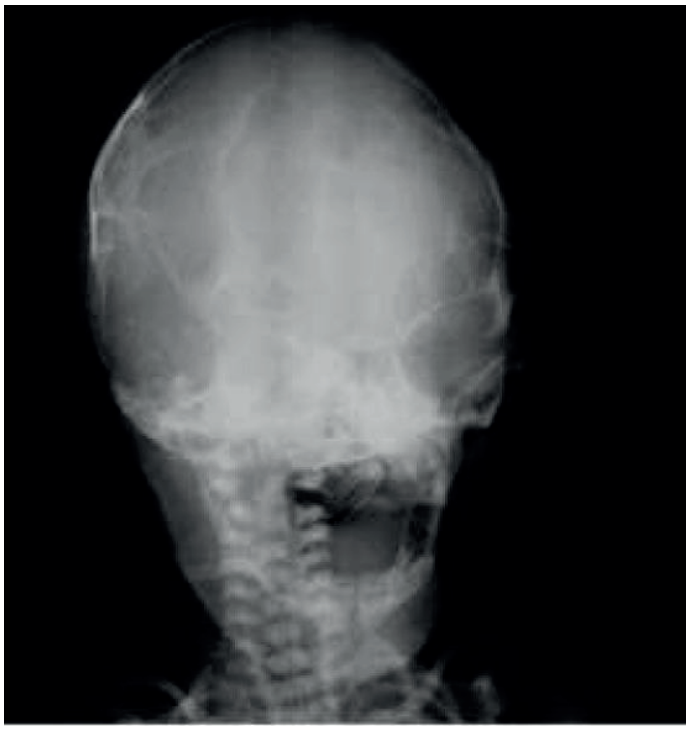

Figure 1. Skull X-ray of the patient demonstrating oval lucencies surrounded by dense ridges

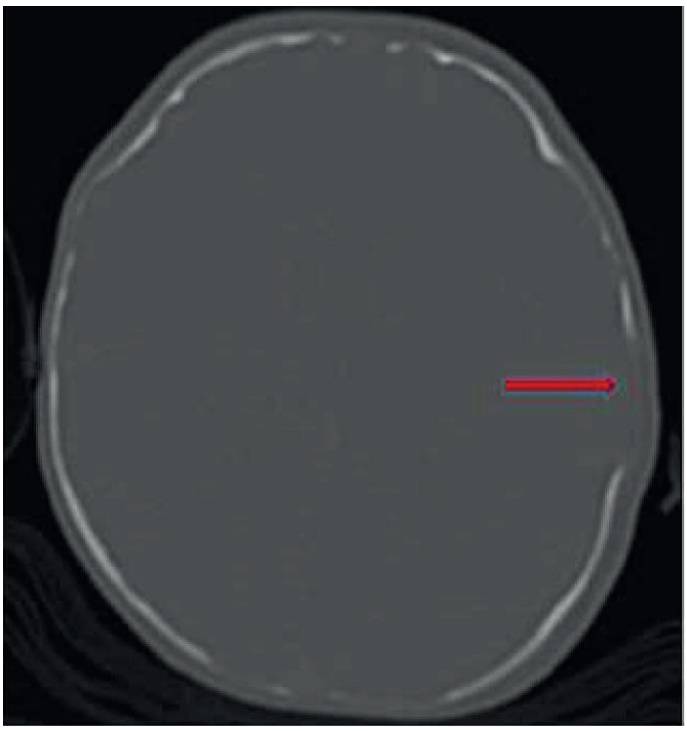

Figure 2. Computed tomography scan of infant demonstrating bone defects in the inner table of the skull

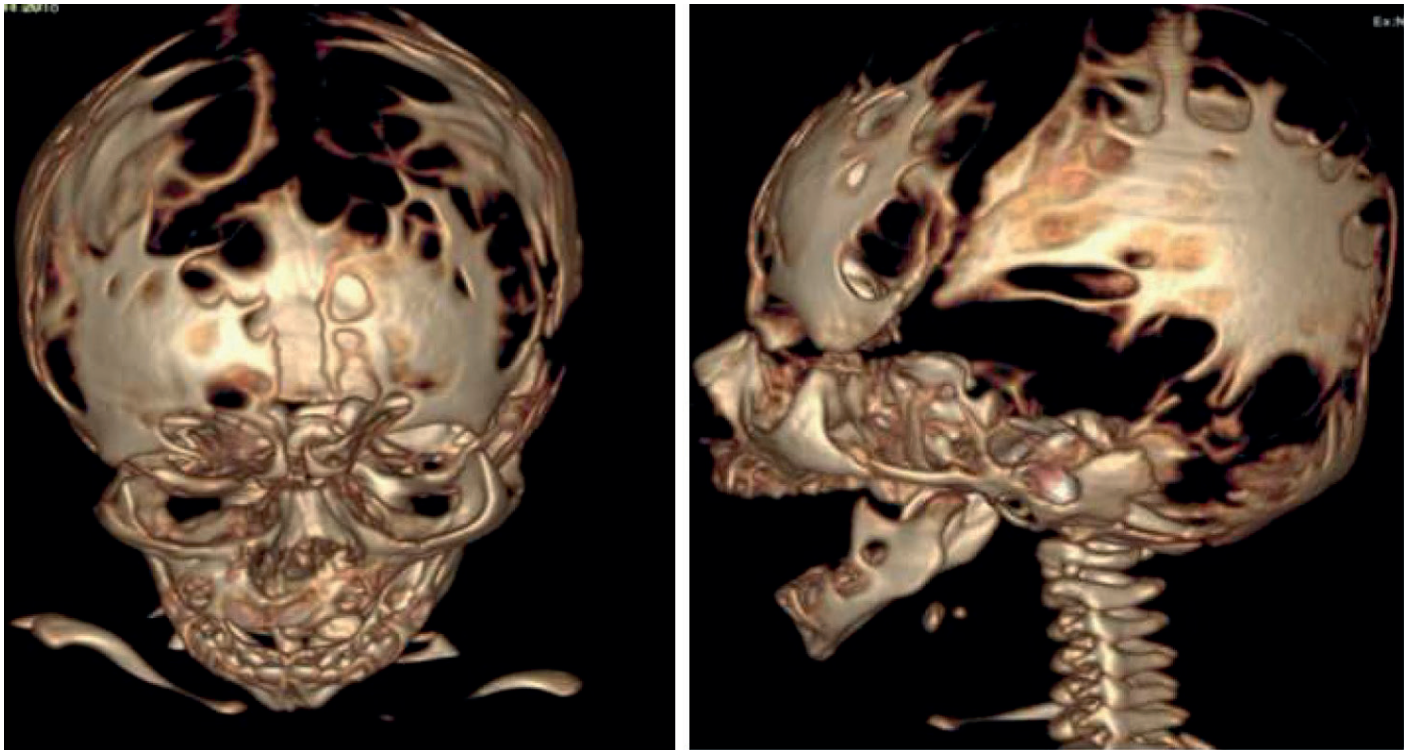

Figure 3a,3b. Three-dimensional computed tomography scan revealed multiple oval and round lacunae at parietal and frontal areas which were in the tendency to join

These clinical, laboratory, and radiological findings suggested the diagnosis of lacunar skull accompanying severe vitamin D deficiency of maternal origin.

The infant was started on vitamin D at 800 IU daily. The mother was also treated with calcium and vitamin D supplementation. The infant remained active and stable on follow-up. Serum calcium, phosphorus, and alkaline phosphate levels also remained at normal levels during a high dose of vitamin D treatment. He was discharged on vitamin D supplementation with breastfeeding and referred to the outpatient clinic for follow-up care, however, no further information was available for this infant.

Informed consent was obtained from the parents at admission.

3. Discussion

The lacunar skull defect was first defined at the end of 19th century in infants with meningocele (1,2). The groups of round and oval pits on the inner surface of the skull in these babies were named Luckenschadel in German literature. Then this fetal ossification disorder characterized by shallow or deep lacunaes in the membranous bones of the skull was called craniolacunia or lacunar skull, considering a term to be of international usefulness (3).

A general reduction in bone thickness and delay in ossification in flat bones of the skull appears in cranio lacunia. Thinning of the skull generally involves the inner table and diploe, and rarely extends to the outer table. The defects involve only the inner table of bone and are not palpable from the outside of the skull in craniolacunia. However, when the defects extend to outer table and the individual lacunaes coalesce into larger defects, the term craniofenestrae is used and these can be palpated from the outside (3,4). In the present case, cranio fenestrae is more accurate term to define the large skull defects.

Clinically, lacunar skull may be suspected, but the diagnosis is usually based on the roentgenographic appearance. On conventional X-ray, craniolacunia has a characteristic ‘‘honeycomb’’ or “soap-bubble’’ appearance, with large rounded areas of decreased bone density bordered by a web-like the pattern of thicker bone. However, three-dimensional computed tomography scan clearly reveals the abnormal bone structure (3). Although oval lucent areas were apparent on the X-ray of our patient, multiple oval and round lacunaes were clearly detected at three-dimensional computed tomography scan.

The lacunar skull is usually associated with spina bifida, encephalocele, meningocele or myelocele. Other congenital developmental anomalies such as deformities of the ribs and extremities, cleft palate, microcephaly, and craniosynostosis may be present (1,2). Physical examination of the present case revealed no dysmorphic features, lacunar skull was an isolated defect without associated any malformation.

The etiology of the lacunar skull has been widely investigated, but no distinct explanation has been reported. The proposed mechanism of the development of craniolacunia is, primary disturbance in the development of the membranous bones of the cranium. Since the skull ossification process is the membranous ossification, the lacunaes are often seen on frontal, parietal, and upper occipital bones of the skull. The base of the skull shows normal development, as do the long bones which arise from the endochondral bone formation (3,5). In the present case, normal findings were seen on radiographs of long bones.

Development of lacunar skull begins in utero but disappears by six months of age. Spontaneous regression may relate to remodeling in response to normal expansion of cerebral tissue (2,3). In the present case, after discharge, no further information was available for the patient so we don't have any information about the course of the skull defect.

The differentiation of oval, large lucencies seen in paediatric skull radiograph includes, convolutional markings and copper beaten skull. However, true convolutional markings are normal markings seen as areas of decreased density and occur later, after sutural closure. The copper-beaten skull is a feature of raised intracranial pressure (5-7). There was no sign of increased intracranial pressure in our patient and any evidence of hydrocephalia on the cranial computed tomography scan.

As the lacunar skull is an X-ray finding, prenatal diagnosis is possible via X-ray pelvimetry. However X-ray is rarely used in current obstetric practice so, the lacunar skull is less often recognized by obstetricians (8). Recently, the appearance of this deformity has been noted in the descriptions of Chiari II malformations in the neurosonography literature (9). The diagnosis of a lacunar skull at the antenatal period is important in the management of the delivery of these babies because clinicians should be more cautious when using forceps or vacuum extractors to assist vaginal delivery in a fetus with a lacunar skull deformity. This case was born by uncomplicated vaginal delivery and there was no sign of intracranial haemorrhage.

The present case also has severe vitamin D deficiency of maternal origin. However, there were no signs of vitamin D deficiency in this neonate.

We suppose that lacunar skull deformity and severe vitamin D deficiency are incidental findings in our patient, however as the lacunar skull is a fetal ossification defect, severe vitamin D deficiency may have caused these defects to be more severe. The effects of vitamin D on osteocyte number and connectivity have been suggested previously (10).

To the best of our knowledge, this is the first case of craniolacunia with very severe skull defects in a newborn without any congenital abnormalities.

Author contribution

Study conception and design: SK, ND and BTC; data collection: SK, BTC and YTY; analysis and interpretation of results: ND and AYB; draft manuscript preparation: SK, ND and AYB. All authors reviewed the results and approved the final version of the manuscript.

Ethical approval

The written consent was received while admitting to the hospital.

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Conflict of interest

The authors declare that there is no conflict of interest.

Yazar katkısı

Araştırma fikri ve tasarımı: SK, ND and BTC; veri toplama: SK, BTC and YTY; sonuçların analizi ve yorumlanması: ND ve AYB; araştırma metnini hazırlama: SK, ND ve AYB. Tüm yazarlar araştırma sonuçlarını gözden geçirdi ve araştırmanın son halini onayladı.

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