A Case of Growing Skull Fracture with Appearance of the Sinking Skin Flap Syndrome

Çöken Cilt Flebi Sendromu Görünümünde Büyüyen Kafatası Kırığı Olgusu

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ABSTRACT A growing skull fracture is a late complication of a traumatic linear skull fracture. Untreated growing skull fractures can be associated with headache and progressive neurological deficits. In this case report, an unusual growing skull fracture mimicking sinking skin flap syndrome is presented. An 11-year-old girl, who had had a linear skull fracture 6 years previously, presented with headache, progressive left hemiparesis and right parietal scalp swelling on supine position. The scalp overlying the bone defect was sinking on upright position like the sinking skin flap syndrome. The growing skull fracture, revealed on the plain X-ray and computed tomography, was treated by duraplasty and cranioplasty with methylmetacrilate. The symptom of headache resolved and her walk improved after the treatment.

Key Words: Craniocerebral trauma; skull fractures

ÖZET Büyüyen kafatası kırığı travmatik lineer kafatası kırığının geç bir komplikasyonudur. Tedavi edilmeyen büyüyen kafatası kırıkları, baş ağrısı ve ilerleyici nörolojik bozukluklarla ilişkili olabilir. Bu olgu sunumunda çöken cilt flepi sendromunu taklit eden görünümüyle nadir rastlanan bir büyüyen kafatası kırığını sunuyoruz. Altı yıl önce lineer kafatası kırığı geçiren 11 yaşında kız çocuğu baş ağrısı, ilerleyici sol hemiparezi ve saçlı derinin sağ paryetal kısmında sırt üstü yatınca oluşan şişme nedeniyle başvurdu. Kemik defektinin üzerini kaplayan saçlı deri, çöken cilt flepi sendromuna benzer şekilde dik pozisyonda çukurlaşıyordu. Düz grafi ve bilgisayarlı tomografide ortaya koyulan büyüyen kafatası kırığı, metilmetakrilat ile duraplasti ve kranioplasti ile tedavi edildi. Tedaviden sonra baş ağrısı semptomu düzeldi ve hastanın yürümesi rahatladı.

Anahtar Kelimeler: Kraniyoserebral travma; kafatası kırıkları

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A growing skull fracture (GSF) is a progressive enlarging diastatic fracture, which occurs as a rare complication of head trauma almost exclusively in infants and children.[1] Brain contusion, porencephalic cyst formation and alterations of cerebrospinal fluid circulation generally accompany the dural defect. The incidence of GSF ranges from 0.05 to 1.6% of pediatric skull fractures, and the patients generally present with scalp swelling and neurological deficits.[2,3] It is known that the scalp overlying the defect can appear flat or sunken.[2,4] However, orthostatic sinking of the scalp over the GSF has not been reported.

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The sinking skin flap syndrome (SSFS) is an uncommon long-term complication of craniectomies performed for different disorders, such as intraoperative brain edema, traumatic skull fractures, subdural hematomas and gun-shot wounds. The syndrome is characterized by progressive neurological deterioration of the patient and requires cranioplasty.[5]

The aim of this case report is to emphasize a late complication of a diastatic linear fracture, GSF, presented with appearance of SSFS. The patient we reported had an unusual GSF, the scalp overlying the bone defect displayed orthostatic sinking SSFS. Other symptoms of the patient included progressive headache and left hemiparesis.

CASE REPORT

An 11-year-old girl admitted with headache and right parietal scalp swelling, which was sinking at upright position. She had a history of hospitalization for head trauma following a fall when she was 5 years old. Cranial computed tomography (CT) scan performed during hospitalization had revealed a linear fracture on the right parietal region, near to the vertex. She also had a cerebral contusion and a small subarachnoid hemorrhage at the same site. The patient had mild left hemiparesis while her general condition was good, and she was discharged after having conservative therapy for 16 days. She has received no medical care thereafter until she has developed headache and scalp swelling 6-months previously. Her headache was progressive, and she had some difficulty when walking and holding an object with her left hand. Her symptoms had got worse since 6 months.

On admission, her physical examination was normal, except for the large defect on the right parietal bone extending to the vertex, which was 10-cm in length and 2-cm in width. When the patient was in upright position, the scalp overlying the defect was sank, whereas scalp swelled in the supine position (Figure 1). Neurological examination revealed 4/5 motor strength in her left upper and lower extremities.

The diagnosis of a growing skull fracture was confirmed with characteristic radiological features. The plain X-ray revealed an irregular elliptical skull defect, the margins of which were scalloped (Figure 2). The CT scan demonstrated not only the extent of the bone defect but also the area of encephalomalacia (Figure 3). Magnetic resonance imaging (MRI) confirmed that the herniating tissue was a cystic collection, not the brain parenchyma, and no defect encroached upon the sagital sinus (Figure 4).

The patient was operated under general anesthesia. Duraplasty and cranioplasty with methylmetacrilate were performed. Recovery was uneventful, and the patient was discharged with

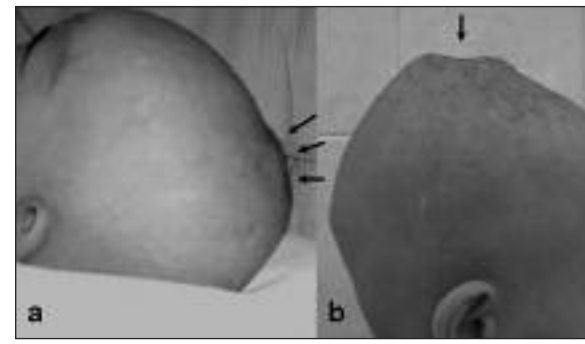

FIGURE 1: a. Photograph of the head at supine position demonstrates right parietal swelling (arrows). **b.** The swelling disappears and the scalp overlying the bone defect sinks at the upright position (arrow).

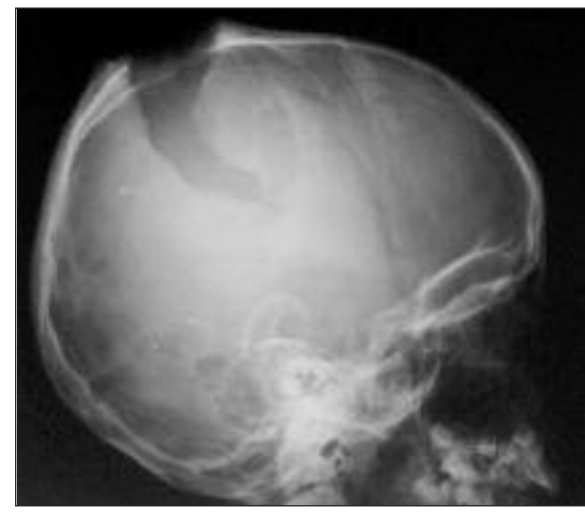

FIGURE 2: An irregular scalloped defect with sclerotic margins is seen on the vertex on lateral head x-ray.

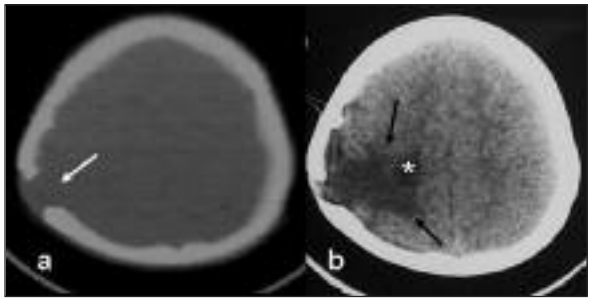

FIGURE 3: a and **b.** Head CT scan shows the bone defect (white arrow) with encephalomalacia (black arrows) around the atrium of dilated lateral ventricle (*).

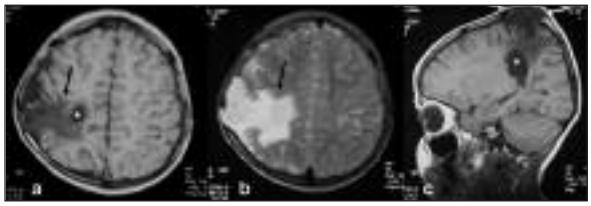

FIGURE 4: Axial T1- **(a)** and T2- **(b)** weighted MRI shows right frontoparietal encephalomalacia (black arrows). Sagital T1- **(c)** weighted MRI demonstrates the cystic collection (white arrows) herniating through the bone defect. The atrium of the lateral ventricle is dilated (*).

resolution of the headaches. The skull deformity was also improved and the orthostatic sinking disappeared. Her walk also improved two weeks after the operation.

DISCUSSION

A GSF, also leading to expanding skull fracture, craniocerebral erosion, leptomeningeal cyst, post traumatic bone absorption, traumatic ventricular cyst and cephalhydrocel, is an unusual complication of skull fractures. The most common causative factors are either a fall or a motor-vehicle accident. Child abuse, birth injury, previous cranial surgery and minor closed-head injury can also lead to GSF.[6] The skull defect is almost always associated with a dural tear. The dura adheres more tightly to bone during infancy and early childhood, and hence it is more easily torn in infants and children when a fracture occurs.[2] The arachnoid bulges through the dural defect by the pulsation of cerebrospinal fluid (CSF), and cyst formation is postulated to take place thereafter.[7-9] Crocker at al. reported a case of growing skull fracture in the absence of a dural tear, which was seen after a head injury with extradural haematoma as a previously unreported cause.[10]

GSF mostly occurs in the parietal bone and rarely occurs in the temporal or occipital bones. Location in the posterior fossa and intradiploic GSF development are very uncommon[11]. The diagnosis is easily established with characteristic clinical and radiological findings. The majority of patients are presented with skull defect and/or swelling, persistent or progressive neurological deficits and seizures. The time interval between the trauma and presentation ranges from less than six months to more than 20 years.[12-15] Local pain is rarely the only symptom, and is considered to be due to traction on the blood vessels in the trapped arachnoid membrane.[2] However, headache is a common presentation of untreated GSFs in late stages.[14]

The plain X-ray reveals an irregular oval or elliptical skull defect. The fracture line can be seen at one end of the defect. The margins are usually everted: scalloped and sclerotic. CT displays the bone defect, cyst and parenchymal injury and ventricular morphology. Three-dimensional reconstructions can help the plan of surgery. MRI, with its unique ability of multiplanar imaging and soft tissue discrimination, shows the exact nature of herniated tissue (normal brain parenchyma, encephalomalacia and/or leptomeningeal cyst) as well as the relationship of the defect with the dural sinuses.[13]

A GSF can be associated with a leptomeningeal cyst, encephalomalacia, cystic parenchymal lesions, porencephaly, unilateral ventricular dilatation, hydrocephalus, pseudomeningoencephalocele, intraventricular cyst and lytic bone lesion mimicking a skull tumor.[2,13,16,17] In addition, in this report orthostatic sinking of the scalp over the GSF such as SSFS is shown.

The SSFS has been described as one of the causes of neurological deterioration after a large craniectomy during a neurosurgical procedure due to the compression of the skin flap onto the brain. This syndrome includes dizziness, undue fatigability, vague discomfort at the site of the defect, a feel-

ing of apprehension and insecurity, mental depression and neurological deficits. In a large majority of cases, most of the symptoms are relieved after cranioplasty.[5]

The SSFS is considered to be caused by the relationship between atmospheric pressure, CSF, and cerebral blood flow (CBF) at the bone defect site. There are three components in pathogenesis of SSFS: cerebrospinal hydrodynamic changes, cerebrovascular or hemodynamic changes and metabolic changes. It has been postulated that the patients with large cranial defects have increased intracranial pressure, decreased CSF movements and decreased CBF.[18,19] The aim of the treatment is to break down the cascade of atmospheric pressure, to expand the brain, to push the flap out and to reconstruct a convex skin flap.[20]

In this case report, a late complication of a traumatic linear skull fracture, a growing skull fracture with the apperance of the SSSF is presented. The orthostatic sinking of the scalp is probably associated with the large encephalomalacia and the low-pressure fluid filling the bone defect. The neurological symptoms of the patient can be attributed to the late complications of GSF. It is known that untreated GSFs may cause delayed-onset neurological manifestations, in addition to cranial growth asymmetry. GSFs, even when they are discovered incidentally without any neurological deficits, should be operated on as soon as possible to prevent further brain destruction.[14] Compression of the sinking scalp onto the brain might also have a role in progression of headache and neurological deficit in this patient. Therefore, we believe that another aim of surgery in this patient is to break down the cascade of atmospheric pressure onto the brain due to the sinking scalp.

The principles of the GSF treatment are the repair of the dural defect with or without a graft, and the use of cranioplasty in significant skull defects.[13,14,21,22] Hayashi et al. have shown that methylmethacrylate is a suitable material for cranioplasty for GSF.[23] Duraplasty and cranioplasty with methylmethacrylate were performed in order to correct the skull deformity and stop the progression of the neurological deficits of our patient. Two weeks after the surgery, headache of the patient relieved and her walk improved, however left upper extremity weakness remained the same.

In conclusion, a growing skull fracture can result in orthostatic sinking of the scalp overlying the bone defect, and it can be associated with headache and progressive neurological deficits. In infants and children, linear skull fractures should be monitored until definite skull bone consolidation is obtained to prevent progressive lesions of both skull and brain tissue, as illustrated in this case. When complications occur, early treatment is necessary to prevent progressive disease.

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