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Subperiosteal Orbital Abscess Complicating Infantile Osteopetrosis: MRI Findings

Case Report

İnfantil Osteopetrozisli Olguda Gelişen Subperiostal Orbital Abse: MR Bulguları

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ABSTRACT

Osteopetrosis (Albers-Schonberg disease) is a rare sclerosing bone disorder in which bone marrow is obliterated. Owing to pancytopenia, infections can occur. In this study an infantile osteopetrosis complicated with subperiosteal orbital abscess is presented and magnetic resonance imaging findings described which to our knowledge no such togetherness has been mentioned before.

Key words: *Osteopetrosis, Orbital subperiosteal abscess, Magnetic resonance imaging*

ÖZET

Osteopetrozis(Albers-Schonberg hastalığı), kemik iliği miktarında azalmaya yol açan nadir bir sklerotik kemik hastalığıdır. Pansitopeniye bağlı enfeksiyonlar gelişebilir. Bu olgu sunumunda, daha önce birlikteliği tanımlanmamış olan infantil osteopetrozisli olguda gelişen subperiostal orbital absenin MRG bulgularını tanımlamayı amaçladık.

Anahtar Kelimeler: Osteopetrozis, *Orbital Subperiostal Abse, MR*

INTRODUCTION

Albers-Schönberg osteopetrosis, a rare heritable bone disease, is caused by failure of the osteoclasts to resorb bone (1,2). This results in diffuse sclerosis of the whole skeleton accompanied by pathological bone fragility and delayed physical development, profound intractable myelophthisic anaemia, neurological deficits, blindness and hearing loss. Osteomyelitis, especially of the jaws and the skull, owing to pancytopenia can occur (3,4,5). In this study we present an infantile case of osteopetrosis complicated with orbital cellulitis and subperiosteal orbital abscess. To our knowledge, osteopetrosis associated with subperiosteal orbital abscess in an infant has not been mentioned before in the English Language literature.

CASE REPORT

A 5 year-old girl who had hepatosplenomegaly, anemia, thrombocytopenia, leukocytopenia and recurrent infections, admitted to the hospital with bilateral proptosis apparent on the left side and a new onset of painful, swollen and red left eyelid . The movements of the left eye had reduced with impaired vision. The eyeball had a swollen, hazy appearance. Her having been taken osteopetrosis diagnosis was learned from her history. In her physical examination hepatosplenomegaly and pancytopenia were detected. These findings were thought to have connection with osteopetrosis diagnosis. Thinking upon preceptal cellulitis, in order to show the intracranial extention of the disease, an MRI study of the patient with 1.5 T MR scanner, in three planes with pre-post contrast SE T1, FSE PD and T2 sequences, was performed. The MR images showed diffuse decrease in diploe spaces and thickening of the calvarial bones, with hypointensity in all sequences. Craniofacial ratio was increased for cranium. Posterior fossa was too small and foramen magnum diameter was diminished (Figure 1).

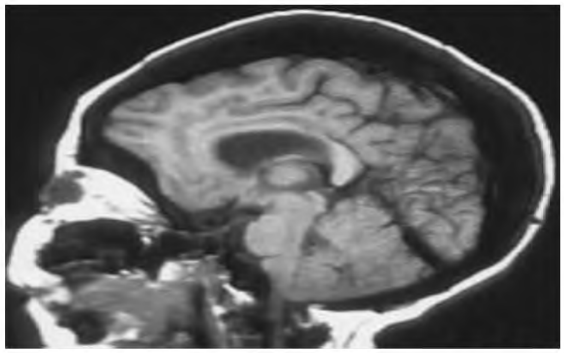

Figure 1: *Involvement of skull in osteopetrosis. Thickness in the inner and outer tabulaes of calvarial bones, and reduction in the diploic space are seen in this midsagittal SE T1W MR image. Posterior fossa is smaller than normal and foramen magnum is narrowed.*

Bilateral proptosis evident on the left side was present. In the medial side of left orbita, in extraconal space, just in the vicinity of laminae papiracea, ovoid shaped subperiosteal abscess was detected. The lesion was hypointense on T1 WI and hyperintense on T2 WI with peripheral enhancement after IV Gd DTPA injection. Also on the left side, preseptal cellulitis was detected with the same signal characteristics and contrast enhancement in preseptal and nasal regions (Figures 2,3,4).

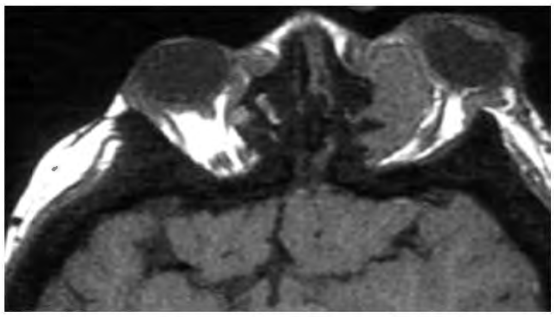

Figure 2: *In the left orbital extraconal space, just in the vicinity of laminae papiracea, a hypointense ovoid lesion deplacing rectus muscles and optic nerve to lateral is seen (axial SE T1W MRI section taken from orbital level). Moreover the left eyelid is thickened and seen hypointense. There is evident proptosis on the left side.*

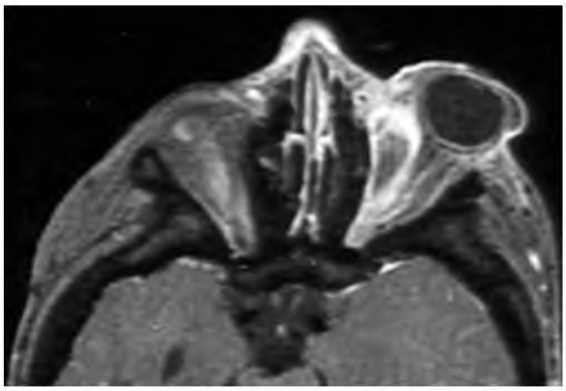

Figure 3: *There is peripheral enhancement in the intraorbital lesion accorded with subperiosteal abscess in this contrast enhanced fat sat SE T1 W axial image. There also is seen diffuse contrast enhancement in preseptal and prenasal region.*

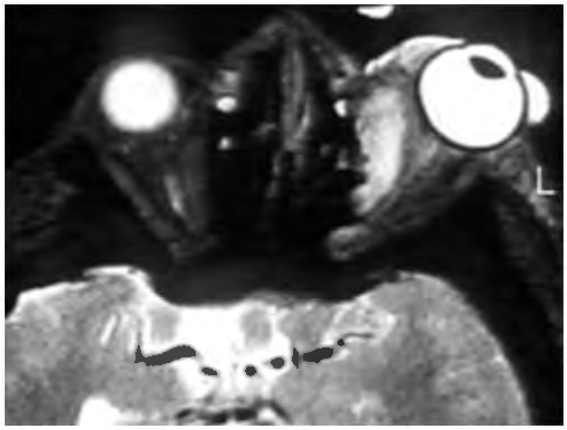

Figure 4: *In axial fat sat SE T2 W image, the lesions described in left orbita and eyelid present hyperintens signal characteristics.*

DISCUSSION

Osteopetrosis, also known as Albers Schönberg disease, is a rare, congenital dysplasia of the skeleton in which the osteoclasts have defective function (6). Since osteoclasts do not function properly, the equilibrium between production and destruction abolishes. The bones of afflicted patients become sclerotic and show modelling defects resulting in either a decrease or obliteration of the marrow cavity and resultant pancytopenia. Other clinical manifestations include bony deformities, cranial nevre palsies from bony overgrowth, pathological fractures, osteomyelitis, and hepatosplenomegaly secondary to extramedullary hematopoiesis (7). Osteomyelitis in maxilla, jaw is reported in the literature (1,3,4,5). Srirompotong S. et al. reported one patient with childhood form of osteopetrosis presented with chronic otitis media and brain abscess. In our patient, there was subperiosteal orbital abscess and preseptal cellulitis. Subperiosteal cellulitis or abscess formation is often associated with multifocal sinusitis with either direct spread through the sinus wall or via thrombophlebitis. Specifically, this most commonly results from inflammatory changes involving the ethmoid air cells or frontal sinuses. A medial subperiosteal abscess (SPA) of the orbit is the most common serious complication of sinusitis in children. Direct extension of infection through congenital osseous dehiscences or involvement of the thin bony walls of the orbit by osteomyelitis can lead to the formation of subperiosteal abscess. But in our case no such a relationship has been detected because the neighborhood sinuses were all normal aerated (Figure 4). We think that infections in osteopetrosis are the consequence of pancytopenia. But infection preference to orbita and preseptal section in osteopetrosis is uncertain. In the presence of craniofacial and orbital infections as in our case, along with other systemic findings, one should also suspect osteopetrosis. In our osteopetrosis case, the subperiosteal orbital abscess has no different imaging findings from the analogous', seen in otherwise healthy individuals. Orbital cellulitis is characterized hypointense on T1 W and hyperintense on T2 W MR images. Fat saturated T1 weighted images after contrast agent injection, is the most sensitive technique in the early detection of orbital inflammation (8,9). The distinction between SPA and the more benign preseptal infectious disease is difficult to make clinically especially in a young child in whom an ophthalmological evaluation is often difficult.

Computerized tomography (CT) and magnetic resonance imaging (MRI) is the

investigation of choice in making this distinction. Both CT and MRI show the extent of involvement of soft tissues by infection, but CT is more precise in demonstrating the bony changes.
Whether accompanied by osteopetrosis or not, subperiosteal inflammatory disease of the orbit is initially treated with intravenous antibiotic therapy with surgery reserved for those patients who do not respond to medical treatment and in whom a medial SPA is confirmed by CT. Conventionally, the abscess is drained via an external incision and an ethmoidectomy is performed at the same time. More recently, successful drainage of SPA's has been accomplished endoscopically via an intranasal approach with less morbidity (1,3,5).

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