Eosinophilic Granuloma With Arachnoid Cyst: A Case Report

Araknoid Kist ile Birlikte Eozinofilik Granülom: Vaka Sunumu

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Short running title : Eosinophilic granuloma with arachnoid cyst

Abstract: A 9 – year- old boy was admitted with a right parietal local swelling and tenderness. Plain X-rays and computerized tomography scan showed an osteolytic lesion at the right parietal cranium and a giant arachnoid cyst at the left temporoparietal region. The lesion was removed, histological examination revealed an eosinophilic granuloma. Immunohistochemical analysis of the specimen were positive for the S 100 protein and CD 1a cell surface molecule. No further surgery was performed for the arachnoid cyst, as each pathologic condition is considered apart. Also to our knowledge, neither co-existance of eosinophilic granuloma and arachnoid cyst nor eosiniphilic granulome of the skull secondary to aracnoid cyst have not been described previously.

Key Words: Eosinophilic granuloma, arachnoid cyst, co-existance

Özet: Sağ parietal bölgesinde lokal şişlik ve hassasiyet şikayeti ile başvuran 9 yaşındaki erkek çocuğun yapılan direkt grafi ve bilgisayarlı tomografi tetkikleri sonucunda, sağ parietal cranium bölgesinde osteolitik lezyon ve sol temporoparietal dev araknoid kist tespit edildi. Lezyon eksize edildi ve yapılan histolojik ve immün histokimyasal inceleme sonucu eosinofilik granülom olduğu tespit edildi. Her iki patolojik durumun birbirinden bağımsız olduğu ve tesadüfi birlikteliğin saptandığı düşünüldüğü için araknoid kist için cerrahi girişim uygulanmadı. Ayrıca bildiğimiz kadarı ile araknoid kist ve eosinofilik granülom birlikteliği veya araknoid kiste sekonder gelişen eosinofilik granülom durumu daha önce literatürde bildirilmemiştir.

Anahtar Kelimeler: Eosinofilik granülom, araknoid kist, birliktelik

INTRODUCTION

Langerhans' cell histiocytosis (LCH) is a rare disorder affecting predominantly children. This term encompasses a spectrum of clinical conditions, ranging from a single, sometimes self limited osteolytic bone lesion to a fulminant, disseminated process (12).

The etiology of this disorder is unknown (8). Historically, three clinical entities were described. The triad of calvarial defects, exophthalmus and diabetes insipidus is known as Hand-Schüller-Christian disease. A fulminant progressive proliferative disease of infancy is known as Letterer-Siwe disease. And a solitary osteolytic lesion of bone is known as eosinophilic granuloma (EG) (1,6,8). The predominant presenting manifestations are bone pains, swellings and lytic lesions on radiology (1,4). The diagnosis of LCH is confirmed by demonstration of langerhans cells in the lesions (2,4,6).

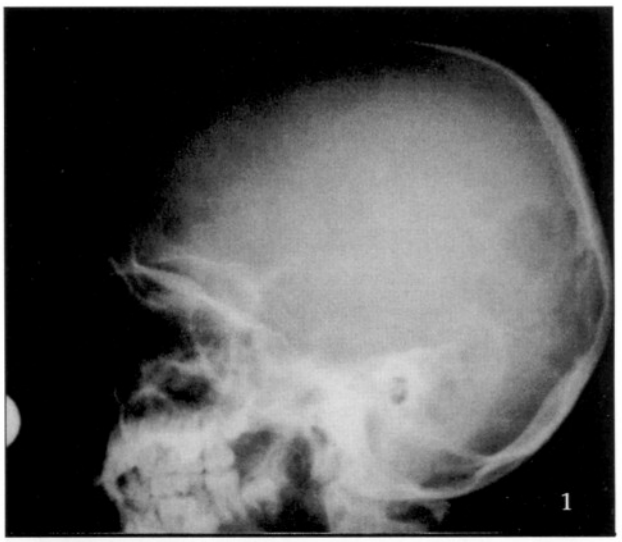
Arachnoid cysts (AC) are benign developmental cysts that occur in the

cerebrospinal axis in relation to the arachnoid membrane (5). The cysts mostly contain clear, colorless fluid resembling normal CSF (10). Computed tomography (CT) scan, magnetic resonance imaging, cisternograms and/or ventriculograms are the methods of evaluation. Despite numerous pathologic studies, the mechanism of formation of these cysts is not completely understood.

Most arachnoid cysts that become symptomatic do so in early childhood. The presentation varies with location of the cyst. Many authors recommend not treating arachnoid cysts that do not cause mass effect or symptoms, regardless of their size and location (5,7). Association between EG and AC is not described. To our knowledge this is the first case reported.

CASE REPORT

A 9- year- old boy visited our department complaining of right parietal local tenderness. No other symptoms or physical findings were found on the initial evaluation. His neurological examination found to be normal. The results of laboratory tests including serum blood chemistry profiles, sedimentation rate, leukocyte count, hematocrit concentration, serum and urine osmolarity were normal. The plain radiography of the skull showed an osteolytic lesion at the right parietal bone featuring sharp margins (figures 1, 2). CT scan showed an osteolytic lesion at the right parietal cranium (2*2,5cm) and an arachnoid cyst (4*6*7,5 cm) at the left temporoparietal part of the brain (Galassi type 3) (figures 3, 4). A clinical work up including a total body plain radiography showed no other bone lesion.



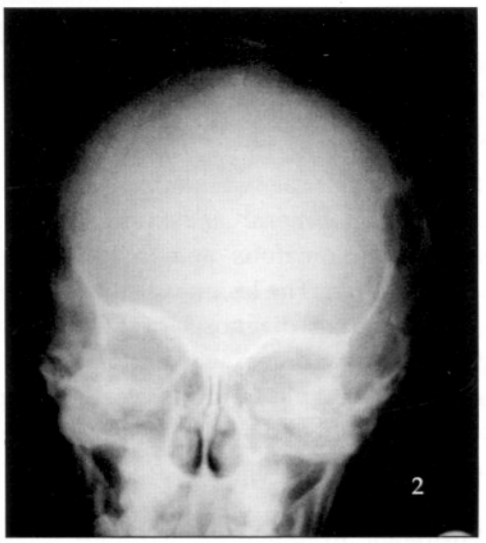
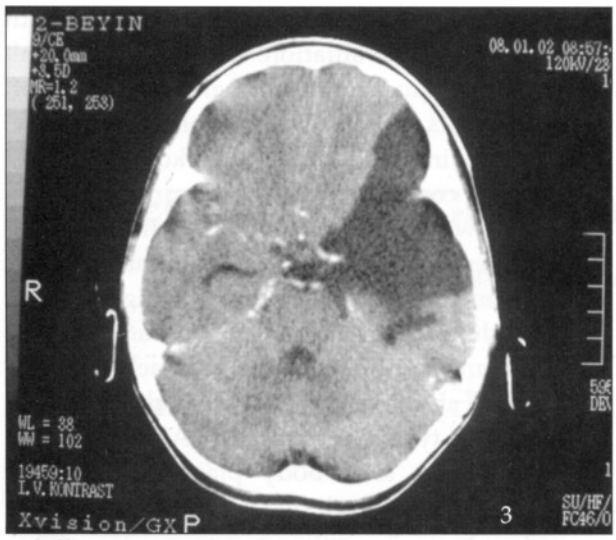


Figure 1, 2 : Plain x-rays, 1) lateral and 2) towne, reveal a right parietal, well circumscribed osteolitic lesion.



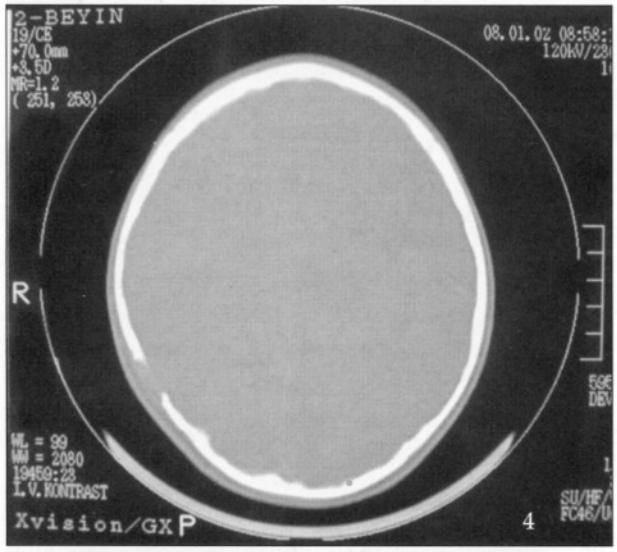
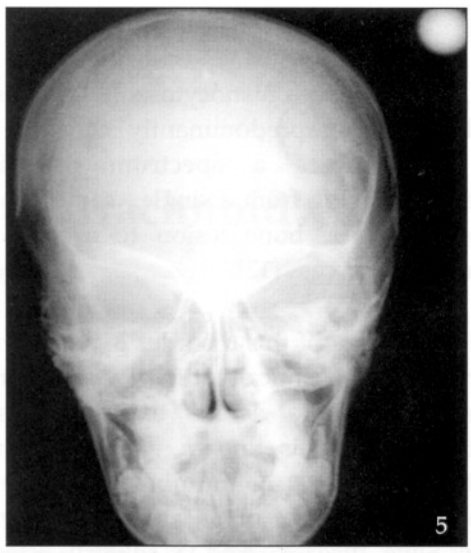


Figure 3, 4 : Computed tomography axial views, 3) large hypodense area at the left temporoparietal region without apparent mass-effect and enhancement, 4) a right parietal bone defect



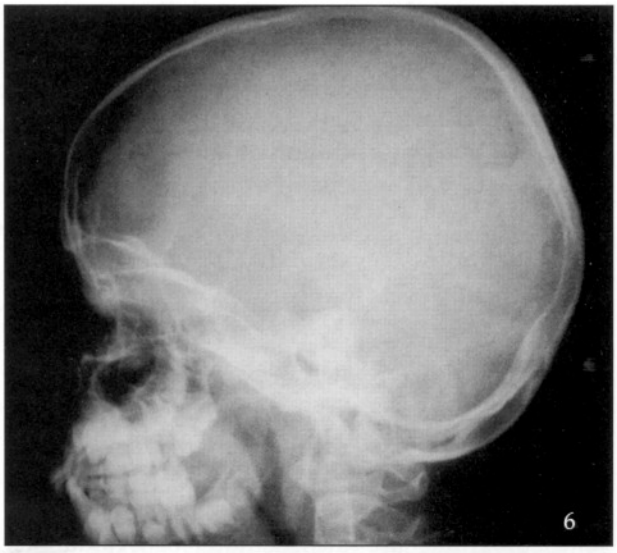


Figure 5, 6 : Post-operative plain x-rays 5) towne and 6) lateral, demonstrating the closure of the bone defect with autologous split-craniotomy flab.

Surgical removal was performed by supine position under general anesthesia. The lesion appeared granulomatous and yellow-brown on gross examination. The lesion was totally removed and intraoperative diagnosis of EG was made. Repair of the lytic defect was performed with an autologous bone split graft harvested from the adjacent calvarium and surgery was completed by primary skin closure (figures 5, 6).

Histologic examination of tumor showed a mixture of histiocyte-like cells with convoluted nuclear grooves and indentations, multinucleated giant cells and a mixed inflammatory infiltrate of eosinophils, neutrophils and lymphocytes.

Immunohistochemical examination were positive for the S 100 protein and cells are also reactive for CD 1a (a cell surface molecule). There were no tumor cells in the cut off margin of the skull.

DISCUSSION

LCH is a unifocal or multifocal disorder of bone or soft tissue or both. Eosinophilic granuloma

is the mildest form of LCH. The exact incidence of EG of the skull is not well known (3). The most common symptoms include local tenderness and an enlarged skull mass (%90) (3). A solitary EG is almost a localized process in older children and adults. As it may resolve spontaneously, observation can be an alternative treatment choice (11). The other treatment choices are craniectomy, curettage, intralesional injection of steroid, radiotherapy or combination (3,8,9).

We describe the case of a 9- year -old male patient with a right parietal local tenderness. Imaging by radiography and computed tomography showed an osteolytic lesion at the right parietal cranium and a arachnoid cyst at the left temporoparietal part of the brain. There was no history of trauma. Histologically the lesion was composed of multinucleated giant cells with S100 and CD1a positive langerhans cells and eosinophilis. So the two pathological entities found, thought to be incidental. Each pathological condition is considered apart while planning the treatment. EG is exised surgically. It is considered to be found co-incidentally and no surgical treatment was done for AC. The patient will be followed by clinical, radiological controls, psycomotor evaluating and treatment will be planned according to these parameters.

To our knowledge, nothing exists in the literature regarding EG of the skull secondary to AC. Moreover we found no description of this association using a medline search.

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