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Aneurysmal bone cyst of petrous bone associated with sixth cranial nerve palsy and fatal outcome

Altıncı kraniyal sinir felci oluşturan ve ölümle sonuçlanan petröz kemik yerleşimli anevrizmal kemik kisti

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Abstract

Aneurysmal bone cysts are benign but locally aggressive bone tumors often located in vertebrae, long tubular bones and flat bones. A small percentage of aneurysmal bone cysts arise from the skull base. We aimed to present a rare case of aneurysmal bone cyst located in the petrous portion of the temporal bone in a 13-year-old boy. It was unique because it was the only case with early aggressive recurrence and fatal outcome and also was the only patient in this entity who had sixth cranial nerve palsy as the only presenting symptom. Although rare, petrous apex aneurysmal bone cyst should be considered in the differential diagnosis of isolated or coexistent sixth nerve palsy or a petrous apex mass, especially in the presence of typical MRI findings.

Keywords: Aneurysmal bone cysts, petrous bone, sixth cranial nerve palsy.

Öz

Anevrizmal kemik kistleri sıklıkla vertebra, uzun tübüler ve yassı kemiklerde yerleşen, benign fakat lokal agresif tümörlerdir. Anevrizmal kemik kistlerinin küçük bir yüzdesi kafa tabanından kaynaklanır. Biz burada, 13 yaşında bir erkek olguda, temporal kemiğin petröz kısmında yerleşmiş nadir bir anevrizmal kemik kisti olgusunu sunmayı amaçladık. Erken agresif rekürrens göstermesi ve ölümle sonuçlanması nedeniyle eşsiz bir olguydu, ayrıca tek başvuru yakınması 6. kraniyal sinir felci olan bu gruptaki tek hasta idi. Petröz apeks yerleşimli anevrizmal kemik kisti, nadir olmasına rağmen, özellikle tipik MR görüntüleme bulgularının varlığında, izole ya da eşlikçi 6. kraniyel sinir felcinin ve petröz apeks kitlelerinin ayırıcı tanısında düşünülmelidir.

Anahtar Sözcükler: *Anevrizmal kemik kisti, petröz kemik, altıncı kraniyel sinir felci.*

Introduction

Aneurysmal bone cysts (ABC) are rapidly growing, benign, lytic, expansive lesions with well-known histology but obscure pathophysiology (1). They are more commonly located in vertebrae, long tubular bones and flat bones and are usually detected in the first two decades of life (2). ABC arising from the skull base is a rare condition with an incidence of 3-6% of all ABC cases and ABC located in the petrous portion of temporal bone is extremely rare (3). Magnetic resonance imaging (MRI) is the method of choice in the diagnosis, which usually shows a well-defined multiloculated lesion with fluid-fluid levels and heterogenous internal enhancement.

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Complete surgical removal is the best treatment option and is curative in most cases (5). Recurrence of temporal bone ABC is rare.

We present an additional case of ABC located in the petrous portion of the temporal bone in a 13-year-old boy. Our case report differs from the other reports in that 6th cranial nerve palsy was the only symptom at presentation; and that after complete removal of the tumor it recurred and caused the death of the patient.

Case Report

A 13-year-old boy presented to our hospital complaining of double vision that had been present for approximately one month. Neurologic examination showed diplopia and limited abduction of his left eye showing 6th cranial nerve palsy while he was otherwise normal. Cranial and temporal bone computed tomography (CT) showed an approximately 2 cm expansive, lytic and destructive

lesion located in the petrous apex of the left temporal bone. There were high and low density areas and punctate calcifications within the lesion (Fig-1a). MRI with TSE T2W axial, TSE T1W axial and postcontrast T1W axial images revealed a well-defined multiloculated cystic mass in the petrous portion of the temporal bone. There were multiple cysts with fluid-fluid levels (Figure-1b, arrow), surrounded by thin, T2 hypointense septae. Fluid-fluid levels and hyperintensity on T1W images within the tumor probably represented hemorrhage. The mass showed heterogenous internal enhancement on postcontrast scan. Depending on CT and MRI findings, this was diagnosed to be a benign mass and a suggestion of an ABC was made, although location was unusual. He underwent operation. The surgeons discovered a bony defect on the petrous apex covered by a membrane. On opening the membrane, they found a semisolid hemorrhagic mass with cavitation and a gross total excision was performed. On histopathologic examination, the diagnosis of ABC was made (Figure-1c), although an area, suspicious for osteoblastoma was detected. The pathologists were not sure whether it was a primary ABC or an ABC arising from a pre-existing lesion such as osteoblastoma.

Postoperative CT showed no residual tumor. As his complaints regressed postoperatively, he was discharged. In the fifth postoperative week, he came back with nausea and vomiting. MRI showed that the tumor, consisting of hemorrhagic cysts with fluid-fluid levels, had enlarged when compared to its size before the operation (Figure-1d). There was also peritumoral edema, mass effect on the 4th ventricle, brain stem and mesencephalon. He refused further treatment (reoperation or radiotherapy). In the ninth postoperative week, he was hospitalized due to progression of his symptoms. A repeat MRI showed that the tumor was even larger and more aggressive than it was in the previous MRI (Figure-1e). Increasing mass effect on the 4th ventricle and brain stem caused obstructive hydrocephalus. Reoperation was not accomplished due to accompanying neurologic, cardiovascular and pulmonary problems and the patient died two weeks after hospitalization.

Discussion

Aneurysmal bone cysts are benign, but locally aggressive bone tumors, consisting of thin-walled blood-filled cysts lined by connective tissue with giant cells and trabecular bone. They are typically found eccentrically in long bone metaphyses and posterior elements of vertebrae (3). ABC located in the petrous portion of temporal bone is extremely rare. The age of our patient is compatible with the fact that ABCs are usually encountered in the first two decades of life (2,3).

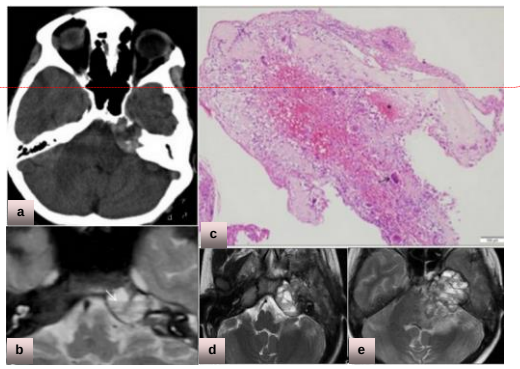

Figure-1. a. Cranial CT axial image shows an approximately 2 cm expansile, lytic and destructive lesion, with heterogenous density and punctate calcifications. **b.** MRI TSE T2W axial image reveals a well defined multiloculated cystic mass in the petrous portion of the temporal bone. **c.** The wall of the cyst lines blood-filled spaces (asterisk) and is composed of thick septa with multinucleated giant cells (arrow) (hematoxylin-eosin). **d.** Follow up MRI in the fifth postoperative week. TSE T2 axial image shows that the tumor enlarged when compared to its size before operation. **e.** MRI in the ninth postoperative week. TSE T2W axial image reveals that the tumor is larger and more aggressive. Increasing mass effect on the 4th ventricle and brain stem caused obstructive hydrocephalus.

Temporal bone ABCs most commonly present as swelling in the temporal region (6). Other symptoms such as hearing loss, 3rd-5th and 7th cranial nerve paralysis, headache, decreased vision, ptosis and even seizure, intracranial hemorrhage and recurrent meningitis have been reported. Our case differs from the other reports in that 6th cranial nerve palsy was the only symptom at presentation. This can be explained by the small size of the tumor and its location in the petrous apex. The portion of the abducent nerve passing through Dorello's canal at the petrous apex was probably affected by the tumor. Our patient did not have swelling in the temporal region, since the tumor was located in the petrous apex, deep in the temporal bone, as opposed to more common locations like superficial mastoid bone. Nevertheless, most of these symptoms, including pyramidal findings and obstructive hydrocephalus developed in our patient after recurrence, due to tumor growth toward the cerebellum, brain stem, temporal lobe and basal cisterna.

Abducent nerve palsy is associated with hypertension, diabetes, trauma, multiple sclerosis, neoplasm etc. while in one quarter of cases the cause remains undetermined (7). Neoplasms constitute a much less frequent cause of sixth nerve palsy than hypertension or diabetes. The differential diagnosis of a petrous apex lesion include pseudolesions (asymmetric marrow space development), infectious and inflammatory pathologies (cholesterol granuloma, cholesteatoma, mucocele),

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vascular lesions (arteriovenous malformation), dysplasias and neoplasms (8). As our case report indicates, petrous apex ABC, although rare, should be considered in the differential diagnosis of isolated or coexistent sixth nerve palsy or a petrous apex mass, especially in the presence of typical MRI findings.

Computed tomography shows that the lesion is intradiploic with expansion of the bone and thinning of bony tables. MRI is the method of choice in the diagnosis showing an expansive, well defined multiloculated cystic mass with hypointense peripheral rim and internal septations. Fluid-fluid levels are characteristic but not specific findings for ABCs, which can also be seen in other lesions like giant cell tumor, osteoblastoma, telangiectatic osteosarcoma, chondroblastoma, solitary bone cyst and fibrous dysplasia (9). The cysts have heterogenous signal due to blood products of varying ages. Heterogenous internal enhancement is noted on postcontrast scans. While the typical CT finding for ABC is preservation of bony tables, the tumor in our case was expansive and destructive; inner and outer tables were not discerned. MRI findings were typical for ABC, although internal enhancement was more intense than expected, especially on follow up MRI images when the tumor recurred. Intensely enhancing tissue that is of low signal intensity on T2W images surrounding cystic spaces have been described as fibrous elements, which are thought to be more abundant in the more cellular or solid variant of ABCs (4). The tumor in our patient can be considered to be a more solid variant of ABC, especially after recurrence.

Recurrence of ABC of the calvaria is rare, whereas recurrence of ABC of other bones may be as high as 50% (4,10). Recurrence has been related to younger age, larger tumor, presence of mitosis, incomplete surgical removal and dural involvement. The success of surgery is strongly associated with the rate of recurrence; while total excision is curative in most cases, simple curettage and subtotal excision may exhibit high recurrence rates varying from 20% to 50%. Our patient represents the only case of temporal bone ABC with early aggressive recurrence and fatal outcome, the reasons for which are not clear to us. Although gross total excision had been performed with no residual tumor on postoperative CT, one might retrospectively argue that a small residual tumor, undetectable by CT in the postoperative bed, could have caused recurrence. ABC is either primary or secondary to a preexisting lesion like giant cell tumor, osteoblastoma, chondroblastoma, nonossifying fibroma, angioma, fibrous dysplasia etc. Presence of a preexisting lesion has been reported to increase the likelihood of recurrence. No clear underlying lesion was found in our case, although an area suspicious for osteoblastoma was noticed.

Radiotherapy has not been used as a treatment option in the past because of risk of malignant transformation. It is now advocated for cases that are not suitable for surgery, and cases with recurrent or residual tumor (10). Radiotherapy was not used in our patient, which could have probably provided a better outcome. We suggest that when there is any suspicion that surgical removal is incomplete, radiotherapy should be added to treatment regime to prevent recurrence and infavorable outcome.

In conclusion, we present an ABC of the petrous bone, representing the 6th case in this location. It differs from the previous reports in that sixth cranial nerve palsy was the only symptom at presentation. Petrous apex ABC, although rare, should be considered in the differential diagnosis of isolated or coexistent sixth nerve palsy or a petrous apex mass, especially in the presence of typical MRI findings.

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