Lymphangioma of the Skull: A Case Report

Kafatasında Lenfanjiom: Olgu Sunumu

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Abstract: Lymphangioma is a relatively common tumor of soft tissue, but a rare primary lesion of bone. Very few cases of skull lymphangioma have been reported to date. We describe a patient with solitary lymphangioma of the skull that was treated by total excision and autogenous bone grafting. We also review the literature on this form of neoplasia.

Key Words: Lymphangioma, skull, tumor

Özet: Lenfanjiom, yumuşak dokunun kısmen sık, fakat kemiğin nadir primer lezyonudur. Günümüze kadar pek az kafatası lenfanjiomu olgusu bildirilmiştir. Yazarlar total olarak eksize edilip otojen kemik grefti ile tedavi edilen bir kafatası soliter lenfanjiomu bildirmişler; lenfanjiom ile ilgili literatürü incelemişlerdir.

Anahtar Kelimeler: Lenfanjiom, kafatası, tümör

INTRODUCTION

Lymphangioma is a relatively common soft tissue tumor that most often arises in the neck and axilla (5,7,9,10,15,16). Although this neoplasm can affect internal organs, central nervous system (CNS) involvement is uncommon, and bone involvement is even more unusual. Skull lymphangioma is particularly rare, but the literature does contain a few cases that have involved the cranium (6,12,13), skull base (15), scalp (10), and vertebrae (2,4,11). The case of skull lymphangioma documented in this report is significant because there have been only a few such cases described to date.

CASE REPORT

A 28-year-old female was admitted with the complaints of constant headache and a swelling near the midline of the frontal area of her skull, both of which had been present for 6 months. The swelling was approximately 1 cm in diameter when it was first noticed, but had enlarged in the past 2 months. There was no history of trauma or systemic illness.

Physical examination revealed a firm, painless, fixed swelling of 5 cm diameter on the frontal aspect of the cranium. The patient's neurological examination and laboratory values were normal.

Skull x-rays showed a radiolucent mass lesion in the frontal bone region, with radiating hyperdense zones. The radiographs also indicated that the mass had invaded both the inner and outer tables of the bones of the skull (Figure 1). Computerized tomography (CT) scanning revealed a 5x5 cm hyperdense extracranial mass in the frontal bone. CT also confirmed invasion of the inner and outer bone tables, and showed the radiating hyperdense sections that had been noted on the plain x-rays (Figure 2). Bone scintigraphy was normal, and we found no other organ involvement.

Surgical treatment involved a bifrontal craniotomy. The bone in the affected area was thin and bluish, and dissection revealed a cystic structure filled with straw-colored fluid. We were able to totally excise the tumor without opening or damaging the dura mater. The final step in the operation involved cranioplasty with a graft harvested from the patient's sixth rib.

Histopathologic examination of the removed tissue revealed numerous dilated vascular spaces lined by flattened atrophic endothelium, and these apparently new-formed channels were interspersed with bony trabeculae. There was no evidence of malignancy. The histopathologic diagnosis was lymphangioma (Figure 3). The patient's postoperative course was uneventful, and she required no further treatment.

DISCUSSION

Lymphangioma is a relatively common neoplasm of soft tissue that is characterized by slow progression and well-circumscribed borders, and typically contains new-formed lymph spaces and channels with cystic structures (5,7,9,10,15,16). The tumor has been documented in all organs, but 75% of cases affect the neck, 20% the axillary region, and the CNS is rarely involved (5,10,15).

Lymphangiomas generally present as multiple lesions, both in soft tissue and bone (2,7,8,15,17). They do not often manifest as a primary bone lesion, but Brickel and Broders reported the first such case in 1947 (1). Although lymphangiomas have been found in all types of osseous tissue, particularly long bones, skull involvement is extremely rare (2,5-8,12,13,15,17). Kopperman and Antoine reported the first primary lymphangioma of the skull in 1974 (12), a case that is one of only three descriptions of primary skull lymphangioma published to date (6,12,13).

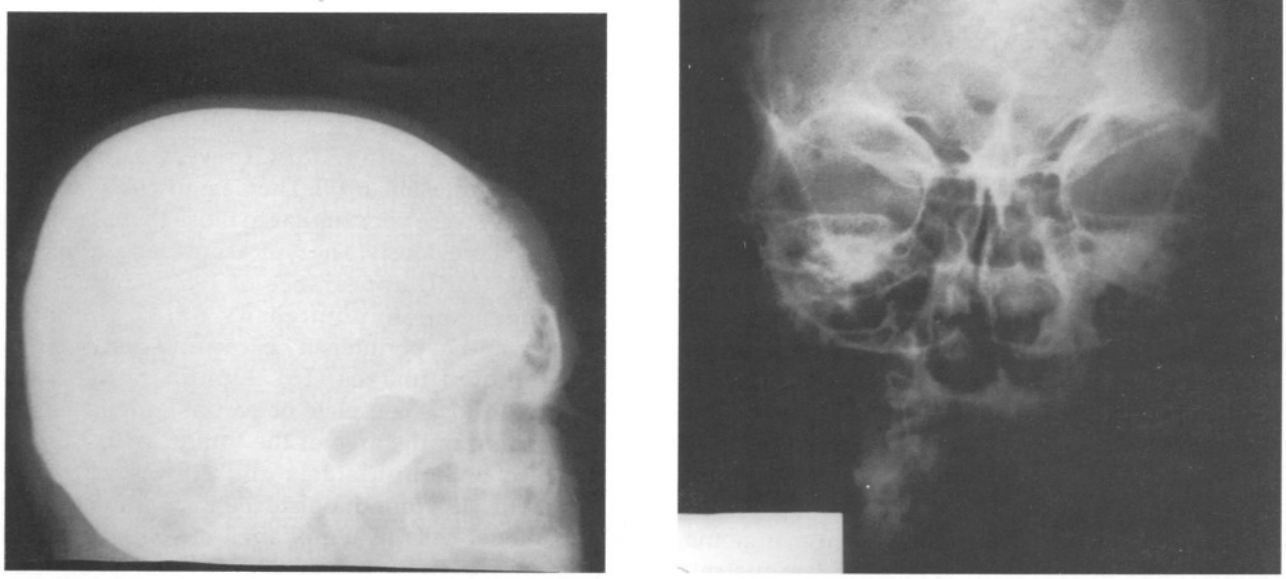

Figure 1: Lateral and anteroposterior radiographs of the skull reveal an expanding radiolucent mass in the frontal bone. The described radiating hyperdense areas are visible.

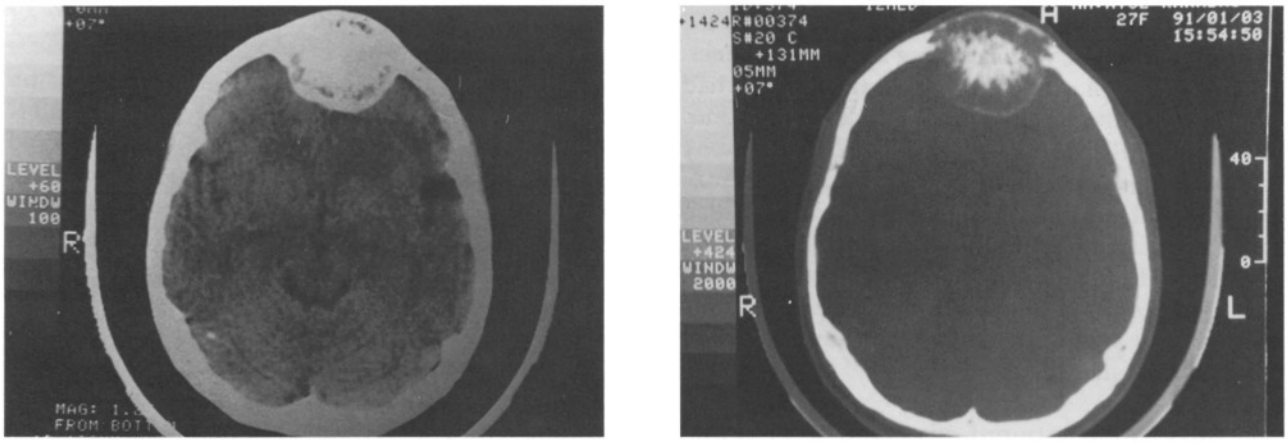

Figure 2: A CT scan shows an expanding hyperdense mass with radiating hyperdense areas located in the frontal bone of the cranium.

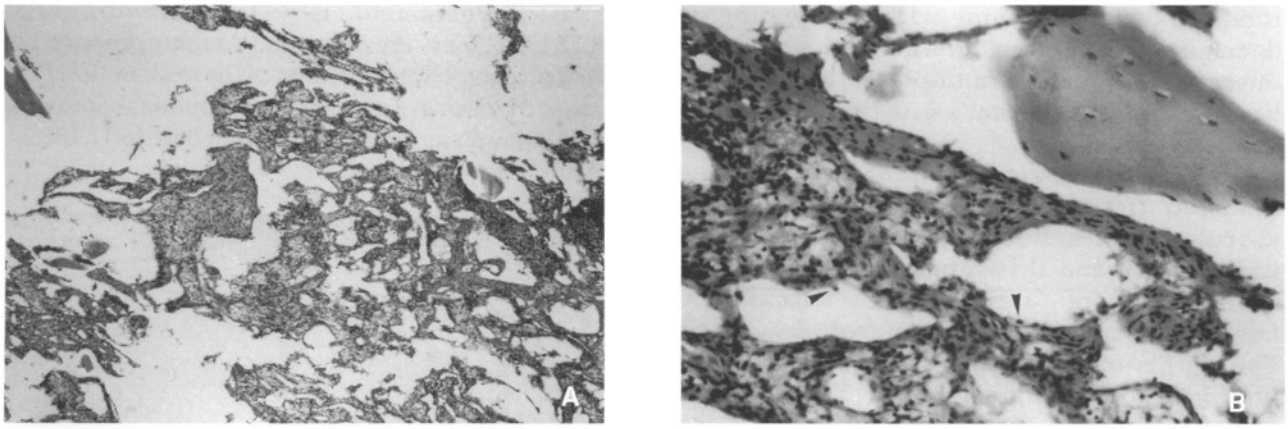

Figure 3: Photomicrographs of the excised specimen: A- Islands of dilated cystic blood vessels are seen between spongy bony trabeculae (H&E x40); B- Higher magnification of the same tissue shows communicating blood vessels adjacent to a remnant of spongy bony trabeculum. Note the atrophic endothelium overlying the dilated lymphatic channels (arrowhead). (H&E x200)

Regarding individuals affected and features of the disease, this neoplasm is not thought to be hereditary (14). It is most often seen in the first two decades of life (6), and arises with equal frequency in men and women (5,6,14). Our patient was a 28-year-old female. The most common complaint at presentation is the presence of a painful or painless mass (5,8,17). Spontaneous bleeding in the mass has also been reported in some cases (5). Our patient's only sign was a painless swelling that she had noticed 6 months prior to presentation, and that had enlarged in the 2 months prior to her initial assessment.

Concerning the diagnosis of lymphangioma, radiographs of the skull typically demonstrate involvement of one or both tables of the cranium, expanding bone or loss of bone mass, and a foamy or irregular-shaped lytic lesion with slightly sclerotic borders (6,9,12, 13,15,17,18). CT reveals a mixed lytic and sclerotic lesion (6,13). The CT and x-ray findings in our case were very similar to those that have been described previously. The typical magnetic resonance imaging (MRI) appearance of lymphangioma is heterogeneous hypointensity on T1-weighted images, and hyperintensity exceeding that of fat on T2-weighted images (16). Lymphangiography typically shows complete or partial lymph channel obstruction in the area of the tumor, with varying degrees of lymph stasis and collateral vessel formation (18). Immunohistochemical studies of the endothelial cells in lymphangioma lesions reveal immunoreactivity to Factor VIII-related antigen (FVIII-Rag), cluster determinant (CD31, CD34), and anti-Ulex Europaeus Lectin I (UEA-1) (14).

The three different histological types of lymphangioma are cystic lymphangioma, capillary or simple lymphangioma, and cavernous lymphangioma (4,5,8). The cystic form is a multilocular cystic mass known as "cystic hygroma" that occurs most often in the neck or axillae of children. No bone involvement has been documented with this type. The lesion in capillary or simple lymphangioma is characterized by capillary-sized lymphatic channels. This type usually arises in the skin, but bone involvement has been observed. The mass in cavernous lymphangioma contains larger lymphatic channels, and is known to arise in bone, soft tissue, and viscera. The growth consists of endothelial cells and connective tissue, and thus is considered to originate from mesodermal remnants. It is considered a benign congenital malformation rather than a tumor, and is characterized by osteolytic and cystic components. When blood cells are present, the cystic component can be confused with aneurysmal bone cysts and hemangiomas (3-6,13). When this form occurs in bone, there are often abnormalities of the haversian canals and of the lymph canals in the periosteum (1). The differential diagnosis for lymphangioma of the skull includes aneurysmal bone cyst, eosinophilic granuloma, osteitis fibrosa cystica, hystiocytosis X, hemangioma, solitary plasmocytoma, osteoma, meningioma, and metastasis (3,4,6,11,13,15,17).

Surgery is the definitive treatment for lymphangioma of the skull, and total excision is always necessary. Studies have shown that curettage alone is not successful (5,6). Bone grafting has generally yielded poor results because the grafted bone tends to become involved with the angiomatous process, and new bone formation is compromised (6); however, grafting is required when tumor removal results a large bone defect (2). Recurrence is almost always associated with subtotal tumor removal, and usually occurs within a year of the initial surgery (10,16). Radiotherapy is not effective when applied alone, but may be used as adjunctive treatment in cases of subtotal excision or tumor recurrence (2,4,6). Currently, chemotherapy is not used to treat this disease (6). Although sclerosing agents have been applied to these tumors in the past, this method is no longer considered a good alternative to surgery (5).

In conclusion, the case of skull lymphangioma described in this report is one of a small handful in the literature. Although this lesion is extremely rare, lymphangioma should be considered in the differential diagnosis of any solitary expanding or lytic lesion of the cranium.

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