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Solitary epithelioid hemangioendothelioma of the metacarpal

Metakarpta soliter epiteloid hemanjiyoendoteliyom

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Hemangioendotheliomas are vascular tumors which have an intermediate clinical behavior between hemangiomas and angiosarcomas. The epithelioid subtype of hemangioendothelioma, which is rarely seen in bone, has the potential to metastasize and may be confused radiologically with benign or malignant lesions. A metacarpal origin of this tumor is extremely rare. In this article, we present a 42-year-old female case with an hemangioendothelioma in the second metacarpal which was initially managed as a benign lesion before the patient was referred to our center. The potential malignant nature of epithelioid hemangioendothelioma has been ill-defined. We recommend excision with a wide surgical margin. Amputation may be necessary to perform excision in bones such as the metacarpals or phalanges.

Key words: Bone tumor; epithelioid hemangioendothelioma; metacarpal tumor.

Hemanjiyoendoteliyomlar, hemanjiyomlarla anjiyosarkomların arasında klinik bir davranışa sahip olan vasküler tümörlerdir. Nadiren kemikte görülen hemanjiyoendoteliyomun epiteloid alt tipinin metastaz yapma potansiyeli vardır ve radyolojik olarak benign ve malign lezyonlarla karışabilir. Bu tümörün metakarp kökenli olması son derece nadirdir. Bu yazıda ikinci metakarpında hemanjiyoendoteliyom saptanan ve kliniğimize sevk edilmeden önce başlangıçta benign bir lezyon olarak tedavi edilen 42 yaşında kadın bir olgu sunuldu. Epiteloid hemanjiyoendoteliyomun muhtemel malign özelliği yetersiz tanımlanmıştır. Biz geniş sınırlarla eksizyon önermekteyiz. Bunu metakarp ve falanks gibi kemiklerde uygulamak için amputasyon gerekli olabilmektedir.

Anahtar sözcükler: Kemik tümörü; epiteloid hemajiyomaendoteliyom; metakarp tümörü.

Hemangioendothelioma belong to a category of vascular tumors that are less malignant than conventional angiosarcomas.[1] A subtype known as epithelioid hemangioendothelioma was proposed by Weiss and Enzinger in 1982, and other subtypes include kaposiform, hobnail (Dabska or retiform) and polymorphous.[1,2] Of these, the epithelioid subtype is the most aggressive.[1]

Epithelioid hemangioendothelioma occurs mostly in adults and has been encountered as a primary tumor in a wide distribution of body tissues.[2-13] In bone tissue it occurs most commonly in the lower extremities.[7] In the bones of the hand it is very rare, and to our knowledge it has been described in the metacarpal in only one patient previously, as part of multicentric disease.[7,14-17] We describe here a female patient with a solitary epithelioid hemangioendothelioma of the second metacarpal, a condition that to our knowledge has not been previously reported.

CASE REPORT

A 42-year-old woman presented at our clinic one week after undergoing surgery at another center for a mass on the dorsal surface of her right hand, just proximal to the index finger. She explained that she had experienced pain and swelling for two months before that operation. The procedure was simple curettage, and surgical specimens were collected. Plain radiographs showed a lytic lesion in the second metacarpal of the patient's right hand (Figure 1).

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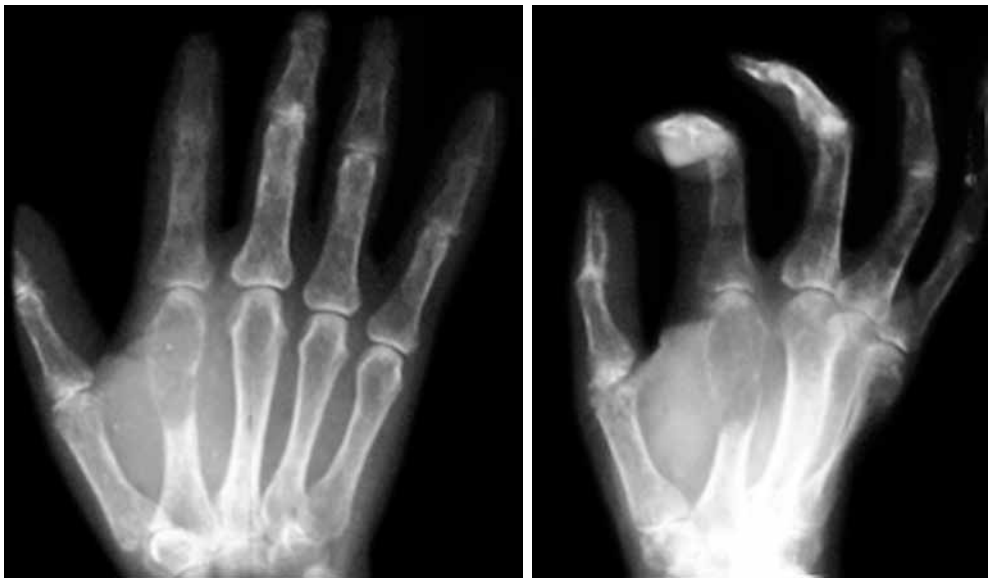

Figure 1. This anteroposterior and oblique plain radiograph of the patient's right hand shows a lytic lesion with cortical expansion and thinning in the second metacarpal.

On computed tomography (CT), the lesion appeared to be lytic with expansion and destruction of the cortex (Figure 2). When histopathologic analyses revealed the mass to be epithelioid hemangioendothelioma, the patient was referred to our hospital.

At our hospital, the patient underwent magnetic resonance imaging (MRI) with contrast (Figures 3a, b), and this revealed an enhancing expansile lesion (Figure 3a). Edema was also apparent in the flexor and extensor tendons. Ultrasonography (US) of the regional lymph nodes and CT of the lung showed no metastatic disease. Plain radiographs of the pelvis, vertebral column and extremities showed no lesions. Bone scintigraphy could not be performed due to technical problems.

The second finger and metacarpal were amputated, sparing the metacarpal base (Figures 4a, 5). Histopathologic examination revealed infiltration of the cortex with mildly atypical cells with cytoplasmic vacuoles. In many fields the stroma was myxoid. The tumor's rich vascular structure was also apparent (Figure 4b).

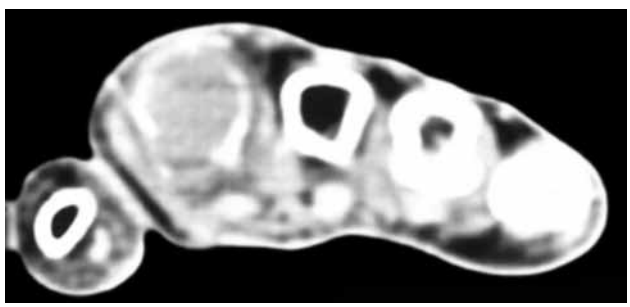

Figure 2. Computed tomography showed cortical thinning and expansion, with cortical destruction on the palmar and dorso-ulnar aspects of the second metacarpal.

On follow-up, the patient underwent MRI of the hand and CT of the lung every three months in the first year, and every six months thereafter. Plain radiographs in the 18th month of follow-up showed no evidence of tumor in the remaining metacarpal base (Figure 5). The patient is now in the 30th month of follow-up and has had no further complaints.

DISCUSSION

In the series of patients with epithelioid hemangioendothelioma of bone reported by Tsuneyoshi et al.,[17] one patient had this tumor in a metacarpal as well as in the ipsilateral scapula. The authors of that study suggested that multicentric disease has a better prognosis than solitary tumors.[17] However, Kleer et al.[7] in their later series of similar patients argued that instead of multicentricity per se, the important factor in multicentric disease may be involvement of the viscera. The patient we describe here provides an example of a solitary epithelioid hemangioendothelioma in the metacarpal, a condition which appears not to have been described before.

Clinically, patients with epithelioid hemangioendothelioma in bone may present with pain and swelling and less commonly with pathologic fracture.[6,15-17] In the series by Tsuneyoshi et al.,[17] multicentric tumors were common, occurring in

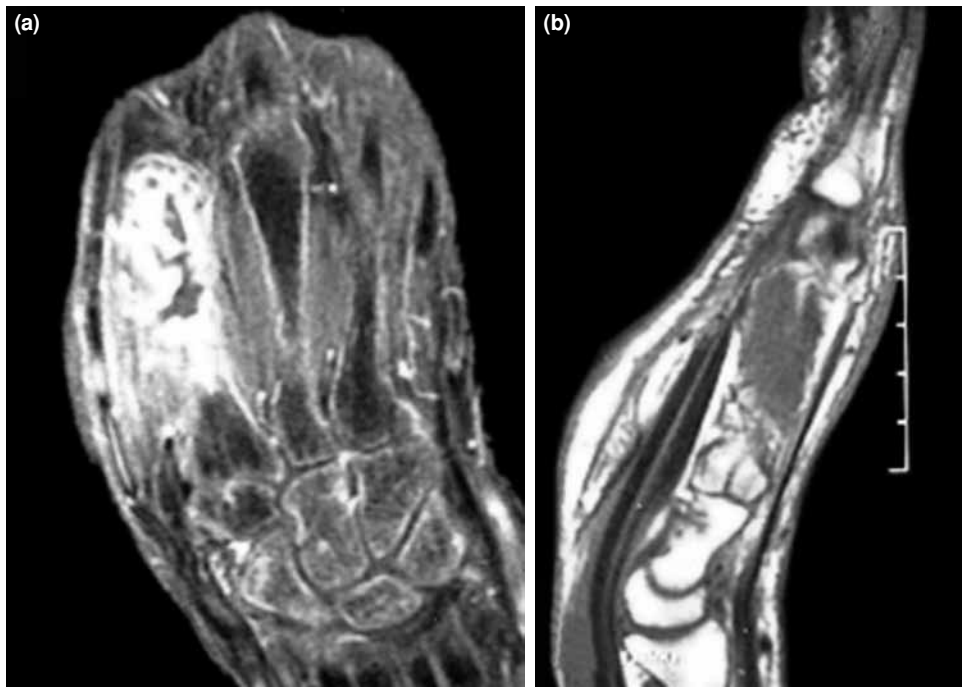

Figure 3. (a) This post-contrast fast spin-echo T_1 image with fat suppression shows involvement of the entire head of the metacarpal, with cystic/necrotic components inside the lesion. **(b)** This T_1-weighted spin-echo sagittal image shows a hypointense lesion approximately 4 cm in length, with soft tissue involvement on the palmar side of the second metacarpal, with the proximal part of the bone remaining intact.

nine of 14 patients, and in the study by Kleer et al.[7] approximately half of the patients had multiple lesions. The major differences between this tumor's occurrence in the upper versus lower extremities is that it is rarer in the upper extremities, and that its clinical presentation can differ according to the anatomical structures involved. For example, in the lower extremity pathological fractures may be more prone to occur.

On plain radiographs, epithelioid hemangioendothelioma in bone generally has a lytic appearance, with sclerotic changes, cortical destruction and expansion, and unclear tumor margins also being common.[7,16-18] In the series reported by Kleer et al.,[7] periosteal reaction was rare and soft tissue extension occurred in approximately one-fourth of the patients. In our patient, extension of the tumor into soft tissue was apparent on MRIs (Figures 3a, b).

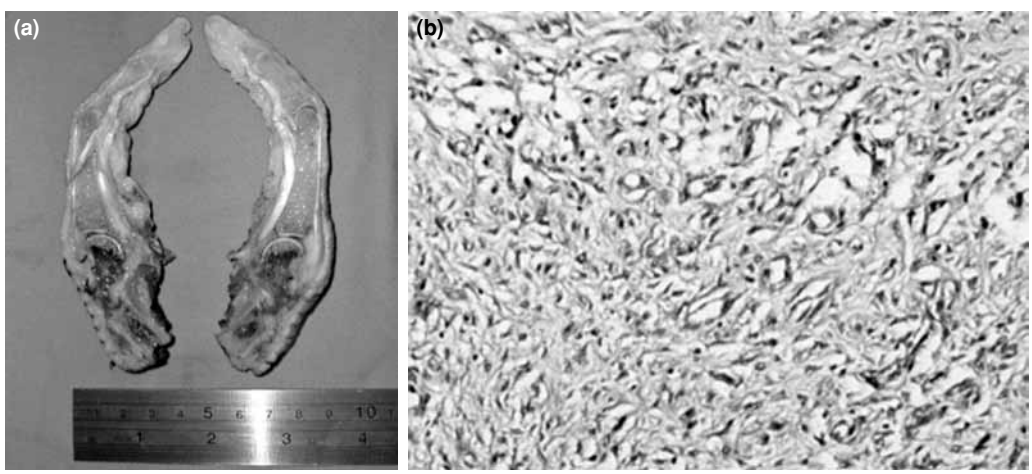

Figure 4. (a) Macroscopic appearance of the amputated finger and metacarpal. **(b)** The tumor cells formed anastomosing channels, had large nuclei and acidophilic cytoplasm, arranged in myxoid stroma. Note occasional cytoplasmic vacuoles (H-E x 400).

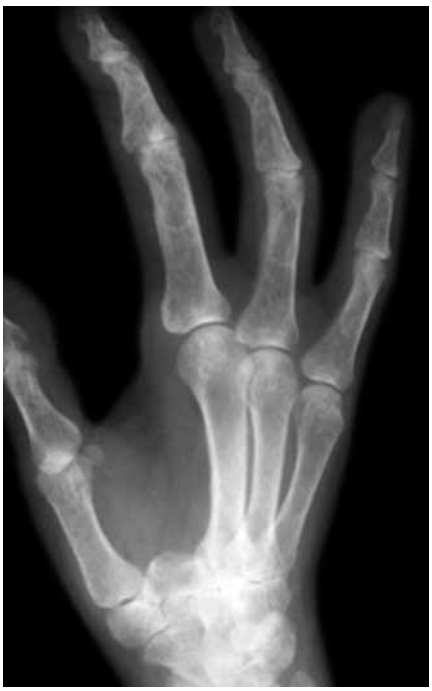

Figure 5. This oblique radiograph taken 18 months after the amputation shows the remaining metacarpal base with no evidence of disease.

The differential diagnosis of epithelioid hemangioendothelioma can be difficult, because tumors can show gradations in histologic features. In our patient, the tumor's presence in the hand suggested the possibility of benign lesions such as enchondroma, tuberculosis, giant cell tumor, aneurysmal bone cyst, chondromyxoid fibroma and epidermoid inclusion cyst, macrodactyly.[19-21] Malignant possibilities included metastasis, chondrosarcoma, Ewing's sarcoma and osteosarcoma.[22-24]

Histopathologically, epithelioid hemangioendothelioma has an epithelioid appearance and less pleomorphism and mitotic activity compared to metastatic or disseminated carcinomas.[2] Weiss and Enzinger[2] reported that stains for mucin fail to confirm mucin production in this tumor, and that vascular differentiation is apparent via reticulin staining or electron microscopy. Other histologic criteria that have been used include hyalinized or myxoid stroma and the presence of antigens such as cytokeratins.[7] The presence of Weibel-Palade bodies and positivity for endothelial markers such as CD31, CD34, factor VIII-related antigen and Ulex europaeus binding suggest that the tumor originates from vascular endothelium.[16,17]

The treatment used for the patient presented here differs from that in previous reports of epithelioid hemangioendothelioma in hand bones. For their patient who had a metacarpal tumor, Tsuneyoshi et al.[17] used curettage. For a patient with a solitary tumor of the phalanx, Kitagawa et al.[16] used curettage and an autogenous bone graft from the iliac crest. Bruegel et al.[14] described a patient who had tumors in the middle, ring and little fingers of the right hand. The tumor in the little finger was the most extensive, and was treated with amputation of the finger; in the other two fingers, the treatment was curettage and grafting. For our patient, due to the soft tissue involvement seen on MRI (Figures 3a, b) and the possibility that tumor cells had been spread during the previous surgery, we preferred amputation as a means of achieving a wide surgical margin.

With an average follow-up period of 4.3 years, Kleer et al.[7] reported that 20% of their patients with epithelioid hemangioendothelioma of bone died from the disease. This shows the need for thorough examination and follow-up in patients with this diagnosis. When the tumor is identified, bone scintigraphy should be used to determine whether the patient has multiple lesions. Regional lymph nodes and lungs should be investigated for metastases with US or CT, and suspected lesions should be biopsied. For treatment we recommend surgery with wide margins if possible and radiation therapy in addition to surgery if wide margins are not possible. Although chemotherapy has been used in patients with involvement of multiple bones or parenchymal organs, its role is still uncertain.[7] Because of the possibility of local recurrence and metastases, we recommend long-term follow-up for all patients with this tumor.[1,7,14]

Declaration of conflicting interests

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