Primary Malignant Fibrous Histiocytoma of Bone in Distal Radius

Radius Alt Uç Yerleşimli Kemiğin Primer Malign Fibröz Histiyositomu

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Primary Malignant Fibrous Histiocytoma of Bone in Distal Radius A case of primary malignant fibrous histiocytoma of bone (MFH-B) located in the distal region of the radius is presented. It is very rare for primary MFH-B to be located in upper extremity particularly in wrist region. The mainstay of treatment for MFH-B is wide resection combined with chemotherapy. This report demonstrates a case of MFH-B of distal radius, treated successfully with surgery combined with chemotherapy.

Key Words: ***primary, radius, malignant fibrous histiocytoma, bone***

Kemiğin primer malin fibröz histiyositomu (KPMFH) üst ekstremite özellikle de bilek çevresinde çok çok nadir yerleşim gösterir. Kemiğin primer malin fibröz histiyositomunun tedavisinin temelini lezyonun geniş olarak rezeksiyonu ile kemoterapi combine edilmesi oluşturur. Bu yazıda, radius alt kesiminde KPMFH tespit edilen ve ceraahi ile kemoterapi combine edilerek başarı ile tedavi edilen bir hasta sunuldu. Lezyon ekstraartiküler olarak küretе edildi. Yüz on dört aylık takip ve kontrolü sonrasında hastada hiçbir lokal nüks veya metastaza ait klinik ve radyolojik bulgu saptanmadı.

Anahtar Kelimeler: ***primer, radius, malin fibröz histiyositom, kemik***

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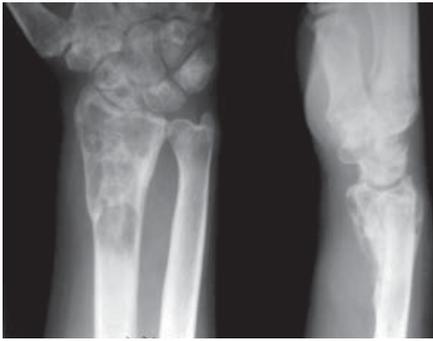E-mail address : drbulentozkurt@yahoo.com

Malignant fibrous histiocytoma of bone was first described as a distinct bone tumor in 1972 by Norman and Feldman as malignant histiocytoma (malignant fibroxanthoma) (1). Although many cases of malignant fibrous histiocytoma in somatic soft tissue have been reported, the description of its bony counterpart are relatively rare, representing 1 to 8% of all primary malignant bone tumors (2,3). Although this tumor may occur at any site, the most common localization site is knee (3). Reports of bony localizations involving the distal forearm and hand, in the literature, are rare (3-5). We present a case with primary MFH-B of distal radius.

Case Report

A 47-year-old man presented with pain in dorsal region of his right wrist. Duration of the symptoms was six months with increasing swelling in the past three months and wrist stiffness in the past 45 days. There was no history of previous trauma, malignancy or recent illness. Clinical examination revealed swelling on distal radius with localized tenderness on the dorsal side. There was swelling on the dorsal and volar region of the distal radius. Active and passive pronation and supination of the forearm and total range of motion of the wrist were limited because of pain. No palpable lymph node was determined. Laboratory tests including complete blood cell count, biochemical values, C-reac-

Figure 1. Preoperative X-Ray of the patient

Figure 2. Preoperative MRI of the patient

Figure 3. Preoperative coronal MRI of the patient
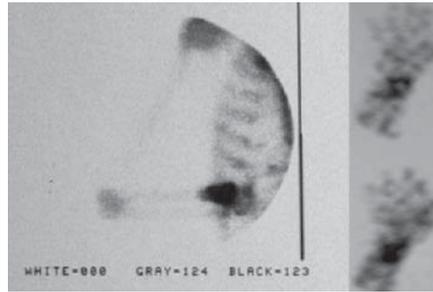
Figure 4. Preoperative scintigraphy of the patient

tive protein, erythrocyte sedimentation rate were normal. A radiograph of the right wrist showed an expanding osteolytic lesion of the distal radius with cortical destruction, extending up to the articular surfaces of both radiocarpal and distal radioulnar joints, with dimensions of $30 \times 35 \times 65$ mm (Figure 1). There was no evidence of pathological fracture. Magnetic resonance imaging of the distal radius showed volar and radial cortical destruction with direct spread into the adja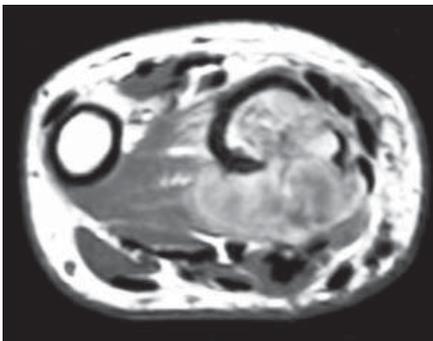cent so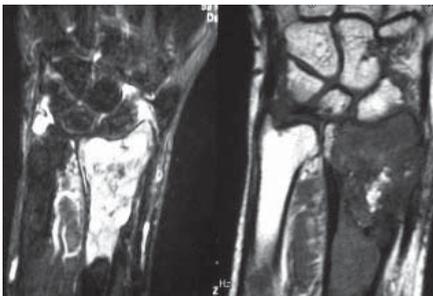ft tissue (M. pronator quadratus). There was bone marrow edema within the adjacent diaphyseal region. Cortical destruction of the radioulnar and radiocarpal articular surfaces was evident (Figure 2, 3).

Whole-body scintigraphy and radiographic bone survey disclosed no additional lesions (Figure 4). The radiograph and computerized tomography scans of the chest and abdomen were normal.

Wide resection of the tumor was performed on January 1999. The radius was cut one inch proximal to the most proximal tumor margin. Wrist extensors, wrist flexors and m. brachioradialis were excised at the same level as the radius. Pronator quadratus, triangular fibrocartilage and distal radioulnar joint capsule were excised. The remaining tendons, distal ulna and carpal bones were preserved. Non-vascularized free fibular graft was obtained from the contra-lateral leg. The fibula was fixed to the radius shaft and the middle ray metacarpal shaft with 14 holes 3.5-mm DCP plate. Cancellous autograft, obtained from iliac crest, was inserted between the carpal bones and fibular head, proximal fibula and radius shaft junctions after excision of the articular cartilages of the fibular head, carpal bones. Midcarpal arthrodesis was performed (Figure 5).

Figure 5. Postoperative posteroanterior and lateral x-ray of the patient after non-vascularized fibular grafting

Histological examination showed a high-grade, pleomorphic sarcoma. The tumor cells being arranged in a storiform pattern and producing varying amounts of collagen. Numerous giant cells were distributed throughout the specimens. The appearances were typical of a MFH-B (Figure 6).

Adjuvant chemotherapy was started three weeks after surgery. Chemotherapy protocol consisted of high-dose methotrexate (8-12 g/m2), adriamycin (60-75 mg/m2,8-h infusion), cisplatinum (120 mg/m2) and citrovarum factor rescue. Chemotherapy was well tolerated and never life-threatening. No cardiac or hepatic toxicity was
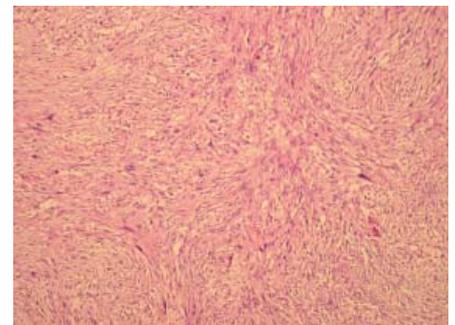

Figure 6. Histopathological features of the patient

Figure 7. Postoperative x-ray of the patient after union

seen. Mucositis requiring mouth care was encountered.

After a sixth year follow-up without any complication, on January 2006, the plate was removed (Figure 7). The patient had satisfactory outcome. He had no pain and complete union at both wrist and proximal fibula-radius junctions was obtained. One month after removal of the plate, a fracture at the distal region of the graft occurred owing to a low energy trauma (Figure 8). After a five months period of conservative treatment with a short arm cast, union could not be obtained. On July 2006, the patient underwent surgery and treated with vascularized fibula and internal fixation (Figure 9). The patient is still under follow-up at 114th month, with no evidence of recurrence or metastasis.

Discussion

Malignant fibrous histiocytoma is an aggressive pleomorphic tumor, that arises most commonly in the soft tissues (6). but bone may be

Figure 8. X-Ray of the patient after fracture followed by minor trauma

the primary site of malignant fibrous histiocytoma, rather than involvement by direct extension of a soft tissue tumor or metastatic spread (1,2).

MFH-B represents 1 to 8% of all primary malignant tumors (3). The majority of the lesions are primary, but 20 to 28% of MFH-B occur secondarily in pre-existing benign bone abnormalities such as Paget's disease, enchondroma, fibrous dysplasia, bone infarcts or cysts, giant cell tumor, aseptic

Figure 9. X-Ray of the patient after revision with vascularized fibula

necrosis, prolonged intake of corticosteroids, previously irradiated either already diseased or normal bone and osteomyelitis (3,5,7). MFH-B has a male predominance ranging from 55-75% of the cases reported in the literature (6,7). Although MFH-B may arise at almost any age, the majority of tumors occur after the fourth decade (5). Our case as an adult man, was a typical example concerning the epidemiology of MFH without any preexisting bone abnormality.

Slow-growing swelling together with local pain is the most common clinical presentation as in our patient (7). Pathological fracture may be the initial symptom (1,3,7).

Although MFH-B may arise in any bone, the metaphyseal parts of the long bones of the appendicular skeleton are the most common localizations (5,7). The majority of the tumors arise around the knee in the lower metaphysis of the femur and upper metaphysis of the tibia (3,7). Reports about the involvement of upper extremity are rare but primary MFH-B has been reported in the proximal humerus, mid-humerus, olecranon, ulna, metacarpal bones, distal phalanx and in radius only one case, to our knowledge, till now on (3,5).

In radiographic assessment, MFH-B typically presents a solitary lytic or permeative lesion, located centrally or eccentric within the metaphyseal region of a long bone with eventual spread into the adjacent epiphyseal and diaphyseal regions. The edges are ill-defined, cortical expansion and destruction with little or no reactive sclerosis is almost always present. Adjacent soft tissue mass may be apparent, but a periosteal new bone

formation is uncommon (5,7). The differential radiological diagnosis includes metastasis from an occult primary tumor, osteolytic osteogenic sarcoma, fibrosarcoma of bone, primary malignant lymphoma, myeloma, and malignant giant cell tumor.

As the clinical, radiological and laboratory data are not pathognomonic for MFH-B, any suspected destructive, lytic osseous lesions should be evaluated histologically by needle or carefully planned open biopsy for diagnosis. Detailed physical examination, CT scan or magnetic resonance imaging of the lesion, radionuclide scanning with technetium99 and CT scan of the chest are necessary for accurate diagnosis and classification of the lesion into its surgical stage.

Histologically, tumor extension through the bony cortex is often present with direct involvement to adjacent soft tissues. The intraosseous tumor spread is characterized by medullary bone marrow infiltration and replacement without reactive ossification or marrow response (7). The spindle fibroblastic-fibrohistiocytic cells, plump cells with histiocytic morphology, and giant cells are always present. Typically, it shows fibrogenic differentiation, often in a storiform (cartwheel) pattern, and other areas of cells have a histiocytic quality and are accompanied by anaplastic giant cells. Prominent collagen production is present between the spindle cells, which are oriented in the typical whorled or storiform pattern. Chronic inflammatory cells are present in majority of the lesions (5).

Biologically, MFH-B is a fully malignant tumor, which not only recurs locally, but also metastasizes via both blood stream and lymphatic channels to distant sites, most commonly to the lungs, or less commonly to the lymph nodes, soft tissues such as liver, heart, kidney, adrenals, pancreas, skin or other bones (1,6). Prognosis depends on local recurrence and metastases. Several authors emphasize that preoperative and/or adjuvant chemotherapy reduces the ever present threat of microscopic residual local or distant (metastatic) disease (4,6).

The mainstay of treatment for MFH-B is aggressive surgical management combined with chemotherapy (3,5,7). Although adjuvant chemotherapy has been shown to improve the survival rate and reduce the incidence of metastasis (2,3,5,6). chemotherapy is not as successful as in patients who underwent inadequate surgical margins (3,5,8).

Numerous alternative procedures, such as autogenous corticocancellous bone grafting, allograft replacement, autogenous vascularized or non-vascularized fibular grafting, custom made prosthesis can be used for the reconstruction after resection of the distal radius. We preferred autogenous fibular grafting and wrist arthrodesis in order to prevent the probable complications of the custom made prosthesis and also wrist instability and degenerative arthritis.

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