CASE SERIES

PRIMARY ANGIOSARCOMAS OF THE BONE: AN EVALUATION OF 4 CASES

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Angiosarcoma is a rare mesenchymal neoplasm that may arise from vascular or lymphatic tissue. Angiosarcoma of bone is a rare high-grade malignant vascular tumour, representing less than 1% of all angiosarcomas. The most common locations of unifocal tumour are the long and short tubular bones, followed by the pelvis, and trunk. The literature regarding treatment and outcome of patients with this tumour is limited. We performed a retrospective study to analyse treatment and survival of four patients with angiosarcoma of bone.

Keywords: Angiosarcoma of bone; Angiosarcoma; Bone tumour

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INTRODUCTION

Bone angiosarcoma is not a common tumour. It constitutes less than 1% of all primary bone sarcomas. Prognosis is generally poor.[1] Skeletal angiosarcoma usually affects a wide range of ages. It affects especially young adults and elderly individuals.[2] Angiosarcomas often tend to affect the long tubular bones of the extremities. The lower extremity bones, especially the femur and the tibia are most involved frequently, followed by the pelvis.[3,4] The treatment and results of patients with bone angiosarcoma are limited in the literature because of the bone angiosarcoma's rarity.[1] Therefore, the objective of this study was to present clinical features, surgical procedures, complications, and outcomes after surgery of the bone angiosarcoma.

CASE REPORTS

Informed consent was obtained from each patient to publish details of their cases.

Case 1:

A 54-year-old man presented with right hip pain for 6 months. There was not an history of any trauma. Weight-bearing activities was painful for patient. Physical examination revealed tenderness but no swelling. Laboratory tests, including erythrocyte sedimantation rate (ESR), C-reactive protein (CRP), and complete blood count were normal. Plain radiography showed intramedullary heterogen radio dense areas and cortical erosions in the proximal femur. Magnetic resonance imaging (MRI) of the femur demonstrated as lightly lobulated subcortical exantric tumoural lesion of 4×3×2 cm dimensions with intact contours and sclerotic borders in the right femoral proximal intertrochanteric region, which has keeping prominent contrast substance and destructing the lateral cortex amounting 5 mm width with periost reaction. Tru-cut biopsy was performed and histopathological result was angiosarcoma. There was no lung metastasis in the CT (Figure-1A, B). Then we decided to operate the patient with wide resection and modular prothesis reconstruction. Spinal anaesthesia was performed, and the patient was placed in a lateral decubite position and a long posterolateral incision made, beginning in the proximal-thigh, ending in the mid-thigh. Subcutaneous tissue was passed through. The fascia was passed through. Vastus lateralis muscle was detached from its origin from the trochanter majoris. Trochanter major was osteotomized by cool chisel. After that, distal femoral osteotomy was performed according to preoperative planning.

Tumoural lesion was excised with wide margins. Thereafter, a definitive modular prothesis was assembled. The 3rd generation cementing technique was used. The reduction of the hip joint was done. Then, the abductor mechanism was repaired by fixating to trochanter majoris over the prosthesis. Very serious care was taken to cover the prosthesis completely with muscle tissue (Figure 1C). Immobilization was applied for 2–3 days post-operatively. Then the patients mobilized with weight-bearing exercise. At the first month visit the patient mobilized successfully and there was no wound infection. Lung metastasis was detected at the third month control. Pancreatic head metastasis was detected on the 6th month control. 3 cure IMA (ifosfamide, mesna, adriamicine) chemotherapy was applied postoperatively. The patient is under the follow-up for the last 3 years. Nodules of lung metastasis are stable.

Case 2

A 57-year-old man presented with right hip pain for 4 months. Physical examination of the right hip

joint, there was limitation of motion and pain during movement. Plain radiography showed radiolucency in the right acetabulum and iliac wing. The right hip MRI showed a tumoural lesion of 7.5×4 cm dimensions over the inferior iliac wing and extending to acetabulum which was destructing and thinning the cortical bone (Figure-2). Tru-cut biopsy was done and histopathological result was angiosarcoma (Figure-3,4). Under general anaesthesia the patient was placed on lateral decubite position. After securing the neurovascular bundle, the wide resection of the tumour completed with type 2 periacetabular resection. There maining defect after type 2 pelvic resection was reconstructed with a fresh frozen hemipelvis allograft and pelvic reconstruction plates, cancellous screws. Then graft's acetabulum was reamed and polyethylene acetabular component inserted cementless. After that femoral component inserted. (Figure-5). Immobilization was applied for one week. Then the patient mobilized without weight-bearing. The patient underwent postoperative chemotherapy and radiotherapy. Lung metastasis was detected at the second month control. Following the fifth month, the patient who applied with general condition disorder and malnutrition was hospitalized intensive care service. At the 7th month the patient was exitus.

Case 3

A 61-year-old woman was referred to our oncology clinic because of the left thigh and knee pain. It was noted by the patient that pain started 2 years ago and worsened over time. Physical examination of the patient revealed a soft tissue mass of 8×4 cm on the posteromedial left thigh. Plain X-ray films showed intramedullary heterogen radio dense areas. MRI showed a tumoural lesion which is 8 cm at its widest portion and heterogenously keeping contrast substance with minimal cortical destruction over the posterior distal femur. Tru-cut biopsy was performed and histopathological result was angiosarcoma. Bone scintigraphy showed involvement of the left distal femur (Figure-6). Under spinal anaesthesia, the left thigh was incisized 20 cm medial to the tumoural mass. A modular distal femoral tumour resection prosthesis was assembled after wide resection (Figure-7). The patient took radiotherapy postoperatively due to the surgical margin proximity. Superficial dermatitis occured related to radiotherapy. Debridement operation took place due to wound drainage on postoperative third month. Antibiotherapy was given. Lung metastasis occurred on the first-year follow-up control. The patient is under follow-up.

Case 4

A 36-year-old man was admitted to our clinic because of a pathologic fracture of collumfemoris. Plain X-ray films showed pathologic left femoral neck fracture and subtrochanteric destruction on MRI, there was a tumoural lesion with 8×6 cm dimensions and surrounding the bony tissue on left proximal femur, a fracture on left femoral neck, and cortical destruction over the subtrochanteric region. Tru-cut biopsy was done and histopathological result was angiosarcoma. The patient had lung metastasis on first application. The patient was treated by wide resection and modular proximal femoral tumour resection prosthesis. Four cures of IMA chemotherapy were given postoperatively. This patient some difficulty with walking. At one year follow up visit, lung metastases showed progression and the general condition of the patient was started to debilitate. The patient died on 16th month.

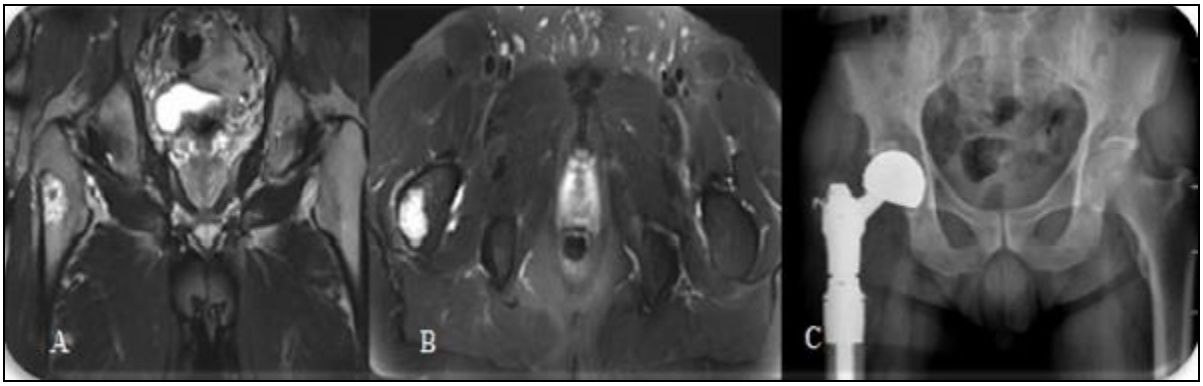


Figure-1: Illustrative case of a 54-year-old man presented with right hip pain for 6 months. (A,B) Coronal MRI showing a slightly lobulated subcortical exantric tumoural lesion with intact contours and sclerotic borders in the right femoral proximal intertrochanteric region (C) Postoperative radiograph showing tumour resection prothesis of the right proximal femur

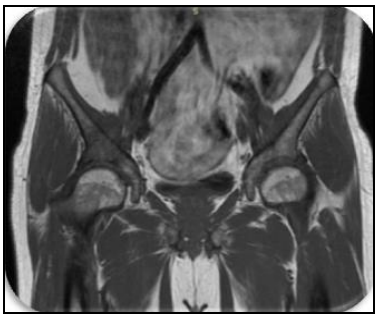


Figure-2: Illustrative case of a57-year-old man presented with right hip pain. Coronal MRI showed tumoural lesion over the inferior iliac wing and extending to acetabulum which was destructing and thinning the cortical bone and keeping contrast substance heterogeneously

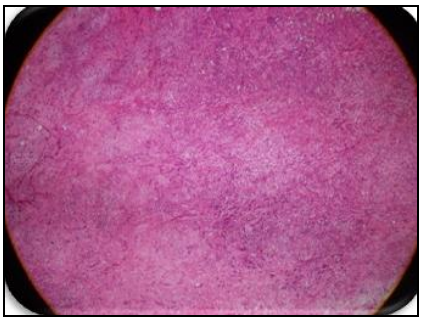


Figure-3: Haemorrhagic tumour tissue showing solid growth pattern. H-E, x40

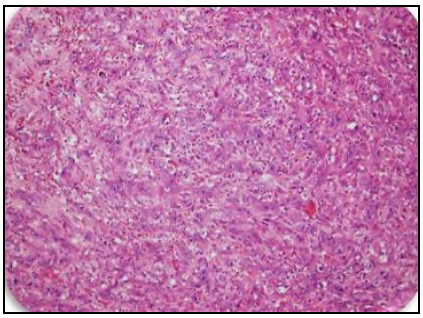
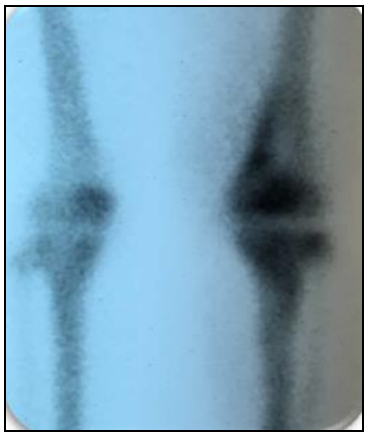
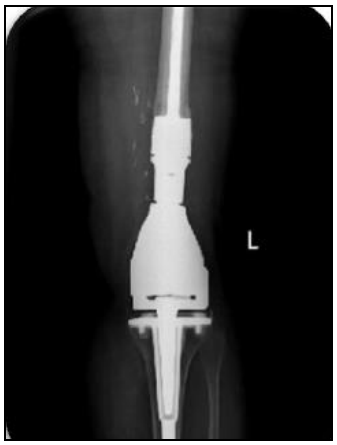


Figure-4: Tumour cells are large, vesicular nucleus and prominent nucleolus. In addition, intracytoplasmic, erythrocyte lumen formation was formed. H-E, x200

Figure-5: (A)Wide resection of the tumour completed with type 2 periacetabuler resection (B) the defects were reconstructed with a fresh frozen hemipelvis allograft and total arthroplasty of the hip joint





DISCUSSION

Angiosarcoma is very rare, malignant tumour comprising 1–3% of adult soft tissue sarcomas.[2,3,5,6]

Primary angiosarcoma of bone is seen in less than 1% of all primary bone sarcomas. It can be seen at all ages.[7] Many terms, including haemangioendothelioma, hemangiosarcoma, haemangioendothelial sarcoma, angiosarcoma and others have been used inter changeably in the literature. Although not precisely defined, most authors use the term hemangioendotheliomatoid entify low-grade malignant vascular tumours and the term angiosarcomatoid entifyhigh-grade malignant vascular tumours.[8] Although the clinical appearance of primary bone angiosarcoma is variable, pain and swelling are often the first symptoms.[9]

The radiographic appearance of this tumour is related to the degree of the tumour. Low-grade tumours have a well-demarcated lytic lesion and a reactive bone around them. High-grade tumours have a more permeable appearance.[8]

Malign vascular tumours can be multicentric. When an angiosarcoma of bone is defined, a skeletal examination is recommended to assess whether the patient has multicentric disease. Multicenter skeletal angiosarcoma has been reported.[10,11] In general, pathological fractures may be seen in high-grade primary bone sarcoma, but the incidence of pathological fractures is unknown.[12] Computerized tomography, magnetic resonance imaging and bone scintigraphy are used to determine the extent of lesion and soft tissue involvement. Three of our cases had bone and muscle involvement, the other one having only bone involvement.

Early diagnosis is the most important issue in the treatment of this tumour. Diagnosis is made by evaluation of microscopic findings after biopsy.[13]

Low-grade tumours show well-formed anastomosis vascular channels that are microscopically depicted by endothelial cells. High-grade lesions may show pleomorphism. Diagnosis in these pleomorphic lesions can only be made by immunohistochemistry. CD31 and CD34 should only be positive in vascular tumours.[8]

Treatment is planned according to each patient's own clinical situation. Solitary lesions are treated with extensive resection if possible. The role of radiation therapy and chemotherapy is being investigated.[9,14,15] Radiotherapy can be used in the treatment of surgically inoperable lesions or in the treatment of multiple lesions. Adjuvant chemotherapy can be added to the protocol for treatment of high-grade lesions. Prognosis is associated with tumour grade at the time of diagnosis. Patients with low-grade lesions have a better chance of long-term survival, while patients with high-grade tumours have a long-term survival rate of less than 20%.[8]

Our all cases treated with wide radical resection of the tumour. Three of the patients were reconstructed with modular tumour resection prothesis, the remaining one was reconstructed with bone allograft, plate and total hip prothesis. All our cases were treated with chemotherapy. The case number three was given radiotherapy because the surgical margin was nearer than 1 mm. Resection margins could not be widened due to the proximity of the lesion to neurovascular bundle. Lung metastasis occured in all of the patients up to first year control. Two patient died in the follow-ups in the first year.

As a result, we would like to emphasize once again that there are four unusual cases and that bone angiosarcoma has poor prognosis. It should be kept in mind, especially in patients over 50 years of age, due to nonspecific radiological findings and early symptoms of pain and characterization. Early diagnosis of bone angiosarcoma is of great importance in minimizing local and systemic complications. Complex resection is the most effective treatment and should be combined with adjuvant chemotherapy treatment.

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