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Primary malignant bone tumours of hematopoietic origin: A single-centre, restrospective study

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Abstract

Tumors of hematopoietic origin are rare tumors that can be confused with benign and malignant tumors of the bone with their clinical and morphological features. Plasma cell myeloma, solitary plasmacytoma, Non-Hodgkin Lymphoma, acute lymphoblastic leukemia/lymphoma, and Langerhans cell histiocytosis are hematological neoplasms that mainly involve the bone. Plasma cell myeloma is the most common primary malignant bone neoplasm, and diffuse large B-cell lymphoma (DLBCL) is the most common type of non-Hodgkin lymphoma arising in bone. In this case report, we will retrospectively investigate the diagnosis and treatment management and postoperative clinical outcomes of patients diagnosed with tumors of hematopoietic origin, evaluated by the Bone and Soft Tissue Tumor Council between 2005 and 2021, and emphasize the importance of the approach to these tumors.

Keywords: hematopoetic myelodysplasia, multiple myeloma, bone neoplasms, leukemia

1. Introduction

Neoplasias of hematopoietic origin are seen extremely rarely in bones. The most widely seen hematological neoplasias primarily involving the bone are plasma cell myeloma, solitary plasmacytoma, non-Hodgkin's lymphoma, acute lymphoblastic leukemia/lymphoma, and Langerhans cell histiocytosis (1). Although tumors of hematopoietic system origin are rarely seen, the clinical and morphological characteristics may be confused with malignant bone tumors and may also be confused with benign proliferations of the bone. This should be kept in mind in the differential diagnosis.

In this study, a retrospective investigation was made of the demographic data obtained from patients presented to the Bone and Soft Tissue Tumour Council,the diagnosis and treatment management of these tumors and the importance of the approach to these tumors are emphasized.

2. Material and Method

A retrospective examination was made of patients who presented at the Orthopaedics and Traumatology Polyclinic between January 2005 and December 2021, underwent advanced examination and were diagnosed with bone metastasis of hematopoietic origin. A record was made of the patient's age, gender, complaints, and definitive diagnosis. The clinical findings, radiological images, and pathology results, if present, were evaluated. All the patients included in the study were diagnosed radiologically and /or pathologically with bone tumors of hematopoietic origin.

3. Results

The evaluation was made of a total of 40 patients with bone tumors of hematopoietic origin diagnosed following bone biopsy or postoperatively from operation material, who presented at the Orthopaedics and Traumatology Polyclinic between 2005 and 2021.

The patients comprised 22 females(%55) and 18 males(%45) with a mean age of 57.45 years. The patients were classified as 18 multiple myelomas, 11 plasmacytoma, 9 non-Hodgkin's lymphoma, 1 Langerhans cell histiocytosis, and 1 amyloidosis (Table 1).

Table 1. Distribution of hematopoietic origin tumours

Hematopoietic origin tumours	n=40	%
Multiple Myeloma	18	45
Plasmacytoma	11	27.5
Non-Hodgkin's lymphoma	9	22.5
Langerhan's cell histiocytosis	1	2.5
Amyloidosis	1	2.5

Tumour localization was determined as mostly (42.5%) in the proximal femur. Involvement was seen in the femur in 20 patients, the humerus in 5, the tibia in 1, the pelvis in 6, the vertebra corpus in 1, the clavicle in 1, and the radius in 1. There was seen to be extramedullary involvement in the proximal thigh in 2 patients, in the arm in 1, in the anteromedial thigh in 1, and in the groin in 1. The diagnosis was made following a pathological fracture in 14 patients (37.5%).

Treatment was planned according to the patient's age,

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clinical condition, and tumor localization. The most frequently selected treatment was wide resection and tumor resection prosthesis (n=27), followed by curettage, cementation, and intramedullary nailing (n=6). Conservative follow-up with a splint was applied to 1 patient. Vertebroplasty was performed on 1 patient with vertebral involvement. The remaining 5 patients were referred to the medical oncology unit, with the decision to conservative orthopedic follow-up. Surgical treatment was performed on 2 patients diagnosed with multiple myeloma due to multiple involvement (Fig. 1 and 2).

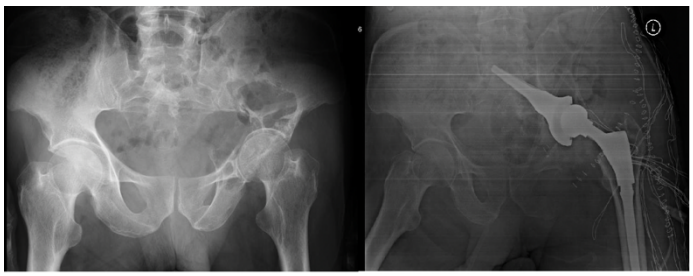

Fig. 1. In a 69-year-old male patient with multiple myeloma metastasis in the left iliac wing, a modular tumor prosthesis was applied following iliac wing resection

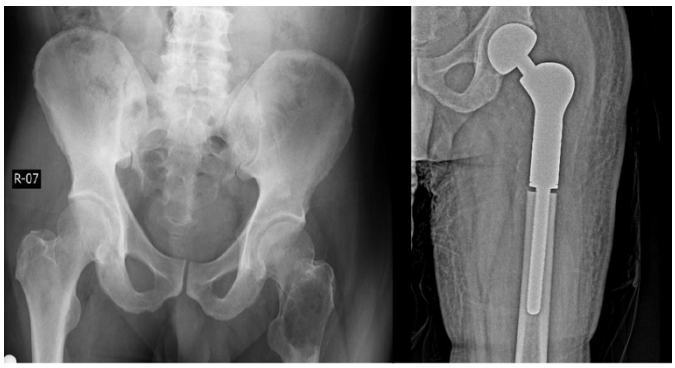

Fig. 2. A 53-year-old male patient with multiple myeloma metastasis in the left proximal femur was treated with proximal femur resection prosthesis.

4. Discussion

The skeletal system is the organ most frequently affected by metastatic cancers, and tumors originating from the breast, prostate, thyroid, lungs, and kidneys have a tendency to spread to the bones (2, 3). Neoplasias of hematopoietic origin are seen extremely rarely in bones. The hematopoietic-origin neoplasias primarily involving the bone are plasma cell myeloma, solitary plasmacytoma, non-Hodgkin's lymphoma, acute lymphoblastic leukemia/lymphoma, and Langerhans cell histiocytosis.

Multiple myeloma is the most frequently seen primary malignant bone neoplasm, and the complication seen most often in patients with multiple myeloma is bone involvement. Osteoclast over-activation is a process that disrupts the balance of bone remodeling, including the inhibition of osteoblasts, osteocytes, and bone marrow stromal cells (4). Of the 40 patients in this study with metastasis of hematopoietic origin, 18 (45%) were diagnosed with multiple myeloma. Due to multiple involvements in 1 patient diagnosed with multiple myeloma, a bilateral humerus shaft and femur proximal third fracture developed, and these were treated surgically (Fig. 3).

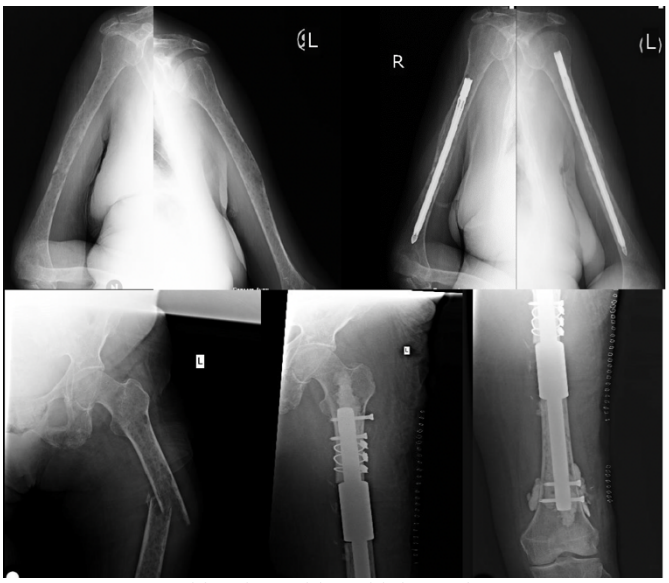

Fig. 3. A 67-year-old patient with multiple myeloma metastases was treated with bilateral humerus intramedullar nailing, and an intercalar tumour prosthesis for a left femur shaft fracture

Solitary plasmacytomas are rare and constitute <5% of plasma cell neoplasms. Solitary bone plasmacytomas generally form in the spine and skull, almost always emerge in the head and neck, and are more common than extramedullary plasmacytomas, which can spread to regional lymph nodes (5). In the current study, 11 (27.5%) patients were diagnosed with plasmacytoma, and involvement was determined in the proximal femur in 6 of these, in the distal femur in 1, in the clavicle in 1, in the proximal humerus in 1, and in the pelvic region in 2.

The most common subtype of malignant lymphoma is diffuse large B-cell lymphoma. Primary bone lymphoma (PBL) is rare and constitutes <1% of all non-Hodgkin's lymphoma. PBL is approximately 3% of all malignant and primary bone tumors (6). All bones in the skeletal system can be involved, but the long bones are most often affected, and the most commonly affected bone is the femur (7). In the current study, 9 (22.5%) patients had non-Hodgkin's lymphoma involvement, and of these 9, a diagnosis of extramedullary involvement was made in 6 cases.

Langerhans cell histiocytosis is originating from the uncontrolled proliferation and accumulation of immature myeloid dendritic cells of bone marrow origin and is an uncommon but serious inflammatory neoplasia. Any bone may be affected, but >50% of bone lesions are in the skull, ribs, and pelvis. Clinical presentation may be with pain, swelling, a soft tissue mass, sometimes bone deformity, and bone fracture (8). In the current study, an 11-year-old male patient was applied with curettage and grafting because of a lesion in the acetabulum, and the pathological diagnosis of the cyst material was consistent with Langerhans cell histiocytosis.

Bone metastases cause severe morbidity. Pain, restricted movement, hypercalcemia, pathological fracture, spinal cord or nerve root compression, and bone marrow infiltrations are

among the main complications (9). Metastatic destruction of the bone reduces the weight-bearing capacity of the bone and initially causes deterioration of the trabecular structure and microfractures, followed by loss of bone integrity. In the current study, 14 (37.5%) patients with bone involvement presented with complaints of pathological fracture.

Radiographs are generally the first step in the evaluation of bone metastases. Osteolytic and osteoblastic lesions may be determined. Scintigraphy is a highly sensitive method in screening bone metastases. In the early period, an increase in osteogenic activity is helpful in the determination of bone metastasis. Computed tomography (CT) and magnetic resonance imaging (MRI) are expensive but very sensitive and specific methods (9, 10). Prophylactic surgical procedures such as curative resection and reconstructive prostheses lead to better survival rates than osteosynthesis of pathological fractures in some patients (11). Arthroplasty, intramedullary nailing, and bone cement injections are other surgical procedures for the treatment of bone metastases.

Limitations of the current study were primarily the retrospective design and relatively low number of patients. A reason for the low number of patients was that only patients with bone metastases of hematopoietic origin who presented at the Orthopaedics and Traumatology Clinic were included. Finally, the treatment section was not given in detail in the study. Therefore, there is a need for further studies to examine larger patient populations with a primary malignant bone tumor of hematopoietic origin and discuss surgical and non-surgical treatments in more detail.

Tumors of hematopoietic origin are rarely seen, but the clinical and morphological characteristics may be confused with malignant bone tumors, so this must be kept in mind in the differential diagnosis. Patients of advanced age, in particular, presenting with atraumatic fractures, must be screened in respect of malignancy, must be examined in detail in respect of pathological fracture, and a treatment plan must be made. With the appropriate diagnostic tools and a multidisciplinary approach, adjuvant treatment selections, and the selection of extremity-sparing surgery as a result, it is possible to obtain successful treatment results.

Conflict of interest

The authors declared no conflict of interest.

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None to declare.

Authors' contributions

Concept: İ.B., Design: T.C., O.M, Data Collection or Processing: Ş.Ş., T.C. Analysis or Interpretation: N.D., H.S.C., Literature Search: Ş.Ş., O.M, Writing: H.S.C

Ethical Statement

Approval for the study was granted by the Institutional Ethics Committee (decision no: B.30.2.ODM.0.20.08/342-459 dated:27.07.2022).

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