

Non-Hodgkin's Lymphoma of Talus: A Case Report

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Abstract

Introduction: Primary non-Hodgkin's lymphoma of the bone (PLB) represents about 3% - 5% of all extranodal non-Hodgkin's lymphoma (NHL) cases and 7% of primary bone tumors. It may occur at any age. The peak incidence for PLB is in the fifth and sixth decades of life, and it has a slight male predominance. The most commonly affected sites are the long bones. A palpable mass due to soft tissue extension of the bony disease is seen in almost half of all cases.

Case Presentation: We report the case of an NHL (diffuse large B-cell lymphoma) in the talus bone of a 17-year-old boy who was referred to our center in 2011 - 2012, which had an odd presentation. The patient's first diagnosis was a simple ankle sprain, and he underwent conservative treatment. Given the patient's deteriorating symptoms, further paraclinical evaluations were conducted, and fracture of the talus was diagnosed, which was thought to be a cause of further osteonecrosis. Finally, because of lack of pain relief and due to MRI images, the suspicion of malignancy was raised. Open biopsy of the talus showed NHL (diffuse large B-cell lymphoma).

Conclusions: The present case is interesting because it expresses how such lesions can be met with diagnostic confusion.

Keywords: Primary Non-Hodgkin's Lymphoma, Talus, Combined Therapy, Osteonecrosis

1. Introduction

Primary non-Hodgkin's lymphoma of bone (PLB) represents about 3% - 5% of all extranodal non-Hodgkin's lymphoma (NHL) cases and 7% of primary bone tumors. It may occur at any age. The peak incidence for PLB is in the fifth and sixth decades of life, and there is a slight male predominance. Most patients complain of localized pain or swelling (1-5). The present case is interesting because it shows how such lesions can be met with diagnostic confusion.

2. Case Presentation

Our patient was a 17-year-old male student whose chief complaint was ankle pain and swelling due to falling from a height (1 meter). The patient was referred to the Rasoul-e-Akram Hospital in Tehran, Iran (2011 - 2012). Physical examination showed normal lower extremity alignment with functional range of motion of all joints. The patient's gait was considerably antalgic. His ankle was stable. The skin and neurovasculature were intact. There was mild tenderness over the anteromedial aspect of the ankle. The patient's initial x-rays showed no pathology (Figure 1), and his symptoms were treated by wearing a short leg cast as ankle sprain. After six weeks, the ankle swelling, pain, and tenderness were not relieved, and the patient was referred to

an orthopedic surgeon in Tehran, Iran. Physical examination revealed no palpable mass or lymphadenopathy. Examination of other limbs revealed no abnormalities. Laboratory studies, including CBC (complete blood count), serum calcium, ESR (erythrocyte sedimentation rate), CRP (C-reactive protein), and serum protein electrophoresis were normal. All instruments were calibrated before use (models of used devices included MRI, Inginia, Philips 1/5 tesla; CT scan: Toshiba, 16 slices). Initial radiographs of the ankle and foot did not show any pathological changes; thus, the patient underwent an ankle CT scan (Figure 2). A fracture was seen in the talus body, and the patient was treated with a short leg cast. Twelve weeks after the patient's presentation, ankle swelling, pain, and tenderness became worse. Because symptoms persisted, the patient consulted a foot and ankle orthopedist, who believed that the symptoms might be caused by osteonecrosis of the talus. Thus, the patient underwent an ankle MRI with and without contrast. The radiologist reported that the findings of the MRI were compatible with talus osteonecrosis, and the patient was treated conservatively. Symptoms and signs worsened after three months of conservative treatment. Significant swelling in the soft tissue surrounding the talus and severe collapse and cortical destruction in the new ankle radiographs were observed. Upon re-evaluation of the MRI, extensive changes in the soft tissue surrounding the talus and the calcaneus were noted. In addition,

cortical destruction in the talus and medial malleolus associated with a change in signal intensity were identified in the talus and the distal tibia (Figure 3).



Figure 1. AP and Lateral View X-Rays Show no Pathology

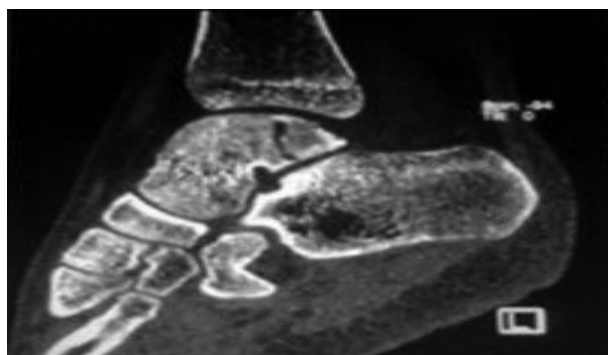


Figure 2. CT-Scan Sagittal View Shows a Fracture in the Talus Body

These findings raised the suspicion of malignancy. Open biopsy of the talus revealed NHL (diffuse large B-cell lymphoma).

3. Discussion

Primary lymphoma of bone (PLB) is a rare disease that comprises less than 1% - 2% of all lymphomas in adults. The cause of primary NHL of bone is not currently known. Certain subtypes of NHL have been revealed to be associated with a virus (Epstein-Barr virus [EBV], human immunodeficiency virus [HIV], and human herpes virus-6 [HHV-6]) (1). However, viral etiology has not been investigated to any great extent in primary NHL of bone. PLB may occur anywhere in the skeleton. The most commonly affected sites are the long bones. The femur has been described as the most common site to be involved when only one isolated site is affected (2-5).

Recently, Beal et al. (6) Studied the clinical characteristics of 82 PLB patients and reported that the most common sites to be involved are the femur (27%), the pelvis (15%), the tibia (13%), the humerus (12%), the spine (9%), the mandible (2%), the skull (1%), the scapula (1%), the radius (1%), and the ulna (1%). The most common presenting symptom is pain without antecedent trauma that is unrelieved by rest. Patients may also present with a palpable mass, swelling, limping, night pain, pathological fractures, spinal cord compression, and systemic B symptoms (fever, night sweats, weight loss) (2-4, 7). Our case presented with pathological fracture after trauma. Diagnosis of the patient was delayed by about six months.

No particular radiological findings are seen in PLB; when it is noticeable, PLB may have heterogeneous appearance, with lytic, blastic, and mixed lesions reported (6, 8). One typical picture is a solitary lytic lesion near the end of a long bone. Additional imaging findings include periosteal reaction, soft-tissue extension, pathological fracture, and cord compression (8). The radiological differential diagnosis includes benign entities (reactive conditions, osteomyelitis) and malignant entities (Hodgkin's lymphoma, sarcoma, neuroblastoma, metastatic disease). In our patient, pathological fracture was the initial presentation. It should be noted that this location of PLB is rare; one case reports a 58-year-old male with primary NHL of the talus and with no history of trauma.

Over the last two decades, combined therapy has become the standard treatment for PLB. A study at the memorial sloan kettering center (9) evaluating various treatment strategies (i.e., radiation therapy alone, chemotherapy alone, or combined-modality therapy), showed significant benefits with combined therapy. Combined modality therapy has been associated with a five-year survival rate of 95%, versus 78% for patients treated with single modality therapies.

More recently, Ramadan et al. (3) reported the use of rituximab in 28 PLB cases. Although there was only a short period of follow-up, they were able to demonstrate that rituximab improved progression-free survival; the three-year progression-free survival rate for patients who received rituximab plus cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) was 88%, compared with 52% for those who received standard CHOP alone.

Our patient achieved complete remission with combined therapy (chemotherapy and radiotherapy). Our patient is under five years of follow-up, and his only significant complaint is limping. The patient's one-year follow-up x-ray is shown in Figure 4.



Figure 3. Ankle T1- and T2-weighted magnetic resonance imaging shows signal intensity changes in the talus and the distal tibia, with abnormal enhancement following contrast injection. There is also soft tissue involvement surrounding the talus.



Figure 4. AP and Lateral Views After One-Year Follow-Up

3.1. Conclusion

No particular radiological findings are observed in PLB; when noticeable, PLB can have a heterogeneous appearance, with lytic, blastic, and mixed lesions reported. One typical picture is a solitary lytic lesion near the end of a long bone. In our patient, pathological fracture was the initial presentation. It should be noted that this location of PLB is rare. Over the last two decades, combined therapy has become the standard treatment for PLB. Our patient achieved complete remission with combined therapy (chemotherapy and radiotherapy). Our patient is under five years of follow-up, and his only significant complaint is limping

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Footnote

Conflicts of Interest: The authors have no conflicts of interest concerning the publication of this paper.

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