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Case Report

Primary Bone Lymphoma of the Patella, Rare Presentation of Primary Non-Germinal Center B Cell-Like Diffuse Large Cell B Cell Lymphoma, Case Report and Review of The Literature

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Abstract

Very few cases of primary bone lymphoma of the patella have been reported in the literature, we are presenting a young adult patient who presented with chronic knee pain that failed to resolve with medications and physical therapy for long time. Magnetic resonance imaging was done that showed abnormal infiltrative lesion which was biopsied and showed typical changes of bone lymphoma and subsequently had PET scan that showed a solitary involvement of the patella with no other lesions elsewhere in the body. Patient was started on chemotherapy and showed excellent clinical response.

Keywords: *Bone Lymphoma; Patella*

Abbreviations

MRI: Magnetic Resonance Image; NSAIDs: Non-Steroidal Anti-Inflammatory Drug; PET Scan: Positron Emission Tomography

Introduction

Primary bone lymphoma (PBL) was presented the first time in 1928 and described by Parker and Jackson as a clear clinical entity in 1939. It affects bones as the primary site and described as a type of lymphoma with malignant lesions [1,2]. PBL is an unusual presentation that accounts for only 2% of all primary bone tumors [3]. The disease has a male dominance with males two times more commonly affected compared to females [4] Almost PBL cases presented by themselves clinically at median age > 30 years [4]. In the literature review, the majority of reported cases were diagnosed as non-Hodgkin diffuse large B-cell lymphoma [5]. Diagnosis of PBL depends on the integration of clinical presentation, imaging studies, and confirmed with microscopic evaluation of bone biopsy an immunohistochemical studies. The patella is an extremely rare site of PBL. To date, only five cases have been reported as PBL of the patella among the adult population [6-10]. The Importance of differentiation of PLB from other types of bone tumors is Significant because PBL has a better prognosis and response to therapy compared to cases with systemic lymphoma with secondary bone involvement [11]. Radiologically, PBL has the same features of histiocytic lymphoma, including ill-defined margins around osteolytic lesions [12]. Because of the rarity of this disease, more related studies and case reports are needed to provide perception about this obscure lymphoproliferative malignancy. Here, we are reporting one rare case of primary bone Lymphoma involving the patella of the left knee. Meantime, clinical, imaging, and immunohistochemical aspects, therapeutic options, and related literature review are discussed. To the best of our knowledge, this is the sixth case reports of PBL of a patella in the English literature.

Case Report

A 40-year old male patient was referred to the orthopedic oncology clinic for further evaluation and management regarding an abnormal lesion in the patella. Patient is previously healthy, reported a history of recurrent anterior knee pain for 2 years that failed to respond to medications and frequent courses of physical therapy. Pain is associated with swelling, difficulty in knee bending, climbing up and down stairs and affecting his functional activities. Physical examination showed anterior patellar tenderness with soft tissue swelling medially as well as inferiorly under the patellar tendon. Knee showed full range of motion in flexion and extension with pain at full flexion. There was no local or regional lymph node enlargement and no hepatosplenomegaly. The rest of general physical examination is unremarkable. Plain radiographs of the knee in AP and lateral projection showed heterogeneous bone density of the patella, with a permeative lytic lesion involving the inferior aspect of the patella with cortical disruption (Figure 1A). Magnetic resonance imaging in sagittal post contrast enhanced T1 fat-saturated showed an enhancing lesion at the inferior patella with an enhancing soft tissue component involving Hoffa's fat pad, and coronal T1 weight MR image through the patella, showing abnormal dark T1 signal of the entire patella indicating replacement of the normal bone marrow by a tumor, with disruption of the inferior patellar surface and soft tissue component near the patellar tendon attachment (Figure 1B and 1C). PET - Computed tomography shows a lytic patellar lesion with increased FDG uptake consistent with an aggressive osseous process raising the suspicion of malignancy. The whole-body PET scan revealed FDG uptake in the left knee, with no other abnormal focus of FDG uptake in the body (Figure 1D and 1E).

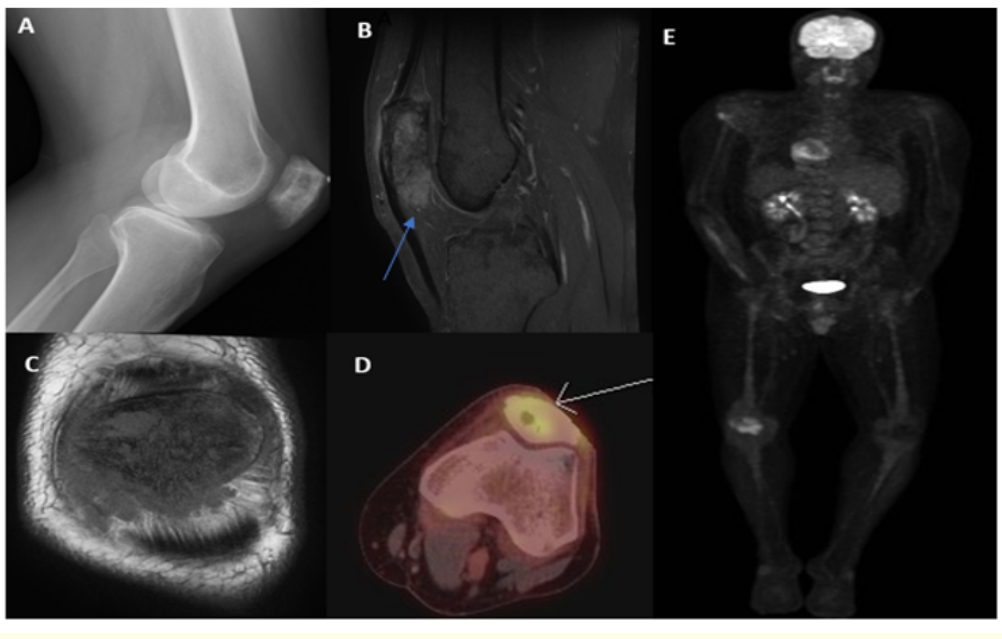

Figure 1: *A- Lateral Knee Radiograph showing heterogenous bone density of the patella, with a permeative lytic lesion involving the inferior aspect of the patella with cortical disruption. B- Sagittal Post contrast enhanced T1 fat-saturated MR image showing an enhancing lesion at the inferior patella with an enhancing soft tissue component (blue arrow) involving Hoffa's fat pad. C- Coronal T1 weight MR image through the patella, showing abnormal dark T1 signal of the entire patella indicating replacement of the normal bone marrow by tumor, with disruption of the inferior patellar surface and soft tissue component near the patellar tendon attachment. D- Axial PET/CT image at the level of the knees shows a lytic patellar lesion with increased FDG uptake consistent with an aggressive osseous process (white arrow). E- Scout PET image showing FDG uptake in the left knee, with no other abnormal focus of FDG uptake in the body.*

Open biopsy was done and a sample was taken from the soft tissue component and sent for histopathological examination. Histological sections of the patellar mass reveal infiltration of the connective and adipose tissue by atypical large cells with abundant cytoplasm, round and occasionally irregular nuclei with vesicular chromatin, prominent nucleoli and frequent mitosis. All immunohistochemical tests were performed on formalin-fixed (6 - 72 hours), paraffin-embedded sections using ultra-view universal DAB detection system from FDA approved Ventana and interpreted with proper controls. The large cells are diffusely positive for CD45, CD20, BCL-6, and MUM-1. However, they are negative for CD5, CD10, BCL-2, CD30, and EBV by EBER in situ hybridization. Ki-67 proliferation index is high at 70 - 80%. Cytogenetics studies showed absence of MYC, IgH/BCL-2 and BCL-6 gene rearrangements. The diagnosis was consistent with diffuse large B-cell lymphoma (DLBCL), non-germinal center subtype (Figure 2). Bone marrow biopsy revealed a normocellular bone marrow with no evidence of lymphoma. Overall, these findings were most consistent with the diagnosis of primary bone adult B-cell lymphoma of the patella. The case was discussed with the lymphoma team and was referred for medical oncology for treatment by chemotherapy. Patient attended to the clinic for follow up three months after treatment and showed excellent response to treatment clinically with marked reduction of the soft tissue swelling as well as resolution of his symptoms.

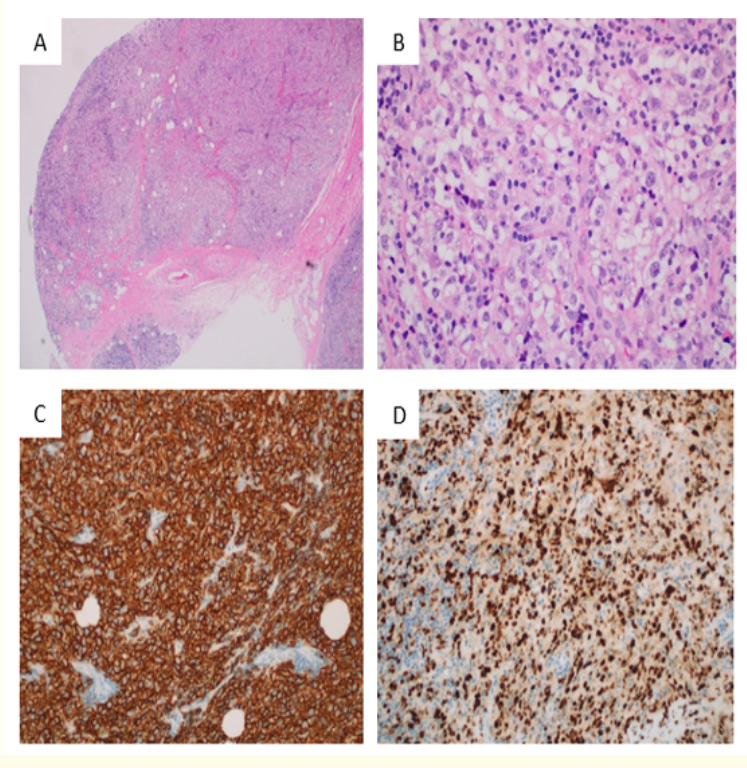

Figure 2: *A- Diffuse infiltration of connective and adipose tissue (H&E, original magnification x40). B- The infiltrate is composed of large cells with irregular nuclei, vesicular chromatin, and prominent nucleoli (H&E, original magnification x400). C- Diffuse strong expression for CD20(Immunohistochemistry, original magnification x100). D- Ki-67 proliferative index is high at 70-80% (Immunohistochemistry, original magnification x100).*

Discussion

PBL is a malignant bone tumor that can affect any bone in the skeleton without affecting other internal organs or distant lymph nodes. PBL is a rare tumor and it is less than 1% of all non-Hodgkin lymphomas and around 3% of all malignant bone tumors. Tumor of the

patella is generally rare, with mainly benign etiology and about 0.12% of all bone tumors [13,14]. PBL generally presented with localized pain, swelling, and occasionally presented with a decreased range of motion. Radiographically, PBLs presents as a lytic bone lesions in radiographs with similar radiological findings with the PLB of the patella in our case [15]. To this date, as we found in the literature review only five cases reported as PBL in the patella. Our patient presented with knee pain more than one year with no affection on range of motion, which are same symptoms of the other five cases in the literature review mentioned in table 1. Only two cases with MRI report to compare with our MRI finding. In our case, the MRI of the patella showed enhancing lesion at the inferior patella with a soft tissue component. Yamamoto., *et al*. [16], showed the lesion to exhibit a low intensity in T1-weighted images and high intensity in T2-weighted images with pathological fracture. Jadidi., *et al.* [10] showed decreased signal on T1-weighted images, increased signal on STIR, and avid contrast enhancement.

Author, year	Age	Sex	CC	X-ray of patella	PF	IT	Biopsy	Treatment	Prognosis
Chandra 1999	52	F	Knee pain	Destruction, a moth-eaten	+	CD20, CD45	Large B-cell lymphocytes	Chemotherapy, Radiation	After 13 month CR
Hughes 2000	76	M	Knee pain, arthritis	Destruction	+	-	-	Patellectomy, chemotherapy, radiation	Nodal recurrence of femur
Agarwal 2001	72	M	Knee pain, arthritis	Lytic area, a moth-eaten	-	CD3, CD45	T-cell lymphoma	Chemotherapy	NA
Yamamoto 2016	56	F	Knee pain, arthritis	Lytic area	+	CD20, CD79a	Diffuse large B-cell lymphoma	Chemotherapy, Radiation	DOD
Jadidi 2019	58	F	Knee pain, decrease ROM	Lytic area	-	CD45, CD2, CD3, CD4, CD5, CD8, CD25, FOXP3	T-cell lymphoma	Chemotherapy, Radiation	NA
Our case 2020	40	M	Knee pain	Lytic area	-	CD45, CD20, BCL-6, MUM-1	Diffuse large B-cell lymphoma	Chemotherapy	NA

Table 1: *Details of 6 cases on primary malignant lymphoma of patella.*
CC: Chief Complaint; ROM: Range of Motion; PF: Pathological Fracture; IT: Immune Type; DOD: Die of Disease; CR: Complete Response; NA: Not Available.

The treatment of primary bone lymphoma was based on multidisciplinary team approach with the main treatment modality is chemotherapeutic agents. The indications of surgery are very limited to obtaining biopsy as well as to fix fractures if the patients presented with fracture. The disease usually carries a good prognosis and low risk of recurrence and systemic spread.

Conclusion

Primary bone lymphoma of the patella is a rare bone tumor. The diagnosis is based on high index of suspicion with the presence of aggressive bone lesion associated with a large soft tissue component and confirmed by biopsy. It is also important to rule out any systemic involvement to diagnose the case as primary bone lymphoma. Treatment is usually chemotherapy and carries a good prognosis.

Summary

Primary bone lymphoma is a rare disease accounting for less than two percent of primary bone tumors. Most of the lymphomas are non-Hodgkin diffuse large B-cell lymphoma. The patella is an extremely rare site of presentation and the commonest presentation is anterior knee pain that failed to respond to medical treatment. Treatment consist of chemotherapy and carries a fairly good prognosis.

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