A Case Report of Primary Extranodal Non-hodgkin Lymphoma First Presentation as a Soft Tissue Swelling Around the Wrist

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What to Learn from this Article?

Extranodal Non-Hodgkin Lymphoma as a differential of Swelling around the wrist.

Abstract

Introduction: Primary musculoskeletal extranodal non-Hodgkin lymphoma is a rare presentation and account for 5% of all primary extranodal non-Hodgkin lymphomas. Treatment uses a combination of chemotherapy and radiotherapy with good prognosis in unifocal manifestation. We report an unusual case of primary musculoskeletal extranodal lymphoma presenting as a soft tissue swelling around the wrist.

Case Report: A 75 year old lady was referred to the Orthopaedic Outpatients Department with a painless, slowly growing mass on the dorsum of the right wrist. Clinical examination revealed a 6 X 9 cm round painless mass on the dorsum of the distal radius adherent to both the underlying structures and skin. MRI of the wrist showed a large mass causing extensive osteolysis of the distal radius and extending proximally with abnormal replacement of the marrow. The patient was brought to theatre for biopsy and subsequent histopathological examination confirmed a B-cell non-Hodgkin lymphoma. The patient was referred to the Haematology Service for further treatment and follow-up. She received chemotherapy and radiotherapy with satisfactory results.

Conclusion: Lymphoma presenting as a soft tissue mass is relatively uncommon and can easily be confused with a wide variety of inflammatory conditions, more common neoplasias as well as infectious diseases (tuberculosis). Though rare, extranodal lymphoma should be regularly included in the differential diagnosis of mass lesions.

Keywords: extranodal lymphoma; B-cell non-Hodgkin lymphoma; soft tissue mass.

Introduction

Primary musculoskeletal extranodal non-Hodgkin lymphoma is a rare presentation and account for 5% of all primary extranodal non-Hodgkin lymphomas [1]. At least one quarter of non-Hodgkin lymphomas arise from tissue other than lymph nodes and even from sites which normally don't contain lymphoid tissue [2,3]. These forms are referred to as primary extranodal lymphomas. Musculoskeletal lymphomas are rare entities and only 5% of these are primary extranodal non-Hodgkin lymphomas [4]. We describe an unusual and dramatic presentation of a patient diagnosed with this rare condition emphasising on the importance of having a good knowledge of the existence of other not so common possible diagnoses.

Case report

A 75 year old lady was referred to the Orthopaedic Outpatients

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Author's Photo Gallery


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Figure 1, 2: Macroscopic appearance of the mass.

Figure 3, 4: Radiographs showing malunion of the distal radius with areas of osteolysis .

Department with a painless, slowly growing mass on the dorsum of the right wrist. She first noticed the swelling a year previously and attended her local doctor who felt it was a ganglion. Past medical history included hypertension, osteoarthritis, polymyalgia rheumatica and malunion of a right distal radius following a distal radius fracture sustained 5 months prior for which she declined surgical intervention (Fig 3,4). Clinical examination revealed a radially deviated wrist with a classic dinner fork deformity. There was a 6 X 9 cm round painless mass on the dorsum of the distal radius adherent to both the underlying structures and skin which did not transilluminate and with necrotic area of overlying skin (Fig.1,2). General physical examination was unremarkable with no obvious lymphadenopathy, no breast masses, no organomegaly and clear lung fields. Her right hand had capillary refill of under 2 seconds, palpable radial and ulnar pulses and normal sensory and motor exams. Mantoux test was negative making TB an unlikely cause. Blood count, renal and liver function were within normal range, ESR 74 (elevated), haemoglobin 11 g/dl. On ultrasound scan there was a large vascularised soft tissue mass infiltrating the tendinous structures on the dorsum of the right wrist. MRI of the wrist showed a large mass causing extensive osteolysis of the distal radius and extending proximally with abnormal replacement of the marrow in the distal one third of the radius. The soft tissue component of the mass involved the entire extensor tendon groups and infiltrated through the subcutaneous tissues to the dermis (Fig. 5,6,7). CT thorax abdomen and pelvis and whole body bone scan confirmed that this was an isolated lesion with no evidence of dissemination. The patient was brought to theatre for biopsy following consultation from the National Musculoskeletal Tumour Centre. Subsequent histopathological examination confirmed a B-cell non-Hodgkin lymphoma (diffuse large B-cell lymphoma -DLBCL- right wrist-stage 1EA). The patient was referred to the Haematology Service for further treatment and follow-up. She received chemotherapy (R-CHOP C6 plus Rituximab and R-CHOP C4) and radiotherapy (IFRT) with satisfactory results that caused the mass to shrink considerably (more than 50% after the first cycle of radiotherapy) and subsequent CT scans didn't show any evidence of metastases. Despite her good response to the therapy, this lady died from causes not related to her primary diagnosis (pulmonary embolism).

Discussion

Common sites for the development of extranodal lymphoma are the skin, head, neck and gastrointestinal tract [2,8,9,10]. True primary lymphomas of soft tissues are extremely rare and there are few well-documented cases in the literature [5]. Only a few cases of NHL affecting musculoskeletal system have been described in the literature. The atypical presentation leads to difficulties in timely diagnosis and treatment. Although there is no specific radiologic feature to point to the diagnosis, the appearance of a permeative lytic lesion [9] corroborated with the anatomical location should raise suspicion. Infiltration of the subcutaneous fat is a striking feature in a majority of cases on CT and MR[11].

A definitive diagnosis requires histological examination of the specimen. DLBCL is one of the most common histological type among NHL and typically presents as a destructive infiltrating mass[2,12]. Not having a specific clinical or radiological characteristic makes it easy to confuse extranodal NHL with a series of conditions like ganglia, hemangiomas, lipomas, nonspecific inflammatory process, sarcoidosis, neolplasia (metastastasis, sarcoma, plasmocytoma, Hodgkin's lymphoma), connective tissue diseases, mycobacterial infections, especially in patients with immune compromise, fungal infections (histoplasmosis, cryptococcosis)[6]. A high index of suspicion, proper radiological imaging and histological diagnosis are essential in avoiding misdiagnosing this condition.

Treatment of primary extranodal non-Hodgkin's lymphomas uses combinations of chemotherapy and radiation therapy. Operative treatment is reserved for the treatment of complications. The prognosis of primary non-Hodgkin's lymphomas is reflected by 10-year-survival-rates without recurrence of more than 80% in unifocal manifestations [1]. A literature search using PubMed with Mesh terms "lymphoma" AND “muscular” AND “skeletal” AND “b cell” retrieved 69 results in English language of which 15 were relevant to this topic. 2 of the article were excluded as they

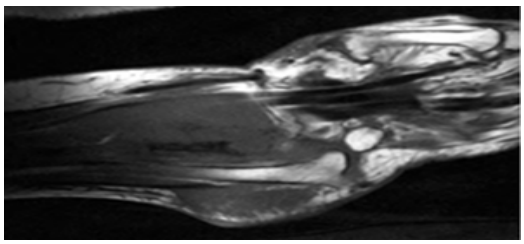

Figure 5: MRI - coronal view of the wrist (T1 weighted) .

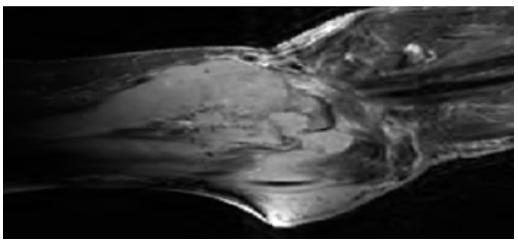

Figure 6: MRI - coronal views of the wrist (T2 weighted) .

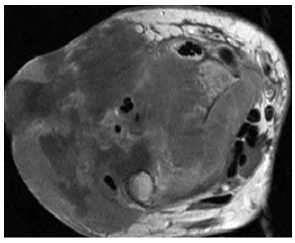

Figure 7: MRI - axial view of wrist (T1 weighted).

Authors and year of publication	Title	Research design	Sample details	Treatment	Conclusion
Morihiro Katsura, Hirokazu Nishina, Yasushi Shigemori, and Takaya Nakanishid 2015	Extranodal lymphoma originating in the gluteal muscle with adjacent bone involvement and mimicking a soft tissue sarcoma	Case report	52 yo female with DLBCL in the left gluteal muscles	8 cycles of R-CHOP chemotherapy	Mass has completely disappeared
Azam Alamdari, Neda Naderi, Soheil Peiman, and Farhad Shahi. 2014	Non-Hodgkin lymphoma with primary involvement of skeletal muscle	Case report	32 year old male with DLBCL of paraspinal, iliopsoas and gluteal muscles	R-CHOP chemotherapy	Complete remission without relapse
Matikas A, Oikonomopoulou D, Tzannou I, Bakiri M. 2013	Primary abdominal muscle lymphoma.	Case report	84 year old female with DLBCL within the abdominal muscles		Discussion over the features of the disease and pathological diagnosis
Guastafierro S, Falcone U, Petriccione L, Rossiello L, Cappabianca S, Rossiello R, Colella G. 2011	An unusual cause of facial swelling: primary extranodal non-Hodgkin lymphoma of the masseter muscle.	Case report	49 year old male with DLBCL of the masseter muscle	R-CHOP chemotherapy	No relapse
Liapi A, Dhanasekar G, Hock YL, East DM. 2006	An unusual case of primary extranodal non-Hodgkin's lymphoma in the muscles of facial expression.	Case report	1 patient with NHL involving the muscles of facial expression	chemotherapy	No relapse
De Giorgi S, Piazzolla A, De Giorgi G, Cimmino A, Parisi G, Ricco R. 2004	Non-Hodgkin's lymphoma in the gluteal region: a case report.	Case report	60 yo female with NHL of the gluteal muscles	wide-margin surgery	No recurrence
Borazan A, Ustün H, Ecirli S. 2003	Primary non-Hodgkin lymphoma of skeletal muscle coexistence with cutaneous infiltration.	Case report	73 year old male with DLBCL upper extremity		Unable to obtain full text
Choudhury J, Yalamanchil M, Friedenberg W. 2002	Skeletal muscle lymphoma.	Case report	1 patient with NHL of muscle of the lower extremity		Unable to obtain full text
Baddour LM, Haden KH, Allen JW. 2001	Primary skeletal muscle lymphoma presenting as refractory cellulitis.	Case report	55 year old female with B-cell NHL of the muscles of the right torso		Unable to obtain full text
Bertoni F1, Sanna P, Zucca E, Roggero E, Cavalli F. 1998	Primary extranodal lymphoma of skeletal muscles: a report of four cases.	Case series	3 of 4 cases were diagnosed with DLBCL	CHOP-like regimen alone or a combined with radiotherapy	Unable to obtain full text
Keung YK, Liang R. 1996	Report of a case of primary skeletal muscle lymphoma and review of the literature.	Case report			Unable to obtain full text
Jeffery GM1, Golding PF, Mead GM. 1991	Non-Hodgkin's lymphoma arising in skeletal muscle.	Case series	4 patients with NHL		Unable to obtain full text

described musculoskeletal involvement in HIV positive patient diagnosed with NHL. Given the heterogeneity of symptoms and presentations in these patients, the two articles were considered non representative for the condition described in this paper. Two papers describe a mass histologically T cell lymphoma.A number of 11 papers were related to the topic in discussion.

All describe case reports or cases series demonstrating the rare nature of this condition. Where full text was obtained, it is shown the importance of including NHL among the differential diagnoses. Radiological and histopathological examinations are paramount in making the correct diagnosis and delivering the correct treatment. Chemotherapy is the treatment of choice with added radiotherapy in selected cases.

Conclusion

Lymphoma presenting as a soft tissue mass is relatively uncommon and can easily be confused with a wide variety of inflammatory conditions, more common neoplasias as well as infectious diseases. Extranodal involvement by lymphoma has been described in every tissue of the body with multiple different appearances even within one organ system. Thus, it should be regularly included in the differential diagnosis of mass lesions [7]. Diagnosis is formally established by pathology.

Clinical Messege

Extranodal lymphoma is a rare entity that can mimic a wide range of more common conditions. It is important to have a high index of suspicion when assessing a mass and differential diagnosis should always include lymphoma.

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