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Primary Bone Lymphoma Revealed by Medullary Compression

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Abstract

Primary lymphoma of bone (PLB) is a rare entity characterized by nonspecific clinical signs that explain the delay in diagnosis. From a case report of PLB involving in multiple spine sites, we review the main features and the importance of histological evidence for diagnosis.

Keywords: *Primary lymphoma of bone; Percutaneous biopsy; Treatment; Medullary compression*

Introduction

Malignant lymphomas are defined by a cellular proliferation of lymphoid tissues, both ganglionic and extra-ganglionic. Primary bone lymphoma or PARKER's and JAKSSON's sarcoma can be distinguished as a histological variety of non-Hodgkin's lymphoma, the particularity of which is that it affects a single or multiple bone site with or without local lymph node involvement, without any other sign of the disease in other sites and without any blood spread within 06 months of diagnosis [1,2].

We report a case report of primary multifocal bone lymphoma complicated by bone marrow compression in a 70-year-old man.

Case Report

A 70-year-old diabetic patient, followed for corticosteroid-dependent peripheral thrombo cytopenia, developed paraplegia while maintaining a satisfactory general condition. He reported inflammatory rachialgia for three years. In biology, he had inflammatory anemia and moderate hypercalcemia. His Lacticodeshydrogenase level was twice normal. Spinal magnetic resonance imaging (MRI) revealed diffuse involvement of the stepped axial skeleton and pelvis with tumor bone marrow replacement type, D10 vertebral compression with posterior wall recoil, soft tissue extension, and a mass effect on the spinal cord **(Figure 1).**

Further exploration by sternal puncture and osteo-medullary biopsy were without signs of malignancy. As part of the extension workup, a bone scan revealed diffuse involvement of the axial skeleton, skull, costal grill, and right sacroiliac. The thoracoabdominal CT scan was without abnormalities suggesting primitive lesions. In view of the need for histological evidence, a bone biopsy of the right iliac crest lesion was performed, showing the appearance of large cell B lymphoma. He was referred to the oncology centre for overall evaluation and therapeutic decision. He was then treated with Rituximab

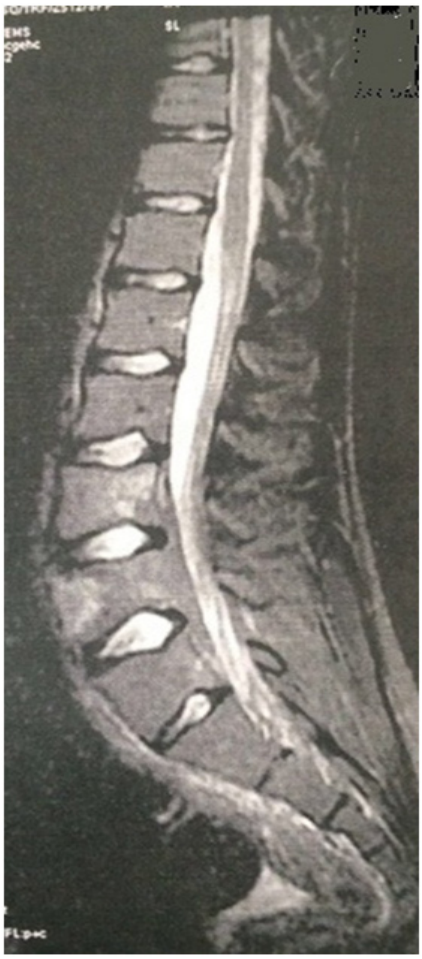

Fig1. *Spinal Magnetic Resonance Imaging (MRI) shows diffuse involvement of the stepped axial skeleton and pelvis with tumor bone marrow replacement type, D10 vertebral compression with posterior wall recoil, soft tissue extension, and mass effect on the spinal cord.*

Discussion

Primary bone lymphoma is a rare entity [3] affecting 2% of all lymphomas and 5% of extra-ganglionic sites [4]. It affects both sexes with a male predominance (sex ratio (M/F) 1.5) and an average age of onset in the 4th decade. It is generally unique, affecting a single bone site with a predilection for the long bones of the femur, tibia, pelvis, then the rachis, mandible, scapula, and rarely the skull [5]. Multifocal involvement [6], as in our patient, is described in 11-33% of cases. Diagnosis is delayed due to non-specificity of clinical and radiological signs with a mean delay between the onset of symptoms and confirmation of the diagnosis of 8 months. This delay was longer for our patient explaining the occurrence of spinal cord compression type complications. The revealing signs may be generally inflammatory bone pain, pathological fractures affecting the long bones or vertebral compression with neurological signs of compression [7] as described in our case. Bone swelling may also be a sign. At the biology we can note anemia, lymphopenia and thrombocytopenia. A biological inflammatory syndrome, an increase in the level of lacticodeshydrogenase and B2 microglobulin can also be seen. Hypercalcemia is noted in 10% of cases of the advanced forms. Bone damage may take different non-specific radiological forms [8-9]. The lesions may be osteolytic in 70% of cases, rarely isolated osteocondensate or a mixed aspect (10-16%) more suggestive of primary bone lymphoma. Standard radiographs may be normal at first. CT scan is indicated as first line for lesion and extensional assessment. However, MRI provides a more accurate mapping of bone marrow involvement and local extensional assessment. Bone scintigraphy is of interest in multifocal lymphoma, where the proximal

femoral and distal tibial involvement and the cranial involvement argue in favour of the primary origin. Positron emission tomography (PET) marked with 18fluorodeoxyglucose (18FDG) has also shown its usefulness in recent years both diagnostically and prognostically. It thus makes it possible to modify the therapeutic strategy during follow-up [10].

Histological evidence is required to confirm the diagnosis, determine the histological type and predict prognosis. Pathologists from the Bone Tumour Reference Network, according to their latest publication in 2018 [11], recommend a radio-guided percutaneous biopsy. Diffuse large-cell diffuse B lymphoma is the most common histological type [12] with its different subtypes determined by immunophenotyping expressing different markers (CD20) that are the target of new therapies, as described in the latest revision of the WHO classification of lymphoid haemopathies [13].

Treatment is age-dependent and depends on the international prognostic index [12-14]. It is based on the combination of multidrug therapy with an anti-CD20 (Rituximab). For localized forms with signs of compression, radiotherapy may be used. It is reserved for young patients and it is associated with a poor prognosis and a very high risk of relapse. Other regimens and molecules are being investigated [12].

Conclusion

Primary bone lymphoma is a rare entity. We reported an observation of multifocal primary bone lymphoma complicated by bone marrow compression in a 70-year-old man. Non-specific clinical signs delayed the diagnosis. Histological evidence is the cornerstone of diagnosis in order to start an appropriate therapy.

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