Primary iliac bone lymphoma with multiple bone involvement evaluated by 18 fluorodeoxyglucose positron emission tomography: A case report

Lymphome primitif de l'os iliaque avec atteinte osseuse multiple évalué par tomographie par émission monophotonique au 18 fluorodésoxyglucose : à propos d'un cas

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Abstract

Introduction: Involvement of bone by lymphoma usually occurs in the setting of widespread systemic disease. The primary bone lymphoma (PBL) is a rare entity, accounting for 3 to 7% of primary malignant bone tumors and less than 2% of all lymphomas. The Multifocal forms are even rarer and can be confused with bone metastases. The diagnosis is guided by the clinic and radiology, but only the histology confirms accurate diagnosis.

Observation: We report a rare case of primitive iliac lymphoma in a 58-year-old man. The pathological study was in favor of a diffuse large B-cell lymphoma. An 18 fluorodesoxyglucose (FDG) positron emission tomography computed tomography (PET/CT) performed as part of the staging assessment revealed multifocal bone involvement.

Conclusion: This case highlights the importance to evoke the diagnosis of primary bone lymphoma, even if it presents in an unusual site, as well as the value of metabolic imaging in assessing the extent of the disease.

Keywords: iliac tumor, diffuse large B-cell lymphoma, 18 FDG PET/CT

Résumé

Introduction : L'atteinte osseuse par le lymphome survient généralement dans le cadre d'une maladie systémique généralisée. L'atteinte lymphomateuse primitive de l'os est une entité rare qui représente 3 à 7% des tumeurs osseuses malignes primitives et moins de 2% de l'ensemble des lymphomes. Les formes multifocales sont encore plus rares et peuvent se confondre avec des métastases osseuses. Le diagnostic positif est orienté par la clinique et la radiologie, mais seule l'histologie le confirme.

Observation : Nous rapportons un cas rare d'un lymphome iliaque primitif chez un homme âgé de 58 ans. L'étude histologique a confirmé le diagnostic d'un lymphome B diffus à grandes cellules. Une tomographie par émission monophotonique couplée à la tomodensitométrie (TEP/TDM) au 18 fluorodésoxyglucose (FDG) pratiquée dans le cadre du bilan d'extension a mis en évidence une atteinte osseuse multifocale.

Conclusion : Ce cas met en évidence l'importance de penser au diagnostic du lymphome osseux primitif, même s'il se présente dans un site inhabituel, ainsi que la valeur de l'imagerie métabolique pour évaluer l'étendue de la maladie.

Mots clés : tumeur iliaque, lymphome B diffus à grandes cellules, TEP/TDM au 18 FDG

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LA TUNISIE MEDICALE - 2022 ; Vol 100 (11) : 804 - 808

INTRODUCTION

Primary bone lymphoma (PBL) is rare, it is characterized by extranodal involvement that can concern bone as well as muscle and subcutaneous tissue (1). Histological confirmation is essential to distinguish it from sarcomatous involvement whose therapeutic management differs considerably (2). Both morphological and functional imaging, such as the 18 fluorodesoxyglucose (FDG) positron emission tomography computed tomography PET/CT, plays an essential role in the different steps of the management of this pathology (3). We present a case of PBL in a relatively rare location.

OBSERVATION

This case report was reported according to the CARE guidelines (4). The authors certify that they have obtained patient's informed consent. A 58-year-old man presented to the orthopaedics department with a 1-year history of progressively worsening pain in the left hip and hemi-pelvis. The pain was resistant to symptomatic treatment. He had a history of total hip arthroplasty for dysplastic coxarthrosis.

Pelvic X-ray was normal and thoraco-abdomino-pelvic computed tomography (CT) scan showed diffuse and heterogeneous sclerotic lesion in the left iliac bone with soft tissue component measuring 96x80 mm extending into ipsilateral iliopsoas muscle. CT-guided bone biopsy of the left iliac tumor mass was performed and the diagnosis of diffuse large B-cell lymphoma was retained. 18 FDG PET-CT was then requested for staging.Imaging was performed 60 minutes after the injection of 206,8 Megabecquerel (MBq) of 18 FDG.

Maximum intensity projection (MIP) images revealed intense tracer uptake in the left hemipelvis and focal uptake in vertebra, right rib, right scapula and sternum (Figure 1). No lymph node involvement was detected.

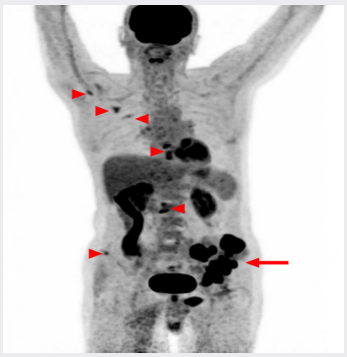

Figure 1. 18 fluorodesoxyglucose (FDG) positron emission tomography computed tomography (PET/CT): Maximum intensity projection (MIP) image showing intense tracer uptake in left iliac bone (red arrow) with several other focal uptake in the axial skeleton (arrow head)

Axial CT, PET and fused images showed mixed bone lesions and intense uptake with a maximum standardized value (SUVmax) of 23.6 at the primary lesion involving the left iliac wing, the acetabulum, and the ipsilateral ischiopubic ramus with a soft tissue mass infiltrating the iliacus, gluteus medius, and obturator externus left muscles (Figure 2). Examination revealed other bone sites affected L2, L5, the spine process and the medial border of the right scapula, the sternal body and the posterior segment of the 5th right rib. In the absence of any other musculoskeletal site of malignancy identified, the diagnosis of PBL was made.

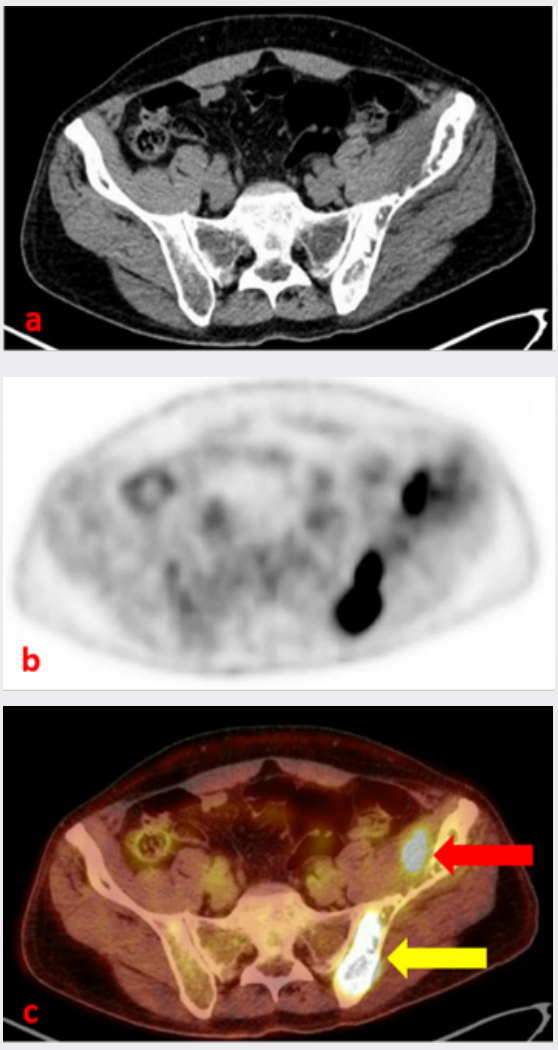

Figure 2. 18 fluorodesoxyglucose (FDG) positron emission tomography computed tomography (PET/CT): axial slices (a: computed tomography (CT) image, b: positron emission tomography (PET) image, c: fusion image). The images show intense uptake of the radiotracer in the iliac bone lesion, maximum standardized uptake value (SUVmax): 23.6 (yellow arrow) with increased metabolic activity in iliacus muscle (red arrow)

DISCUSSION

18 FDG PET/CT combining the sensitivity of metabolic imaging with the specificity of morphologic imaging is a useful method of evaluating primary bone tumours during initial staging (5). PBL is rare representing 3 to 7% of malignant bone tumors (1), less than 2% of all adult lymphomas (6) and 3 to 5 % of all extranodal non-Hodgkin's lymphomas (7). Two forms of bone lymphoma are identified: PBL and secondary bone lymphoma (SBL) (8). SBL is more common, it is a disseminated lymphoma with concomitant involvement of the bone skeleton (9).

The World Health Organization, defines PBL as a neoplasia producing one or more masses in the bone, without associated visceral or lymph node disease (10).

Non-Hodgkin's lymphoma constitutes the majority of PBLs. Diffuse large B-cell lymphoma (DLBCL) is the most common form in 80% of cases (11,12). T-cell lymphomas accounts for approximately 1 to 5% of cases (11,12). Mean age at diagnosis ranges from 40 to 60 years, with clear man predominance 8:1 ratio (11–13). Involvement is mainly unifocal, more rarely multifocal (11). PBL is commonly found in appendicular skeleton, typically affecting the diaphyseal region of the long bones (71%) in particular of the femur, tibia and humerus. In order of decreasing frequency, axial skeleton involvement is seen within the spine, the iliac bone as in our patient, the sternum and the ribs (9,13). Lymphomatous involvement of soft tissue and in particular skeletal muscle is also rare. It has been reported in 1.4% of all musculoskeletal lymphomas. The most common location is the thigh and arm muscles. Pelvis and calf muscles can also be affected (14).

Clinical presentation of PBL is comprised of a wide variety of mostly non-specific symptoms. That is the case with our patient who did not present any clinical signs that could suggest a diagnosis of bone lymphoma. Careful physical examination and imaging studies did not reveal associated hepatomegaly, splenomegaly, or adenomegaly (15).

Indeed, the diagnosis of primary bone lymphoma is often difficult due to the lack of specific clinical and radiological signs (15).

Patient generally reports the notion of localized bone pain which did not reacted favourably to medical treatment or intermittent persisting pain for several months as in our patient. Swelling or pathological fracture can also occur. General signs such as fever or weight loss can be observed (15,16). Spinal injuries can lead to neurological symptoms due to paraspinal extension (17).

The clinical and radiological suspicion of PBL requires the performance of a bone biopsy with anatomopathological, immunohistochemical and molecular study in order to confirm the exact type of the lymphoma (9).

The radiological appearance of primary musculoskeletal lymphoma is usually aggressive bone destruction observed in 70% of cases (13). A mixed aspect is apparent in 28% of cases, and the sclerotic aspect predominates in only 2% of patients (13).

Assessment must be performed to eliminate lymph nodes or visceral localizations and thus confirm the diagnosis of PBL, and also to look for a multifocality and an extension to the soft tissues which are elements of poor prognosis (18).

18 FDG PET/CT is one of the gold standard examinations for staging non-Hodgkin's lymphoma and exploring nodal and extranodal lymphomatous involvement by revealing increased radionuclide activity in affected areas (19). Our patient was classified according to Ann Arbor classification as stage IVE (defined by polyostotic lymphoma) with the involvement of multiple bones without lymph node or visceral disease. Isolated PBL fell into stage IE (defined by single bone lesion) (18). 18 FDG PET/CT is also very valuable in assessing early disease response to therapy, and plays a pivotal role in the detection of recurrent disease, the assessment of residual post-therapy lesion, as well as the detection of relapses (20). An advantage of 18 FDG PET/CT is its ability to detect additional bone lesions with the whole-body acquisition protocol (19). and its combination of anatomical structure and functional metabolism which reflects various FDG uptakes at different stages of malignancy (21), 18 FDG PET/CT helps also to avoid misdiagnosis such as metastatic disease, chondroma or sarcoma suspected primarily in the young subject. Indeed interpretation of the CT part of the 18 FDG PET/CT study and the correlation between FDG uptake and the nature of bone tumors can help distinguish benign and malignant tumor (22). Aggressive bone lesions show a higher FDG uptake than benign ones (range between 3.2 and 6.9 in malignant and 0.7 and 1.35 in benign) (23). 18 FDG PET/CT can help guide the biopsy to the most aggressive tumor component. This information can significantly change the treatment and follow-up.

Although they are the standard imaging modalities for PBL, using only CT and MRI imaging criteria for diagnosis and staging purposes is not sufficient (24). Moog et al. showed that PET/CT has a higher sensitivity and specificity than bone scan for identifying lymphomatous infiltration of skeleton (20). In addition, 18 FDG PET/CT has demonstrated a better diagnostic yield in extra-nodal lymphoma with 97% of sensitivity, 100% of specificity and 98% of accuracy versus respectively 87%, 85% and 84% for CT scan (25).

Many series concluded that FDG PET/CT is a valuable imaging tool for assessing treatment response for patients with PBL (6,7,26,27). In contrast to morphologic techniques which can only show bone changes from lytic to sclerotic after treatment 18 FDG PET/CT shows metabolic response which can be semi-quantified by mesearing SUVmax (28).

And in case of complete response, PET/CT shows no hypermetabolic lesion, with a steep decline in FDG uptake often returning to normal, making this examination very reliable with an excellent negative predictive value (29). Liu used an SUVmax of 2.5 as a cut off value to separate residual lymphoma from metabolically inactive disease (3).

CONCLUSION

PBL is a rare extranodal lymphoma that usually has an excellent prognosis. The assessment of primary bone lymphoma is similar to that of other lymphomas. This case emphasizes the importance of 18 FDG PET/CT in the management of the PBL. It is of great help in distinguishing between PBL and SBL, which have different management and prognosis. It is also provides opportunity to reflect on the importance of initial metabolic imaging in evaluating multiplicity, disease extent. This imaging will be also a baseline review to evaluate treatment response. PBL pathogenesis lack in this study and large immunohistochemical and molecular studies have to be conducted to know more it.

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