Incidental Findings of Acute Myeloid Leukemia in Sjögren's Syndrome Detected by Nuclear Medicine Techniques

Abstract

A 46-year-old female with Sjögren's syndrome previously treated with corticosteroids was referred to our department for suspicious humeral head osteonecrosis. Dual-phase bone scan showed an increased radiotracer distribution in the head of the left humerus. Nevertheless, whole-body scan revealed multiple sites of heterogeneous skeletal uptake. As lymphoproliferative disorder was hypothesized, also based on laboratory examination, F-18 fluorodeoxyglucose positron-emission tomography/computed tomography was performed and showed increased uptake in several osseous structures and in the subcutaneous nodules. Finally, bone marrow biopsy confirmed the diagnosis of acute myeloid leukemia as well as skin nodule biopsy revealed infiltration by malignant cells.

Keywords: *Acute myeloid leukemia, bone scintigraphy, F-18 fluorodeoxyglucose positron-emission tomography/computed tomography, Sjögren's syndrome*

Angelo Castello, Matteo Caracciolo, Luca Urso, Naima Ortolan, Alberto Nieri, Stefano Panareo, Mirco Bartolomei

Department of Oncological Medical and Specialists, Nuclear Medicine Unit, University Hospital of Ferrara, Ferrara, Italy

Sjögren's syndrome is one of the most common chronic inflammatory autoimmune diseases primarily involving the exocrine glands, which affects mostly females in the fourth decade of life.[1] A 46-year-old female with Sjögren's syndrome previously treated with corticosteroids underwent bone scintigraphy for suspicious humeral osteonecrosis. Avascular osteonecrosis is a common complication observed in autoimmune disease patients after corticosteroid therapy.[2] Blood pool images [Figure 1a] showed slight increased distribution of technetium 99m-hydroxydiphosphonate in the head of the left humerus. Delayed images [Figure 1b], performed 3 h after the radiotracer administration, confirmed intense uptake of the left humerus, and suggestive for osteonecrosis. Nevertheless, images also revealed an ill-defined uptake in the humeral diaphysis, ribs, and spine. Whole-body bone scan [Figure 1c] showed multiple sites of heterogeneous radiotracer distribution throughout the skeletal, especially involving the skull, right sacroiliac region, left iliac wing, left femur, and tibia diaphysis.

Therefore, a lymphoproliferative disorder was suspected, also based on high levels of lactate dehydrogenase (2193 U/l), C-reactive protein (670 mg/L), erythrocyte sedimentation rate (97 mm/h), as well as white blood cells (16,83 × 10^3/µl) and increased number of immature elements of the myeloid series in the peripheral blood. Platelets (218 × 10^3/µl) and hemoglobin (11.6 g/dl) were within normality ranges. Indeed, Sjögren's syndrome can be associated with tumors, mostly lymphoma and occasionally T-cell leukemia/lymphoma, although the exact pathogenesis is still under debate.[2,3] F-18 fluorodeoxyglucose positron-emission tomography/computed tomography (18F-FDG PET/CT) has an established role in the diagnostic algorithm of patients with suspicious lymphoproliferative disorders, as well as in in detecting uncommon disease localizations and in response to treatment assessment.[4,5]

Thus, 18F-FDG PET/CT was performed. Maximum intensity projection image [Figure 1d] and axial fused 18F-FDG PET/CT [Figure 1e-h] confirmed the humeral osteonecrosis (black arrow) and diffusely increased and heterogeneous bone marrow activity (red square brackets and red arrows). In addition, 18F-FDG-avid soft tissue nodules were detected in the right scapular region, right axilla, and right thigh (arrowheads). Finally, the results of

How to cite this article: Castello A, Caracciolo M, Urso L, Ortolan N, Nieri A, Panareo S, *et al.* Incidental findings of acute myeloid leukemia in Sjögren's syndrome detected by nuclear medicine techniques. Indian J Nucl Med 2022;37:279-80.

Address for correspondence:
Dr. Luca Urso, Department of Oncological Medical and Specialists, Nuclear Medicine Unit, University Hospital of Ferrara, Via Aldo Moro 8, CAP 44124, Ferrara, Italy.
E-mail: rsulcu@unife.it

Received: 12-08-2021
Revised: 24-11-2021
Accepted: 30-11-2021
Published: 02-11-2022

Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.ijnm_126_21

Quick Response Code:

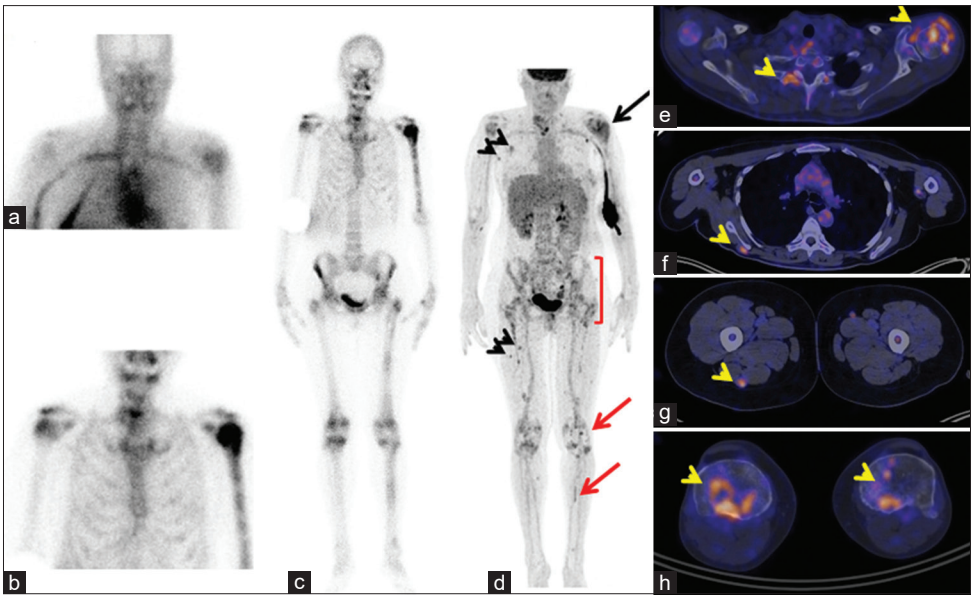

Figure 1: dual-phase bone scan (1a: blood pool images; 1b delayed images; 1c total body delayed images) showed increased distribution of 99m-hydroxydiphosphonate in the head of the left homerus and multiple sites of heterogeneous uptake throughout the skeletal. 18F-FDG PET/CT (1d: maximum intensity projection; 1e-h: axial fused 18F-FDG PET/CT) confirmed multiple sites of uptake, subsequently revealed to be localizations of acute myeloid leukemia

bone marrow biopsy demonstrated acute myeloid leukemia, a hematologic malignancy characterized by the clonal expansion of myeloid blasts in the peripheral blood and bone marrow. Likewise, skin nodule biopsy confirmed infiltration by malignant cells.

This case describes a rare acute myeloid leukemia in Sjögren's syndrome with associated humeral osteonecrosis, based on two-phase bone scan and 18F-FDG PET/CT findings. Although in our case, we did not completely discriminate osteonecrosis whether secondary to corticosteroids treatment or to leukemia, progressive skeletal symptoms in young patients may hide hematologic disease; therefore, a critical review of bone scan in such cases may help in the early settlement of diagnosis.[6,7] Moreover, our case highlights that nuclear medicine could have an important role in identifying possible localizations of malignant disease in Sjögren's syndrome, which may drive biopsy and subsequent clinical management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Vitali C, Bombardieri S, Jonsson R, Moutsopoulos HM, Alexander EL, Carsons SE, *et al.* Classification criteria for Sjögren's syndrome: A revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis 2002;61:554-8.
2. Liao MT, Chien WC, Wang JC, Chung CH, Chu SJ, Tsai SH. Increased risk of bisphosphonate-related osteonecrosis of the jaw in patients with Sjögren's syndrome: Nationwide population-based cohort study. BMJ Open 2019;9:e024655.
3. Voulgarelis M, Skopouli FN. Clinical, immunologic, and molecular factors predicting lymphoma development in Sjogren's syndrome patients. Clin Rev Allergy Immunol 2007;32:265-74.
4. Cheson BD, Fisher RI, Barrington SF, Cavalli F, Schwartz LH, Zucca E, *et al.* Recommendations for initial evaluation, staging, and response assessment of hodgkin and non-hodgkin lymphoma: The Lugano classification. J Clin Oncol 2014;32:3059-68.
5. Panareo S, Urso L, Santi I, Rigolin GM, Cuneo A, Cittanti C, *et al.* Right atrium mass assessed with 18F-FDG PET/CT scan turns out to be an uncommon relapse of testicular diffuse large B-cell lymphoma: A case report. Diagnostics (Basel) 2020;10:987.
6. Feng H, Qiao J, Ding N, Chen W, Qi K, Pan X, *et al.* Sjögren's syndrome complicated by myeloid/natural killer cell precursor acute leukemia: Case report and review of the literature. Case Rep Hematol 2016;2016:8261249.
7. Solav SV, Bhandari R, Solav P. Skeletal scintigraphy manifestations of hematologic disorders. Indian J Nucl Med 2012;27:59-62.