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Histiocytosis with Pulmonary Involvement Mimicking Rheumatoid Arthritis

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Abstract In Langerhans cell histiocytosis pulmonary involvement is often isolated. It is rarely a part of a multifocal form. Tenosynovitis is exceptional. Hand bones involvement is atypical. We report the case of a 75-year-old, non-smoker man, without medical history, who presented with arthritis and fever. Examination found arthritis affecting large and small joints. In biology, there was an inflammatory syndrome. Radiographs of the hands showed erosions of the carpal bones. Doppler ultrasonography of the hands showed active synovitis. Tc99 bone scintigraphy highlighted joint uptake in the hands, knees, spine and right upper jaw. Blood cultures and infectious investigations were negative. Anti-nuclear antibodies and rheumatoid factor were negatives. CT scan showed cystic lung lesions and nodular aspect. Bronchoalveolar lavage fluid revealed positivity of anti-CD1a at 6%. A surgical biopsy showed an infiltrate of eosinophilic cells, positive for CD1a in immunohistochemistry studies. The diagnosis of Langerhans cell histiocytosis was retained.

Keywords: *langerhans cell histiocytosis, lung, arthritis, synovitis, bronchoalveolar lavage*

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1. Introduction

Langerhans cell histiocytosis is a rare disease, mainly affecting children, which consists in various clinical manifestations from a single lytic bone lesion to multisystemic lesions with organ dysfunction [1]. Tenosynovitis is exceptional. Hand bones involvement is atypical [2,3]. Pulmonary involvement is often isolated. It is rarely a part of a multifocal form [4,5]. We showcase in this paper an exceptional case of histiocytosis with pulmonary involvement mimicking rheumatoid arthritis.

2. Case Report

A 75 year-old non smoker man, with no past medical history, was admitted for febrile polyarthritis. He didn't complain of cough or dyspnea.

Physical examination revealed fever at 38.5°C, bilateral and symmetric polyarthritis of the small and large joints (elbows, wrists, knees, metacarpophalangeal joints, proximal and distal interphalangeal joints).

Lab tests showed an increase in the inflammatory markers without hyperleukocytosis. The erythrocyte sedimentation rate was 86 mm/h and C reactive protein was 22 mg/l.

Radiographs of the hands showed multiple erosive changes in carpal bones (Figure 1), narrowing of the metacarpophalangeal and the medial femorotibial joints spaces with osteosclerosis of the subchondral bony plates of the correspondant tibial plateau.

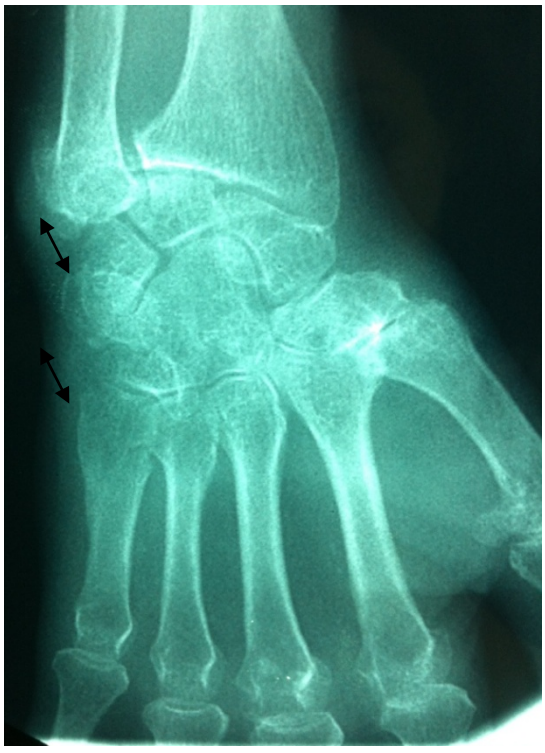

Figure 1. X-Ray of the hand showing erosions of the carpal bones.

Doppler ultrasound of the hands revealed bilateral synovitis mainly of the metacarpophalangeal joints, the distal interphalangeal joint of the third ray of the right hand (Figure 2), the right radiocarpal joint and the sixth compartment of the right wrist.

Blood cultures, HIV, Lyme and Whipple's serologies were negative. There were no arguments for tuberculosis. Antinuclear antibodies, rheumatoid factor and anti-cyclic citrullinated peptide (anti-ccp) antibodies were negative. Giant cell arteritis was ruled out. Transthoracic echocardiography was normal.

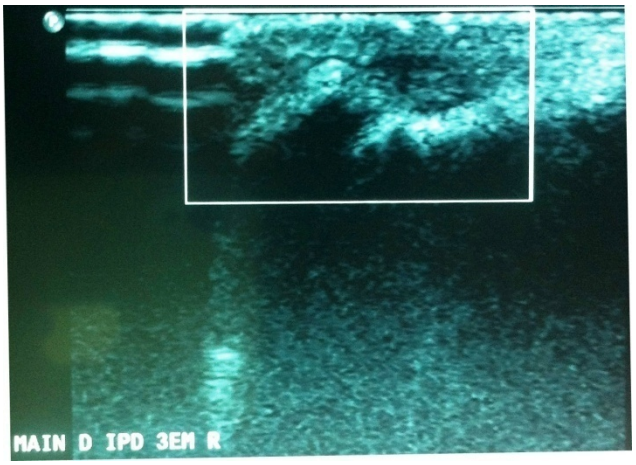

Figure 2. Doppler ultrasonography of the right hand showing active synovitis

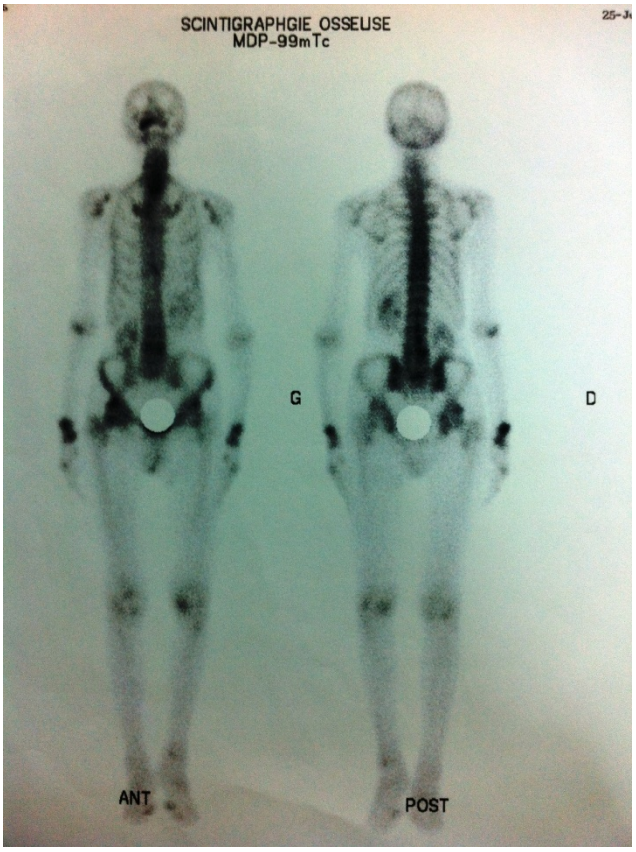

Figure 3. Tc99 bone scan Scintigraphy showing increased uptake on the hands, knees, spine and upper right jaw

Technetium 99 bone scintigraphy showed articular uptake in the hands, knees, spine and right upper maxilla (Figure3).

Chest computed tomography (CT) revealed bilateral, diffuse, infracentimetric, cystic, parenchymal pulmonary lesions (Figure 4). Some nodules had a holey appearance (Figure 5).

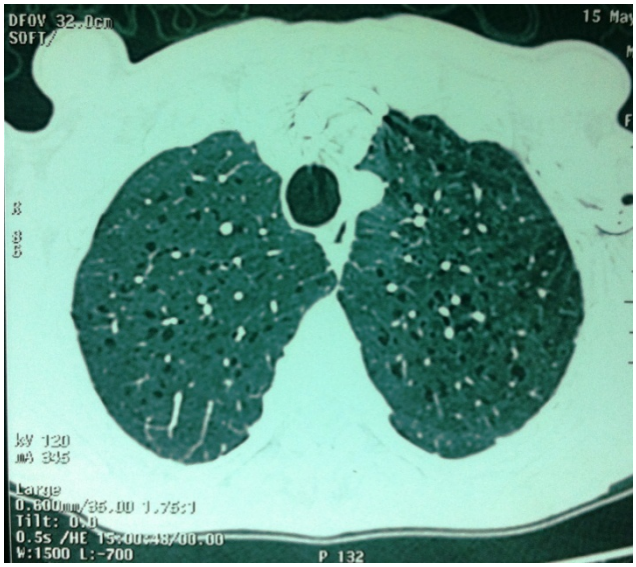

Figure 4. CT scan showing cystic lung lesions

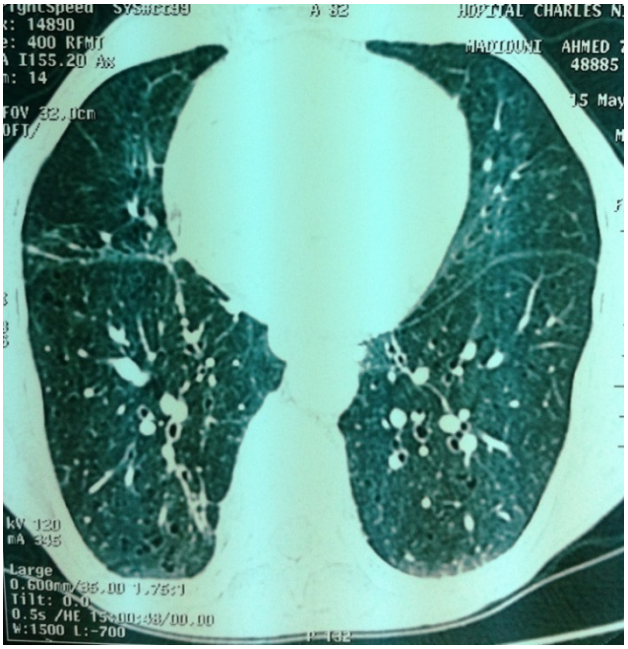

Figure 5. CT scan showing nodular lung lesions

Bronchoalveolar lavage evidenced an alveolitis with a predominance of macrophages and lymphocytes and a positivity of the anti CD1a at 6%. Abdominopelvic CT showed no abnormalities.

Lung biopsy was redoubted because of the high risk of pneumothorax. A synovial knee surgical biopsy was chosen because of local inflammation, uptake in scintigraphy and accessibility for orthopedists. This biopsy showed an infiltrate of eosinophilic cells, positive for CD1a in immunohistochemistry studies.

Based on these findings the diagnosis of Langerhans cell histiocytosis (LCH) was retained.

The patient received glucocorticoid therapy combined with vinblastine followed by etoposide. However, he presented severe sepsis after pulmonary infection and died one year after diagnosis.

3. Discussion

Langerhans cell histiocytosis (LCH) is a rare multisystemic disease, with several clinical presentations varying from an isolated osseous involvement to a multi-organ involvement with a severe form. It affects mainly the young adult and the child [1]. Elderly are rarely affected.

In LCH, osteoarticular involvement is typically presented by bone lesions of the skull, mandible, spine, pelvic and long bone diaphyses [1]. Hands involvement with tenosynovitis is exceptional [2,3]. In our patient, a reactionnal synovitis due to an associated disease was ruled out. Infections, autoimmune diseases mainly rheumatoid arthritis were eliminated. A paraneoplastic syndrome was improbable.

In adults, pulmonary involvement is classically isolated. The main epidemiological feature of pulmonary LCH in adults is its occurrence almost exclusively in smokers, with often important daily smoking [4,6].

In half the cases, pulmonary involvement is revealed by non specific respiratory signs in smokers, dominated by non-productive cough, sometimes associated with exertional dyspnea. Pulmonary involvement is rarely part of a multifocal form [4,5,6]. Our patient had no history of smoking and no respiratory signs.

In pulmonary histiocytosis, chest CT shows typically the association of nodules with irregular borders, cavitary nodules, thick-walled and thin walled cysts [4], as was the case in this observation. In LCH with lung involvement, the discriminating threshold of 5% of CD1a + cells is retained . A value of 5 % is highly suggestive of LCH, but occurs only rarely (less than 25% of cases in some series). False positive with an accumulation of Langerhans cells CD1a + with rates of 2 to 3 % can be observed in the cells of control smokers subjects with no pulmonary diseases [7]. In our patient, bronchoalveolar lavage revealed the presence of Langerhans cells CD1a at 6%, despite the absence of smoking. Lung biopsy was redoubted and not done because of the high risk of pneumothorax [8].

4. Conclusion

Our observation is particular by the occurrence of LCH in a 75 year patient, with atypical revealing symptomatology made of febrile arthritis.

On the other hand, pulmonary involvement in multifocal histiocytosis form, in a non-smoking patient, is extremely rare.

References

[1] Arico M, Egeler RM. Clinical aspects of Langerhans cell histiocytosis. Hematol Oncol Clin North Am 1998;12: 247-58.

[2] Glajchen N, Springfield DA, Lipton JM. Langerhans' cell histiocytosis of the synovium. Am J Roentgenol 1997;169:1751-2.

[3] Santilli D, Lo Monaco A, Cavazzini PL, Trotta F. Multicentric reticulohistiocytosis: a rare cause of erosive arthropathy of the distal interphalangeal finger joints. Ann Rheum Dis 2002;61(6):485-7.

[4] Tazi A. Adult pulmonary Langerhans' cell histiocytosis. Eur Respir J 2006;27:1272-85.

[5] Harpreet S Suri, Eunhee S Yi, Gregorz S Nowakowski, Robert Vassallo. Pulmonary langerhans cell histiocytosis. Orphanet J Rare Dis 2012;7: 16.

[6] Cheng-Wei Li, Man-Hui Li, Jiang-Xiong Li, Ru-Jia Tao, Jin-Fu Xu, Wei-Jun Cao. Pulmonary Langerhans cell histiocytosis: analysis of 14 patients and literature review. J Thorac Dis. 2016;8: 1283-9.

[7] Casolaro MA, Bernaudin JF, Saltini C, Ferrans VJ, Crystal RG. Accumulation of Langerhans'cells on the epithelial surface of the lower respiratory tract in normal subjects in association with cigarette smoking. Am Rev Respir Dis 1988;137:406-11.

[8] Mendez JL, Nadrous HF, Vassalo R, Decker PA, Ryu JH. Pneumothorax in pulmonary Langerhans' cell histiocytosis. Chest 2004;125:1028-32.