CASE REPORT

Mycosis Fungoides-Spectrum of Manifestations

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Abstract

Mycosis fungoides is an unusual form of cutaneous T cell lymphoma that is diagnosed by skin biopsy after years of non specific skin eruptions. In addition to skin lesions extracutaneous manifestations can also occur.

Key Words

Mycosis Fungoides, X-ray, CT, HRCT, Lymphoma.

Introduction

The doctor who first documented the disease mycosis fungoides in a patient was a French doctor named Jean Luis Marc Alibert. He called it mycosis fungoides because of the similarity of the spots on the skin to raised mushrooms. The very first case of the disease was documented in 1806 and the name that Dr. Alibert gave it stuck as mycosis fungoides. Mycosis Fungoides represents a relatively small proportion of lymphoma. According to a study group incidence of mycosis fungoides was reported to be 0.36/10 person years in 1973-1992 in United states (1). It inolves various organs including sinonasal cavity, airway, intestinal tract, skin, lymphnode, liver, lung and musculoskeletal system. Radiological demonstration of disease progression beyond the primary site is clinically important because systemic dissemination in most of the entities leads to a dramatic change in prognosis and treatment (2).

Case Report

A 80 years old male patient presented with a history of diffuse asymmetrical erythematous plaques in skin of chest, abdomen and thigh for last 10 years which were unresponsive to treatment .There was increase in severity of lesions in the form of formation of nodules .Also patient complained of progressive dyspnoea and pain ,swelling with restricted movement of the hands and feet for the last 1 year. Haematological investigations revealed anaemia and leucocytosis. Biopsy of skin lesion was consistent with the diagnosis of Mycosis fungoides. Radiological investigations like X-ray chest, X-ray of musculoskeleton system, CT abdomen and HRCT chest was done to determine the systemic involvement. Chest X-ray revealed prominent interstitial markings in lower lung fields. HRCT Chest findings revealed small nodules distributed in left upper zone *Fig.1(A)* along with cylindrical bronchiectasis involving the right lung with septal lines in lower lung fields *Fig.1(B)* .

CT abdomen revealed liver involvement with presence of hypodense non enhancing lesions in liver (*Fig.2*). Skeleton involvement was seen in the form of well defined areas of destruction in the medullary region of phalanges of hand (*Fig. 3*) and foot (*Fig.4*).

Discussion

Cutaneous T-Cell lymphoma include a whole spectrum of clinical and pathological manifestations, most common disease form is mycosis fungoides manifesting as a dermal eruption. Initially characteristic infiltrates in skin are seen as plaques and can mimic psoriasis. This limited stage1 can last for years but eventually progressing to skin nodules and tumours. Later on lymphadenopathy and visceral involvement may occur (3).Bony lesions in these patients are peripheral, affecting the long bones and sparing the axial skeleton. There are areas of discrete medullary destruction, many of which have permeative pattern. Cortical destruction originates in the endocortex and extends to exocortex. However changes are not specific (4). Primary musculoskeletal lymphoma is rare but can occur in bone (reticulum cell sarcoma) or in the skin and subcutaneous tissues (mycosis fungoides). Secondary involvement in the musculoskeletal system is more common and can have a variety of radiologic findings.

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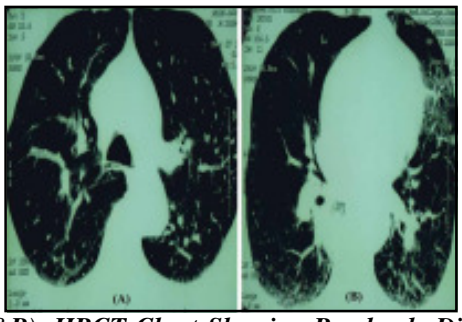

Fig1(A&B). HRCT Chest Showing Randomly Distributed Nodules in the Left Upper Lung and with Bilateral Septal Thickening in Bilateral Lower Lung Fields

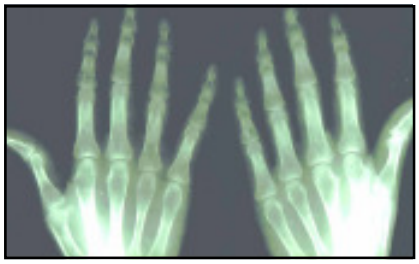

Fig 3. X Ray AP View Showing Well Defined Lytic Lesion in Phalanges of Both Hand

The definitive diagnosis of musculoskeletal lymphoma, however, is difficult to make by using imaging criteria alone. Primary and secondary bone lymphoma can be indistinguishable radiologically and histologically (5).

The HRCT chest in cases of mycosis fungoides showed multiple, dense, peribronchovascular nodules with surrounding ground-glass opacity and several wedge-shape peripheral opacities. The autopsy specimen revealed angiocentric and peribronchovascular involvement of mycosis fungoides and pulmonary infarctions distal to angiocentric infiltration of the tumor cells (6). Probability of developing extracutaneous disease correlates strongly with the extent of skin involvement at presentation (T classification). Those patients with limited patch or plaque lesions are unlikely to progress to extracutaneous disease when they are treated for their skin disease, whereas patients with generalized lesions are more likely , and patients with tumorous or erythrodermic involvement are most likely (7). Mycosis fungoides in a limited form has similar survival rates as normal population. CT for staging and follow up of lymphadenopathy and visceral involvement is clearly indicated in patients with advanced disease (8).

Conclusion

In mycosis fungoides cutaneous as well as extracutaneous manifestations can occur. Although diagnosis is based on clinical and histopathological findings ,it is important for the radiologist to be aware of the whole spectrum of disease since imaging plays a crucial role in prognosis and management.

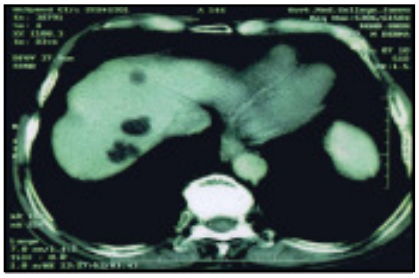

Fig 2.CT Abdomen Showing Multiple Hypodense Non Enhancing Lesions in Right Lobe of Liver in Case of Mycosis Fungoides.

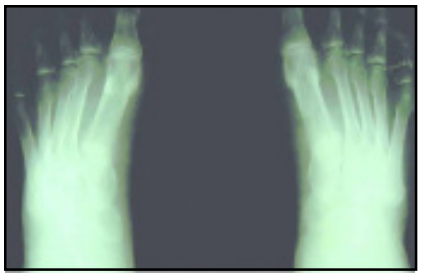

Fig 4. X Ray AP View Foot Showing Lytic in Head of First Metatarsal of Right Foot

References

1. Santucci M,Biggeri A,Feller AC, *et al*.Efficacy of histologic criteria for diagnosing early mycosis fungoides.An EORTC cutaneous lymphoma study group investigation. *Am J Surg Pathol* 2000;24:40-50.
2. Lee HJ, Im JG, Goo JH, *et al*. Peripheral T cell lymphoma :Spectrum of Imaging findings with clinical and Pathologic features. *Radiographics* 2003; 23:7-28.
3. Miketic LM, Chambers TP, Lembersky BC. Cutaneous T Cell Lymphoma : Value of CT in. imaging and determining prognosis .*AJR* 1993;160: 1129 -32.
4. O Reilly GV, C lark TM, Crum CP. Skeletal involvement in mycosis fungoides. *AJR Am J Roentgenol* 1977 ;129(4) : 791-93.
5. Mallory PC, Fishman EK Magid D Russel H. Lymphoma of bone muscle and skin:CT findings. *AJR Am J Roentgenol* 1992;159 (4): 805-09.
6. Ueda T, Hosoki N, Isobe K, Yamamoto S, Matoori K, Shinkai H. Diffuse pulmonary involvement in mycosis fungoides. High resolution computed tomography and pathologic findings. *J thoracic Imaging* 2002; 17(2) :157-59.
7. De Coninck EC ,Kim YH, Varghese A, Hoppe RT. Clinical Characteristics and Outcome of Patients With Extracutaneous Mycosis Fungoides . J Clin Oncol 2001; 19(3): 779-84.
8. Levy AD, Koeller KK. Invited Commentary. *Radiographics* 2003; 23:26-28.