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Unusual Presentation of Anterior Mediastinal Mass

Srilata Puru Naik*, Jayaraj BS and Mahesh PA

Department of Respiratory Medicine, JSS Medical College & Hospital, Mysore, Karnataka, India

Introduction

Approximately half of all mediastinal lesions are asymptomatic and are detected on chest radiographs taken for unrelated reasons. The absence of symptoms suggests that a lesion is (maybe) benign, whereas the presence of symptoms suggests malignancy. In adults, 48-62% of lesions are symptomatic, whereas the percentage of symptomatic lesions is higher in children (58 to 78%). The most common symptoms are cardiorespiratory - in particular, chest pain and cough. Other manifestations are heaviness in the chest, dyspnea, signs of superior vena caval obstruction with facial swelling, and cyanosis. Recurrent respiratory infections are also common.

Cutaneous paraneoplastic manifestations in Hodgkin Lymphoma (HL) have been well described. These include eczema, prurigo, mycosis fungoidosis and erythema nodosum. Pruritus is a well-recognized presenting symptom of Hodgkin lymphoma [1,2].

We report a 28 year old lady who presented with pruritic skin lesions and was later diagnosed to have Hodgkin lymphoma. This serves to illustrate the point that intractable eczema or prurigo should warn the clinician a possibility of underlying sinister process.

Background

The clinical syndrome described by Thomas Hodgkins in 1832 is now taken to be confined to a particular histological type of lymphoma characterised by destruction of lymph node architecture, proliferation of large abnormal cells derived from monocytes and usually Reed Sternberg cells (RS Cells). Nodular sclerosing hodgkins lymphoma tends to be the more benign variant, the other types often progressing from lymphocyte predominant through mixed to lymphocyte depletion. All types of Hodgkins lymphoma may involve the lung, although the lymphocyte predominant does so less frequently than do the others.

Cutaneous Hodgkin lymphoma was first described by the German physician Grosz in 1906. This rare condition is thought to have decreasing incidence in recent decades, owing to the improved treatment modalities of patients with Hodgkins lymphoma. In addition to rarity, skin involvement carries an ominous prognosis but can be indolent if systemic disease was controlled properly. Cerroni et al. found that histology of cutaneous specific manifestations of Hodgkin lymphoma correlates with that of the nodal counterpart in most cases [3-5].

Case Report

A twenty eight year old lady presented with intractable pruritus with dryness followed by the appearance of skin lesions, multiple pigmented papules and macules over the extremities for six months. These lesions were initially noticed over the extremities and later spread proximally to involve her thighs and back. Examination revealed extensive lichenification, dryness and pigmentation over the macules. A clinical diagnosis of a prurigo nodularis was made [6,7].

She had no respiratory symptoms other than mild cough for two months. Though she did not complain any respiratory distress she had suprasternal in-drawing and use of accessory muscles. She had no history of orthopnea, night sweats, syncope or weight loss in the recent past. She had normal hematological profile, normal renal and liver function tests and normal ECG and 2D Echocardiogram. Routine chest radiograph showed scoliosis with mediastinum widening suggestive of a large anterior mediastinal mass.

CT Chest revealed evidence of a large lobulated enhancing lesion measuring about 14X14X10 cms in the anterior mediastinum extending along the right paratracheal region up to hila. The lesion was abutting the right cardiac border and encasing and compressing the superior venacava, right pulmonary artery, right pulmonary veins. There was partial collapse of the right upper lobe. The lesion was extending from the retrosternal space up to prevascular region to left side. With diagnostic possibilities of Lymphoma or Thymoma (Figures 1 and 2).

Skin biopsy showed pronounced orthokeratosis, papillomatosis and irregular acanthosis. Increased collagenisation of dermis with vertically oriented collagen bundles in the papillary dermis and no amyloid deposits. The features were consistent with Prurigo Nodularis.

Right anterior mediastinotomy and biopsy of lesion showed a tumour with cells arranged in lobular pattern separated by thin fibrovascular tissue. Cells were heterogeneous, composed of lymphoid cells, eosinoplils, histiocytes. Lacunar type of Reed-Sternberg's cells were seen. Occational typical Reed-Sternberg's cells also seen. Features were those of Hodgkin lymphoma – Nodular sclerosing type (Figure 3).

Patient showed good response to standard chemotherapy. (ABVD-Adriamycin [doxorubicin], bleomycin, vinblastine, dacarbazine regimen) (Figure 4).



Figure 1: Clinical pictures.

*Corresponding author: Srilata Puru Naik, Department of Respiratory Medicine, JSSMedicalCollege&Hospital,Mysore,Karnataka,India,Tel:919632785978;E-mail: srilatanaik1986@gmail.com

Received December 30, 2014; Accepted February 13, 2015; Published February 17, 2015

Citation: Naik SP, Jayaraj BS, Mahesh PA (2015) Unusual Presentation of Anterior Mediastinal Mass. J Pulm Respir Med 5: 247. doi:10.4172/2161-105X.1000247

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Figure 2: Chest X-ray and CT.



Figure 3: Histopathology of skin and mass.



Figure 4: Post-treatment scannogram.

Discussion

Pruritic skin lesions have been known to pre-date clinically evident B and T cell lymphomas by years. Pruritus and prurigo nodularis have been associated with Hodgkin Lymphoma. The mechanism of itch in malignancy is unclear and has been attributed to histamine release, tumour metabolites, immunological mechanisms and dry skin. The other paraneoplastic skin manifestations in Hodgkin Lymphoma are eczema, mycosis fungoides and erythema nodosum.

Pruritus continues to challenge physicians because of its nonspecific nature and the infrequency with which it is caused by Hodgkin's lymphoma. Clinicians, especially family practitioners, primary care internists, and dermatologists, need to remember Hodgkin's lymphoma as a possible cause of intractable itching. Finally, incidentally discovered anemia, thrombocytopenia, neutropenia, lymphopenia, hypoalbuminemia, or elevated erythrocyte sedimentation or similar

tests followed by an appropriate biopsy or performance of a bone marrow biopsy should provide the additional information necessary to pin down a diagnosis of Hodgkin's lymphoma, if present [8-10].
Conclusion
This case is being highlighted for its interesting presentation and to sensitize physicians to suspect an underlying sinister neoplastic presentation.

to sensitize physicians to suspect an underlying sinister neoplastic disease in patients presenting with chronic dermatosis. High degree of clinical suspicion is required as anterior mediastinal mass may have paucity of clinical symptoms at presentation. Early diagnosis may help in improving the prognosis.

findings encountered either incidentally or in the assessment of fatigue, unexplained weight loss, fever, night sweats, or other constitutional

symptoms may suggest the presence of Hodgkin's lymphoma. Imaging

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