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Pathology

A CASE OF LANGERHANS CELL HISTIOCYTOSIS PRESENTING AS A STERNAL MASS.

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ABSTRACT Langerhans cell histiocytosis (LCH) is a rare disease predominantly seen in young age group, and is a result of clonal proliferation of Langerhans cells. Here, we present a case of 26 years old female patient with weight loss, cough and a sternal mass of short duration. Clinical and laboratory evaluation for tuberculosis were negative. An initial cytologic evaluation was suggestive of LCH which was confirmed on further histopathological evaluation.

KEYWORDS : Langerhans cell Histiocytosis, CD1a, S100, cytology

INTRODUCTION

Langerhans cell histiocytosis (LCH), an uncommon disease predominantly affecting infants and young adults, results from the proliferation of a clonal population of Langerhans cell [1]. In normal physiology, Langerhans cells are specialised cells of dendritic origin seen in the skin [2]. Abnormal clonal proliferation of Langerhans cells; can show a wide spectrum of clinical presentation ranging from a self resolving illness to a progressive multisystem disorder [3]. The variable clinical presentation has resulted in identifying three different forms of Langerhans cell histiocytosis: Letterer-Siwe syndrome, Hand-Schuller-Christian syndrome, and eosinophilic granuloma (EG) [4]. In this case report, we discuss an atypical sternal presentation of LCH where diagnostic cytologic findings were helpful in arriving at an early conclusion.

CASE REPORT

A 26 years old female patient presented to the medical OPD with loss of appetite and cough of one month duration. At the time of presentation she was conscious and co-operative. On local examination a tender swelling identified at the right lower sternum, measuring 3 x 2cms in size; not fixed to overlying skin. General and systemic examination was within normal limits. Clinically, a possibility of Tuberculous lesion was considered. Laboratory investigations were ordered; and showed elevated Erythrocyte Sedimentation Rate. All other parameters were within normal limits. A plane and contrast helical CT scan showed moderately enhancing soft tissue destructive lesion involving lower sternal body and extending up to xiphysternum (figure1). A possibility of malignancy was suggested.

Subsequently, a Fine needle aspiration was done from the lesion. Cytological evaluation of the FNA sample revealed singly scattered and clusters of cells with moderate pale, eosinophilc cytoplasm and vesicular nuclei (figure2). Occasional cells showed nuclear grooves. The smear background showed scattered neutrophils and hemorrhage. Occasional ill defined collections of epithelioid histiocytes. Considering the age of the patient, clinical features of the lesion and cytological appearance on FNAC; a possibility of Histiocytic lesion was considered. Trucut biopsy was taken from the lesion, which showed nodular aggregates of plump histiocytes with well defined cell membrane, pale eosinophiilic cytoplasm and vesicular nuclei. Many nuclei showed grooving and occasional indentation (figure 3 & 4). Interspersed between the histiocytes were numerous neutrophils. Scant intervening stroma showed lymphocytes, neutrophils and ectatic vessels. With this morphology, a possibility of Langerhans cell Histiocytosis was considered; in view of which Immunohistochemical evaluation for CD1a and S100 was done. All the histiocytes showed membranous staining for CD1a antigen (figure 5), and diffuse staining for S100 antigen, which confirmed our diagnosis Langerhans cell Histiocytosis.

DISCUSSION

Langerhans cell Histiocytosis (LCH) is a disorder characterised by clonally proliferating dendritic cells, probably induced either by cytokines like tumor necrosis factor, interleukin 11 and leukemia inhibitory factor 1 or viral infections [5]. Although most patients presenting with LCH are children below 15 years of age; any age or gender can be affected. Lesions with single focus, and multifocal unisystem lesions tends to involve the bone commonly. Also, the bony lesions show an increased propensity to involve the skull, spine, pelvis and mandible. In the case of multifocal multisystem lesions, however; any organ can be involved including bone, skin, liver, hematopoietic system and lymph node [6].

Langerhans cell Histiocytosis (LCH) presenting as a solitary sternal mass is rare, with most patients presenting with pain and tenderness. In such cases the diagnosis, depends on clinical manifestation, cytology, histopathology and electron microscopy.

Appropriate clinical and radiological settings is essential for considering a diagnosis of LCH in cytology, as the findings in cytology might not always be characteristic of the lesion.

Cytologic evaluation of solitary lesions show highly cellular smears with sheets and singly scattered Langerhans cells; admixed with numerous inflammatory cells, predominantly eosinophils along with neutrophils, lymphocytes, plasma cells and multinucleated giant cells. Langerhans cells can be appreciated by the characteristic nuclear grooves and nuclear pseudoinclusions. Variable degree of nuclear pleomorphism; along with brisk mitotic activity can be appreciated in certain cases [7]. Rarely, dendrite-like cytoplasmic processes will be seen [8]. The population of eosinophils varies in different areas of LCH lesion, resulting in scant to numerous cells in cytology smears [8]. However, eosinophils can suggest the diagnosis of LCH.

In our case, the clinical differentials provided were tuberculosis and malignancy. As our case lacked a well formed granuloma, and showed plump histiocytes along with numerous neutrophils; we excluded a diagnosis of Tuberculosis. Also, the smears lacked any malignant cell population even after extensive search. Moreover, the histiocytes showed occasional nuclear grooves. These findings, coupled with age of the patient and radiological appearance of the lesion, led to a possibility of Langerhans Cell Histiocytosis in this case.

In langerhans cell histiocytosis, the biopsy usually show aggregates of langerhans cells with grooved and indented nuclei. Langerhans cells show characteristic positivity for S-100, PNA (peanut agglutinin), MHC class II, CD1a, and langerin (CD207) [9]. Of these, we tested for the expression of S-100 protein and CD1a. Both the markers were expressed by the tumor cells in a diffuse and strong pattern. Birbeck granules are considered the ultrastructural hallmark of LCH [9].

LCH with single organ involvement has a better prognosis than multiple organ involvement. If there is multiple organ involvement, then the prognosis is dependant on age of onset and number and type of organ involvement.

Occasional cases of LCH show spontaneous regression of the lesion with negligible mortality. In cases requiring treatment, the usual management followed is either surgery, radiotherapy, chemotherapeutic management using topical corticosteroids in cutaneous lesions, Mechlorethamine Hydrochloride solution, thalidomide or other combination regimens. The treatment modality considered is based on age, organ affected, systemic involvement of the disease and type of organs affected [10]. In this case, our patient was kept on conservative management as there was only a solitary lesion. However, the patient was subsequently lost for follow up.

CONCLUSION

Our case highlights the significant diagnostic role played by cytological evaluation, which when coupled with appropriate clinical and radiological setting is an effective guide to consider Langerhans Cell Histiocytosis.

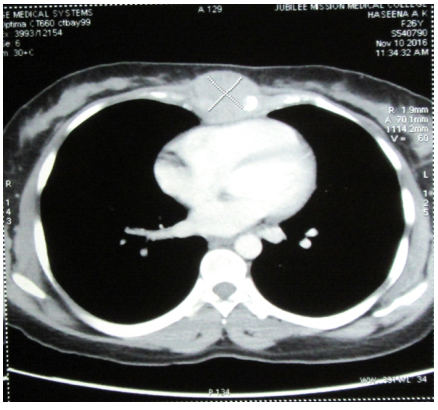

Fig 1: MRI image of chest showing sternal lytic lesion

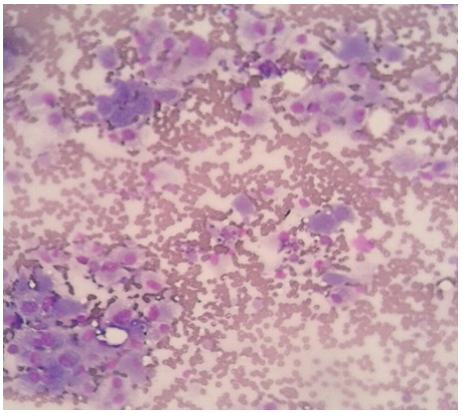

Fig 2: Cytology smears show epithelioid histiocytes in aggregates (Ma-Grunwald-Giemsa stain, magnification: 40)

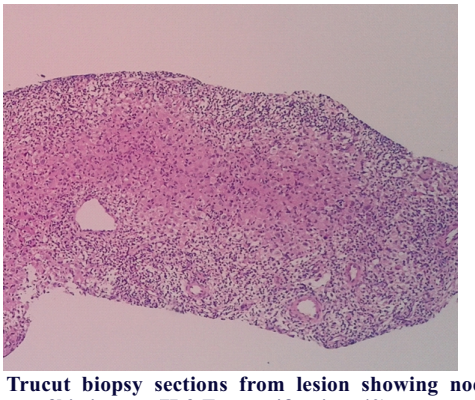

Fig 3: Trucut biopsy sections from lesion showing nodular aggregates of histiocytes (H & E, magnification: 40)

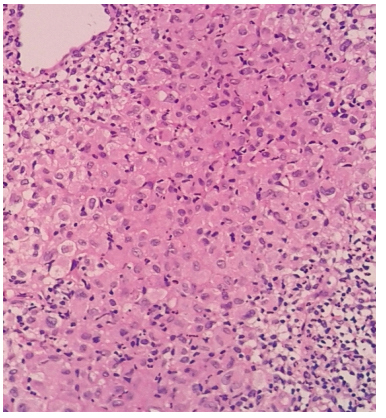

Fig 4 : In higher magnification the vesicular histiocytic nuclei with occasional grooved forms are visible (H & E, magnification: 200)

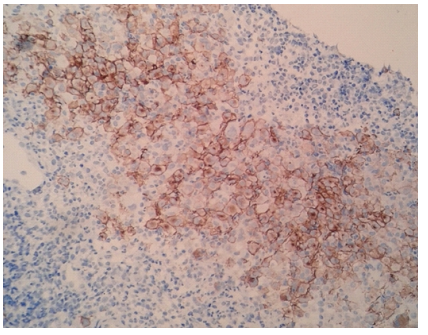

Fig 5 : Immunohistochemical evaluation with anti CD 1a antibody showed strong membranous positivity.

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