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Isolated Axillary Lymph Node Involvement in Adult Langerhans Cell Histiocytosis: A Rare Case Report.

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare disorder, characterised by the abnormal proliferation of Langerhans cells, a type of immune cell, leading to tissue damage and tumor-like lesions in various organ systems. It is a disease of childhood and is less commonly seen in adults. LCH can involve single or multiple organs, including bone, skin, pituitary gland, liver, lung, and lymph nodes. Diagnosis relies on biopsy and immunohistochemical markers such as CD1a and S100. Treatment depends on disease extent, ranging from observation or surgery for isolated lesions to systemic therapy for multisystem involvement. Prognosis varies by organ involvement and disease spread. We here by present a case of LCH involving the axillary lymph node.

Keywords: Langerhans cell histiocytosis (LCH), Axillary lymph node LCH, Histiocyte Society classification, Unifocal disease.

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INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disease involving the proliferation of langerhans in one or more organ systems. It is more commonly seen in children as compared to adults with an estimated incidence of 1-2 instances per million [1, 2]. Common sites include pituitary gland, bone, liver, lung and skin but it can occasionally involve the hematological system, lymph nodes, and lungs [1, 3]. The disease's clinical symptoms vary from single organ involvement to disseminated disease.A biopsy is typically required to diagnose LCH, which shows inflammatory cell infiltration with a high number of histiocytic cells and Langerhans cells that stain positively for S100 and CD1a [1, 2]. Here, we present a case of isolated axillary lymph node LCH in an otherwise healthy male.

Case presentation

A 25-year-old man presented to the surgical outpatient department with a 4–5 month history of a painful swelling in the axillary region with no history of fever, weight loss and loss of appetite. On clinical examination there was no significant past medical history or systemic symptoms. Ultrasonography revealed an oval-shaped hypoechoic lesion measuring 2.0 × 1.3 cm. Fine-needle aspiration cytology (FNAC) yielded a blood-mixed aspirate, and a preliminary diagnosis of histiocytic neoplasm was made, warranting further evaluation. An excisional biopsy of the swelling was performed one month later. Gross examination of the specimen revealed a soft tissue piece measuring 4.0 × 3.0 × 0.9 cm with homogeneous grey-brown areas on cut section. Histopathological analysis demonstrated a histiocytic lesion with periadnexal infiltration by sheets of histiocytic cells exhibiting folded nuclei with longitudinal grooves, and numerous eosinophils. On immunohistochemical staining these histiocytic cells were positive for CD1a and CD68, confirming the diagnosis of Langerhans cell histiocytosis.

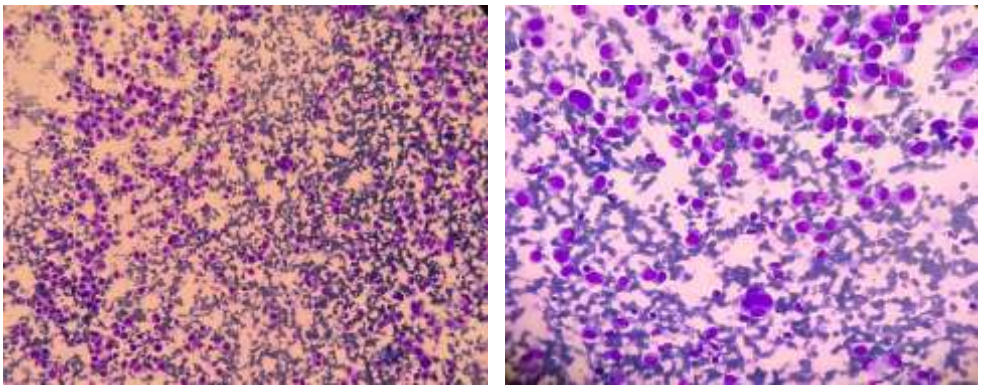

Fig 1 and Fig 2: 100x and 400x : Leishman stained smear showing large, pleomorphic cells with vacuolated cytoplasm with uninucleate to mulinucleated forms with eccentric placed nuclei.

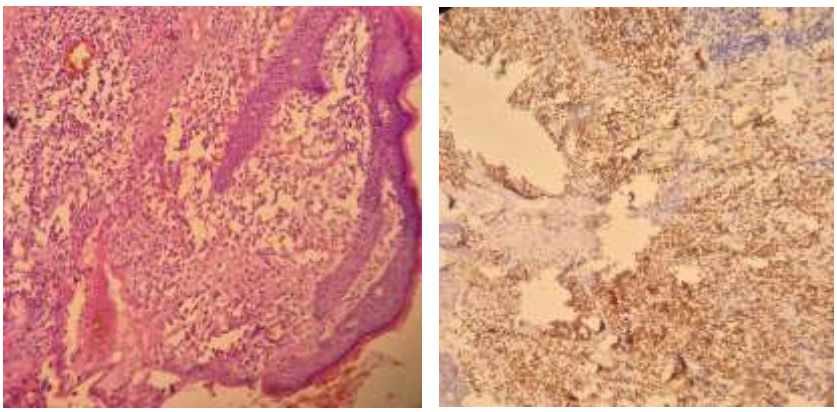

Fig 3: 100x: H/E section revealing periadnexal infiltration by sheets of histiocytic cells and numerous eosinophils.

Fig 4: 100x: CD1a shows cytoplasmic positivity.

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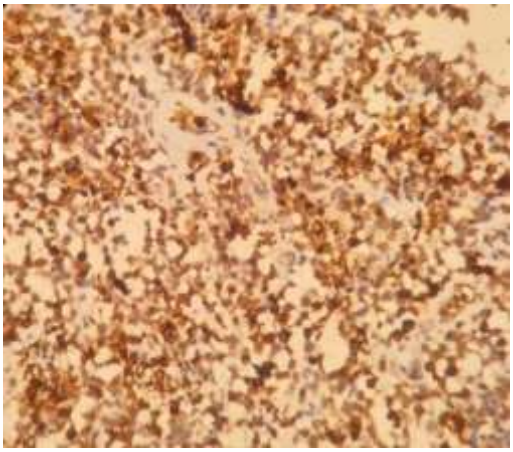

Fig 5: 400x: CD68 shows diffuse cytoplasmic positivity.

DISCUSSION

In 1985, the Histiocyte Society changed the name of of Eosinophilic granuloma, histiocytosis X, Letterer-Siwe illness, and Hand-Schuller-Christian disease to Langerhans Cell Histiocytosis (LCH).The disease's genesis and underlying pathogenic mechanism is unknown, but it may be caused by immune dysregulation that follows an excess of prostaglandin and cytokine production [3].

LCs are dendritic cells that generally dwell in the skin and mucosa and serve as the skin's major antigen-presenting cells. After encountering antigens, LCs move to lymph nodes and offer them to T cells [4].

Histiocytosis is an uncommon illness characterized by the accumulation of cells believed to be originating from dendritic cells or macrophages. LCH, the most common histiocytic disorder, affects the function, differentiation,and proliferation of mononuclear phagocyte cells.[4] Diagnosis requires histopathological examination of tissue biopsy followed by immunohistochemistry anyalsis for IHC markers of langerhan cells like CD1a, CD207 along with S-100 [3].

The LCH Study Group classified LCH into two broad categories: single-system LCH and multi-system LCH. Single-system LCH was classified into two types: single site (unifocal bone, skin, or lymph node) and multiple site (multifocal bone or numerous lymph nodes) [2]. LCH is a rare condition that primarily affects children. Adult cases typically involve a particular organ system, such as the bones or lungs. The clinical aspects of adult LCH are not well characterized due to its rarity [4]. In a 2003 research based on data from the Histiocyte Society's International Registry, the average age at disease onset was 33 years. This registry data included 18 years or older adults only, totaling 274 patients from 13 different countries, collected from January 2000 to June 2001 [5].

The prevalence of involvement of the bone is 80 %, skin 25 %; the pituitary gland 25 %; spleen 15 %; liver 15 %; the hematological system 15 %; lymph nodes 5-10 %; and cranial involvement excluding the pituitary gland 2–4 %. Lymph node involvement has been described in pediatric patients with systemic LCH. However, primary LCH of lymph nodes without involvement of other sites is unusual. The diagnosis of LCH is made using clinical and radiographic findings, as well as histological and immunophenotypic analysis. Histopathologically, lymphnodes involved in LCH frequently exhibit effacement of the nodal architecture and sinuses that are significantly distended by histiocytes. Histiocyte immunohistochemical staining is positive for CD1a, S-100, CD207 and CD-68 [1, 3, 4].

The histopathologic differential diagnosis of LCH presenting with lymphadenopathy covers all illnesses that can mimic LCH depending on the amount of the disease's nodal involvement,such as Kimura's disease, dermatopathic lymphadenitis, and Hodgkin lymphoma. Treatment for LCH is adapted to the scope and severity of the condition at diagnosis. The appearance of LCH as isolated solitary lymph node enlargement is uncommon and appears to be minimal risk for patients due to single system, unifocal involvement. In certain circumstances, isolated LCH has resulted in spontaneous regression of the lesion.The treatment of an isolated lymph node with confirmed LCH may include watchful waiting or

surgical removal. Surgical excisional biopsy is frequently used as the primary technique of diagnosis and treatment, as it was in this case [2, 4].

CONCLUSION

LCH is distinguished by a wide range of clinical symptoms and a biopsy is frequently required for definitive diagnosis. Atypical presentations of LCH pose diagnostic problems and necessitate a high level of suspicion. Our case report emphasizes the atypical presentation of a rare disease and offers us an important lesson: investigate the diagnostic possibility of LCH because the correct diagnosis determines the appropriate treatment.

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