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TISSUE DIAGNOSIS OF TWO DISTINCT EOSINOPHIL RICH LESIONS WITH SIMILAR HISTOMORPHOLOGY

Pathology

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ABSTRACT

Eosinophil rich lesions in histopathology have a wide range of differential diagnosis including conditions arising from inflammatory, reactive, immune mediated, drugs, parasitic infestations to neoplasms of both hematopoietic and non-hematopoietic origin (Nutman, 2007). Some of them are yet to be clearly differentiated by histological criteria, though being genetically different, such as Kimura disease and Angiolymphoid hyperplasia with eosinophilia (ALHE) (Googe et al., 1987). Some close mimics of eosinophil rich reactive and clonal lesions need to be differentiated based on the site of involvement, radiological features and appropriate immunohistochemical evaluation on biopsy tissues. We present here two distinct diagnoses of Langerhans cell histiocytosis (LCH) and ALHE which appear similar in terms of eosinophil rich inflammatory infiltrate on histological examination but have entirely different clinical course.

Summary: Langerhans cell histiocytosis is a locally aggressive slow growing neoplasm while Angiolymphoid hyperplasia with eosinophilia is a reactive lesion. Problem arises sometimes in the tissue diagnosis of these two lesions owing to their similarities in a couple of features. Both have a predilection for head and neck region. Both appear as an inflammatory lesion with eosinophils being a striking feature in biopsy tissues. Since, both the lesions have an entirely different clinical behaviour and prognosis, it is important to differentiate between the two based on the histopathology, clinical as well as radiological features. This paper emphasizes upon the same.

KEYWORDS

langerhans cell histiocytosis, angiolymphoid hyperplasia, eosinophil, ALHE, LCH

INTRODUCTION

Presence of increased eosinophils in biopsy tissue has distinct connotations depending upon the amount (number), site of localization in the tissue and the associated histomorphology. There are many differential diagnoses of increased eosinophils in biopsy tissues. This may include inflammatory conditions such as eosinophilic abscess to parasitic infestation, allergic or hypersensitivity reaction, drug induced changes and to a number of reactive and neoplastic pathologies. We present here the histomorphological features of two distinct eosinophil rich lesions which rely upon the light microscopic identification of striking tissue eosinophilia. The lesions include, Langerhans cell histiocytosis (LCH) and Angiolymphoid hyperplasia with eosinophilia (ALHE).

Langerhans cell histiocytosis is a rare clonal disease of the monocyte-macrophage system characterized by uncontrolled proliferation and accumulation of CD1a positive and CD207+ (Langerin) positive dendritic cells (DCs) as a result of continuous immune stimulation (Allen et al., 2018; Rodriguez-Galindo & Allen, 2020). Clinical presentation is highly variable. However, bone involvement is the most common feature. LCH can involve any bone of the body, of which skull is the most common site in the pediatric population.

Angiolymphoid hyperplasia with Eosinophilia (ALHE) is a lesion which occurs in 3rd to 4th decade of life with slight female preponderance. It has a predilection for head and neck area, especially peri-auricular region (Sharp et al., 1990). It is also called as epithelioid hemangioma owing to its prominent vascular features.

Case report

Case 1.

An 8-year-old male presented with complaints of pain and restricted mobility of left elbow joint for 6 months. MRI showed osteolytic and mildly expansile lesion in olecranon and coronoid process including adjacent shaft of the proximal ulna.

Possibility of chondromyxoid fibroma/eosinophilic granuloma was considered radiologically. The patient did not have any remarkable abnormalities on routine blood, urinary, biochemical and viral investigations. He did have a mild hepatomegaly.

Biopsy was done which showed two grey brown soft tissue bits together measuring 1x0.8x0.3cm. Cut surface was grey brown and soft. No bony tissue was identified.

Microscopic examination showed fibro-collagenous tissue fragments with presence of patchy dense inflammatory infiltrate comprising of plasma cells, lymphocytes, eosinophils including eosinophilic abscesses at places (Figure 1A). Few multinucleated giant cells and plenty of grooved histiocytes (Figure 1B) were present in the entire tissue. Immunohistochemistry showed a strong cytoplasmic immunoreactivity for CD 68 and CD1a as shown in the bottom two photographs of the panel below.

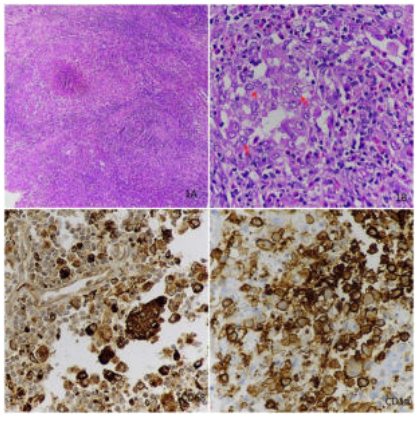

Fig.1A, B (Haematoxylin and eosin stained tissue sections at low and high power view): Densely inflamed fibrocollagenous tissue with an eosinophil rich mixed inflammatory infiltrate (1A). High-power view showing collection of histiocytes with nuclear grooves and scattered eosinophils on the periphery. CD68 (bottom left) and CD1a (bottom right) immunostains show strong cytoplasmic positivity. CD68 also highlights the multinucleated giant cells (Centre of the field).

Case 2.

A sixteen-year-old male presented with post auricular swelling for 3 years. Dull aching pain and occasional pruritis were present. On examination, swelling was non-tender, non-indurated. He did not have any derangements on routine investigations like complete blood count, routine urine examination, random blood sugar etc. His routine viral markers were negative. He did not have any history, clinical signs/symptoms of Tuberculosis or other chronic organ disorders. Biopsy was taken from the postauricular lesion which grossly showed a greyish white, partly skin covered soft tissue measuring 3.3x1.2x0.6

cm. Cut surface was firm, whitish.

Histopathological examination showed a fibrocollagenous tissue with lobular inflammatory cell infiltrate with proliferation of plump cuboidal to epithelioid cell. Intracytoplasmic vacuoles were seen. Moderate to dense chronic inflammatory infiltrate rich in eosinophils, lymphocytes and plasma cells was present in the parenchyma.

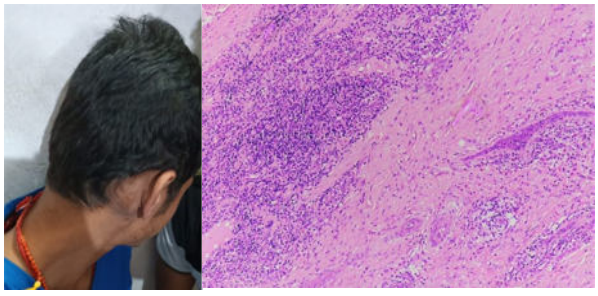

Figure 2: Patient with ALHE presenting with postauricular swelling. Histopathology showing dense inflammatory infiltrate comprising of eosinophils, plasma cells and lymphocytes on a collagenous background. Many thin and thick walled vessels were present with plump endothelial cell lining.

DISCUSSION:

These cases highlight different diseases which show abundance of eosinophils along with lymphocyte and plasma cells in varying proportions under the microscope.

LCH is primarily a disease of childhood but it also occurs in adults, including elderly. It is an uncommon tumor which most commonly presents as osteolytic lesion. Epidermal Langerhans cells are dendritic cells and belong to the bone marrow derived hematopoietic lineage cells (Jaitley & Saraswathi, 2012). The pathophysiology, though not clear, has now been found to initiate somatic mutations of the MAP-Kinase pathway in myeloid precursor cells(Allen et al., 2018). This has also led to LCH being viewed as a myeloid neoplastic disorder by some researchers. However, there are additional immunoregulatory factors at play. On microscopic examination, bone injuries show abundance of polygonal cells with eosinophilic cytoplasm, oval nucleus having coffee bean like longitudinal grooves (Favara & Jaffe, 1994). These typical cells are of histiocytic nature. These histiocytes, along with giant cells, lymphocytes, and eosinophils, may infiltrate soft tissue, mainly skin, lungs, liver, and spleen. The most definitive diagnosis of LCH relies on an ultrastructural demonstration of Birbeck granules.

LCH is grouped into three distinct clinical types: Single-system Single site (SS-s), Single-system Multi-site (SS-m), and a Multisystem type (Ms). LCH can prove fatal if left untreated. Though LCH most commonly presents as a lytic skull lesion and/or otitis in children (Saliba et al., 2008), it was a child with left elbow involvement in our index case. Treatment depends upon the site and number of lesions. Patients with single lesions may respond well to local treatment however, patients with multisystem disease usually need systemic therapy (Rodriguez-Galindo & Allen, 2020). Prognosis for single lesion is far superior to multisystem LCH.

Angiolymphoid hyperplasia with eosinophilia is an uncommon and benign vascular lesion. It clinically presents as angiomatous papules or nodules. The common site of involvement is head and neck region (Wells & Whimster, 1969). There is marked proliferation of blood vessels with distinctive plump endothelial cells (Olsen & Helwig, 1985). ALHE follows a benign clinical course, with no metastases reported so far, and with only few cases recurring when incompletely excised.

Both ALHE and LCH require surgical excision. But, while ALHE has a very timid chances of recurrence, LCH may require an additional chemotherapy in many of the cases.

CONCLUSION:

LCH and ALHE both may commonly affect younger age group and have a predilection for head and neck area. Radiology for LCH and histopathological examination for both the diseases is a mandatory confirmatory diagnostic modality. Since, both LCH and ALHE present as inflammatory lesion having an eosinophil rich infiltrate, it is important to consider the clinical, radiological and appropriate histopathological features coupled with immunohistochemistry for separating them out as they both have different biologic behaviour and treatments.

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