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Conjunctival Langerhans Cell Histiocytosis – A Case Report

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Abstract

Langerhans Cell Histiocytosis (LCH) is a spectrum of diseases with typically three clinical forms viz., eosinophillic granuloma, Hand-Schuller-Christian disease and Letterer-Siwe disease, characterised by accumulation of histiocytes in various tissues e.g., bones, skin, hypothalamus-pituitary region and other organs as well[1]. It is rarely seen in the eyes with an affinity for orbits. We report a patient with LCH primarily involving inferior bulbar conjunctiva without systemic involvement.

Keywords: Langerhans Cell Histiocytosis (LCH), Conjunctiva, Histopathology

Introduction

LCH is an uncommon multisystem disorder which is predominantly seen in children[2]. Currently it is considered to be due to a transient immune reaction provoking the cytokine mediated proliferation of pathologic langerhan cells within hematopoietic marrow of affected bone[3,4,5].The characteristic lesion of LCH consists of langerhan cells, macrophages, eosinophills, T-lymphocytes and plasma cells[6]. Eosinophillic granuloma generally originates in bones while as Letterer-Siwe disease shows soft tissue and visceral involvement with or without bony lesions. Hand-Schuller-Christian disease manifests as a triad of exophthalmos, bony defects of skull and diabetes insipidus[7].

All the patients with unifocal ocular disease require a biopsy to establish and confirm LCH and should undergo a complete systemic evaluation to rule out any malignant disease. The histological diagnosis is based on staining with S-100 and CD1a antigen or finding Birbeck granules on electron microscopy[8,9].

Disease limited to a single site should either be observed or requires a local therapy (curettage, steroid injection or radiotherapy) while as multifocal disease is treated with systemic prednisone with or without chemotherapeutic agents like vinblastine or etoposide depending on the disease extent[10].

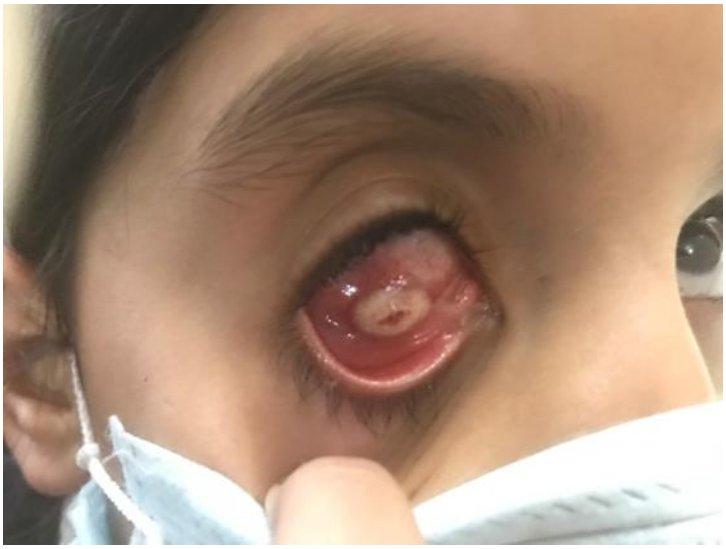
Case Report

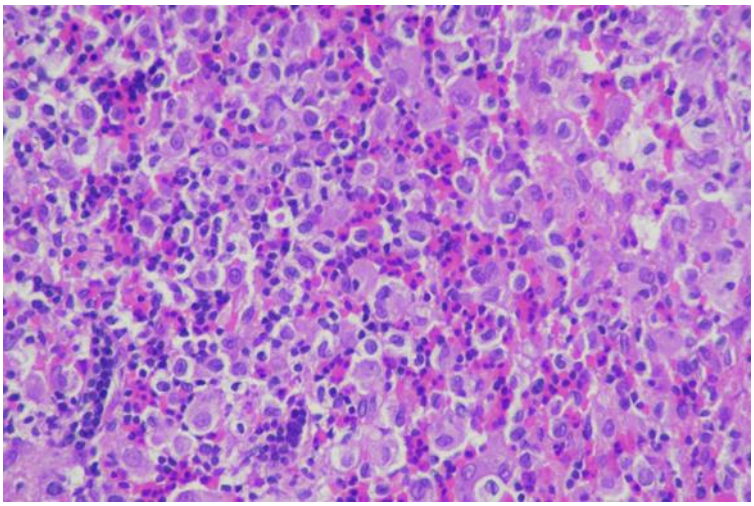
A seven years old girl presented to the Ophthalmology out patient department complaining of a painless swelling in the lower lid of the right eye for 2months.There was no history of discharge, itching, etc. On examination a solid yellowish- white oval mass measuring 2mm×3mm was seen in the inferior conjunctival fornix with surrounding conjunctival congestion. Her vision was 6/6 and rest of the ocular examination was normal.Left eye was normal. Considering it an infectious or inflammatory process, topical Moxifloxacin 0.5% and topical 0.5% Loteprednol were prescribed for 2 weeks. However with no improvement in 2 weeks a foreign body granuloma with abscess formation was suspected and the patient was planned for incision and drainage. However no pus could be drained on incision and drainage so the mass was excised completely and sent for histopathological examination(HPE). HPE revealed the features of LCH (eosiniphillic granuloma) following which the patient was referred to a paediatric oncologist for extensive evaluation to

rule out any systemic association. The diagnosis of LCH was further confirmed by positive S-100 protein and CD1a antigen. A systemic study revealed normal hemogram without cytopenia, normal renal function test, normal orbit and chest X-ray, and a normal whole body CT scan. The patient was kept on syrup Cefixime 100mg/5ml, syrup Ibuprofen and topical antibiotic steroid (Moxifloxacin and Dexamethasone) for 4 weeks.

The patient is on regular follow-ups with a paediatric oncologist and an ophthalmologist. The follow ups made in last 6 months after excision did not reveal any systemic involvement related to LCH . The inferior palpebral conjunctiva had healed well and no relapse of the lesion was seen.

Images





Discussion

LCH is a spectrum of clinicopathological condititons characterised histologically by a monoclonal proliferation of histiocyte like cells, identified as langerhan cells, accompanied by eosinophills lymphocytes, plasma cells and mutinucleate giant cells[11]. Langerhan cells are dendritic monoclonal cells which normally process and present the antigen to the T-lymphocytes and are seen in epidermis, mucosa, lymph nodes, bone marrow. In LCH the monoclonal proliferation of langerhan cells leads to destruction of hard and soft tissue[12].

The International Histiocytic Society defined objective criteria for the diagnosis of LCH which requires two or more than two criteria for the diagnosis; positive staining for adinosine

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triphosphate, S-100 protein antibody, alphamannosidase/peanut lectin binding[13].

LCH is predominantly seen in childhood with a peak incidence between 1 to 10 years with an aggressive course and worst prognosis in ages less than 2 years[14,15].

LCH has both inflammatory and tumorous characteristics, with some researchers advocating the disease concept "inflammatory myeloid tumour."

Conclusion

All the patients with unifocal orbital disease require a biopsy to establish and confirm the diagnosis and rule of any other malignant disease like in this patient. These patients should be referred to a paediatric oncologist for a complete systematic evaluation and subsequent follow ups in order to monitor recurrence or prognosis.

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