Case report

Ocular Langerhans Cell Histiocytosis (A Multisystem Involvement of Langerhans Cell Histiocytosis)

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Abstract

A 2 year old boy was referred from paediatrics OPD to Eye OPD with chief complaint of Multiple conjunctival cyst since 15 days. The patient had a prior history of recurrent chest infections. Requiring hospitalisation since 7 months of age. Upon visual acuity examination, the patient seemed to follow light.Fundus examination of both eyes were within normal limits.H& E sections of the lesions showed collection of histiocytes typical of Langerhans Cell Histiocytosis.Patient was treated with multiple cycles of Vinblastine and predinisolone as per LCH 3 guidelines. Langerhans cell histiocytosis (LCH), the most common histiocytic disorder in children. LCH lesions are granulomatous lesions consisting of pathologic "Langerhans cells" (LCs), lymphocytes (primarily T-cells), eosinophils, and macrophages. The incidence of LCH is estimated to be around 5–10 cases per million children per year and 1–2 cases per million adults per year with a male is to female ratio 1.2:1.Its pathology is 'Misguided Myeloid Differentiation' where state of differentiation of myeloid precursor in which activating MAPK mutation arises determines the extent and severity of disease[1]. The current standard of care for initial therapy is vinblastine/prednisone for one year.

Keywords: Cell, Conjunctival, Patient.

Case report

A 2 year old boy was referred from paediatrics OPD to Eye OPD with chief complaint of Multiple conjunctival cyst since 15 days.The patient had a prior history of recurrent chest infections. Requiring hospitalisation since 7 months of age.He had multiple small umbilicated nodular lesion on cheeks temporal area & forehead.Upon visual acuity examination, the patient seemed to follow light. Fundus examination of both eyes were within normal limits.Patient was advised for hot fomentation and asked to follow up with Chest Xray and CBC report.

CBC reports showed Hb 9.9,TLC 23,500,DLC- P – 67,L :22 ,M- 01, Platelets 2,40,000,Chest Xray showed honeycombing of Lungs with left sided pneumonia.Patient was then followed up in department of paediatrics where PET-CT imaging showed periportal soft tissue hypodensities in both lobes of liver, subcentimetric right upper paratrachealnode.USG abdomen showed heterogenous soft tissue in periportal region in left lobe of liver corresponding to area of tracer uptake of PET-CT.H& E sections of the excised lesions under sedation showed collection of histiocytes typical of Langerhans Cell Histiocytosis[2,3].

Fig.1: PICTURES SHOWING NODULAR CYSTIC LESIONS ON SKIN AND CONJUNTIVA AND HONEYCOMBING OF LUNG WITH LEFT SIDED PNEUMONIA SEEN IN CHEST X-RAY P-A VIEW

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Diagnosis
Langerhans Cell Histiocytosis (skin+ocular+liver)(multisystem system)[3,4], Risk involvement -Liver.

Management
Therapy started with Prednisolone/Vincristine/Cytarabine*6 weeks with repeat therapy at follow-up[5].
Skin and Ocular lesions decreased in size after 2 treatment cycles with USG showing few scattered cystic lesions in right lobe of liver with no definite communications with the biliary radicals and few enlarged periportal nodes[6].

Fig. 2: RESOLUTION OF DERMAL AND OCULAR NODULAR LESIONS WITH SYSTEMATIC IMPROVEMENT POST CHEMO THERAPY

Discussion
Langerhans cell histiocytosis (LCH) is a rare (about 3-5 cases per million children aged 0-14 years), non-malignant disease characterized by proliferation and accumulation of clonal dendritic cells, extreme clinical heterogeneity, and an unpredictable course[7]. Langerhans cell histiocytosis (LCH) is an inflammatory neoplasia of myeloid precursor cells driven by mutations in the mitogen-activated protein kinase pathway[3]. When disease involves the skin, it presents most commonly presents as seborrheic dermatitis or eczematous eruption on the scalp and trunk. Evaluation for involvement of other organ systems is essential, because 9 out of 10 patients presenting with cutaneous disease have multisystem involvement[4]. Clinical manifestations can range from an isolated disease with spontaneous resolution to life-threatening multisystem disorder. Prognosis depends on involvement of risk organs (liver, spleen, and bone marrow) at diagnosis, particularly on presence of organ dysfunction, and response to initial therapy. Systemic treatment incorporating steroids and cytostatic drugs has improved prognosis of multisystem LCH and represents the current standard of care[8]. Poor prognostic markers are multisystem presentation, risk-organ involvement, and poor response to initial therapy[9].

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Conflicts of interest
There are no conflicts of interest.

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