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They Grow (up) Too Fast

Case

A 6-year-old, healthy boy presented with a history of progressively increasing left upper eyelid swelling since 20 days. It was not responsive to the antibiotics started by the pediatrician considering a preseptal cellulitis. On examination, the best-corrected visual acuity was 6/6 in OD and 6/12 in OS. There was mechanical ptosis in the left eye with periocular fullness and soft, ill-defined palpable mass [Fig. 1a]. There was mild limitation of movement in upgaze but no proptosis. Right eye was normal. He was advised orbital computed tomography (CT), PET-CT, and peripheral blood smear (PBS).

What is your next step?

A. Fine needle aspiration biopsy (FNAB)
B. Excisional biopsy with curettage
C. Multilevel incisional biopsy with intraoperative frozen section diagnosis (IOFD)
D. Systemic steroids

Findings

CT revealed extraconal lesion involving superior and lateral recti, lacrimal gland, and temporal fossa with erosion and defect in orbital roof [Fig. 1b]. PBS was normal. PET-CT showed FDG avid lesion with no systemic abnormality. Multilevel incisional biopsy was performed with IOFD.

IOF-light microscopy showed diffuse proliferation of large, noncohesive, round-polygonal cells with eosinophilic cytoplasm, vesicular, grooved nuclei, and inconspicuous nucleoli [Fig. 1c]. Background showed polymorphous infiltrate of eosinophils, lymphocytes, neutrophils, plasma cells, and multinucleate giant cells. Features were consistent with eosinophilic granuloma. Intraoperative intralesional triamcinolone was injected. Cells were positive for CD68, S100, and CD1a, confirming the diagnosis of Langerhans cell histiocytosis (LCH) [Fig. 1d]. Child did well, at a follow up of six months, with complete resolution, bone remodelling and no recurrence. Bone scan was normal [Fig. 1e and f].

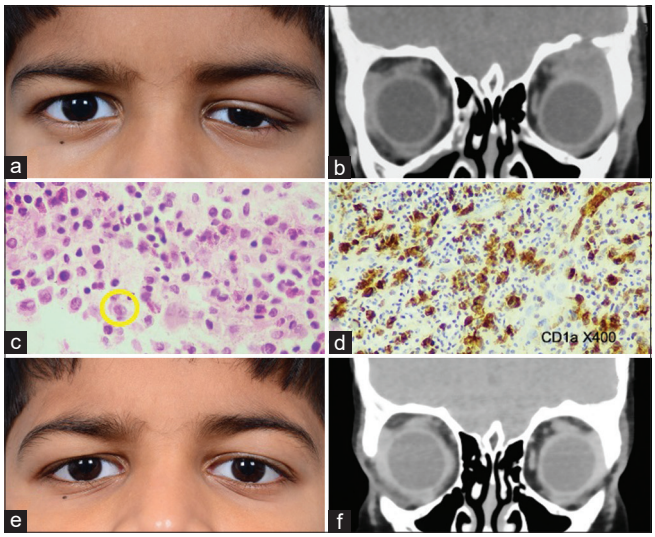

Figure 1: (a) Clinical photograph of a 6-year-old child showing left upper lid edema and ptosis. (b) Coronal section of CT scan showing an ill-defined, extraconal mass with erosion of the orbital roof. (c) Light microscopy showing diffuse round to polygonal cells with grooved nuclei (yellow circle) in a background of eosinophils, lymphocytes, and neutrophils. (d) The cells stained positively with CD1a, confirming the diagnosis of Langerhan cell histiocytosis. (e) The child responded very well to two intralesional injections of steroids and oral steroids with complete resolution. (f) CT scan 6 months post the incisional biopsy showing bone remodeling and no residual lesion

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Quick Response Code:	Website: www.ijoreports.in
	DOI: 10.4103/ijo.IJO_1583_21

Diagnosis: Eosinophilic granuloma

Correct answer: C

Discussion

LCH is a multisystem disease ranging from acute, disseminated Letterer–Siwe disease to chronic, multifocal Hand–Schuller–Christian disease and localized eosinophilic granuloma. Orbital presentation is rare and affects 4/100000 children annually (mean age: 8 years) with a male predominance.[1] Bone erosion is attributed to PGE2 and IL1 produced by histiocytes.[2] It can simulate rhabdomysarcoma, granulocytic sarcoma, primitive neuroendocrine tumor, and lymphoproliferative lesion. Single bone is most commonly involved followed by skin, bone marrow, CNS, and lymph node. FNAB can miss the diagnostic cells. IOFD allows intraoperative initiation of treatment including bone marrow biopsy, tumor debulking, or intralesional steroid injection. Excision with bone curettage is preferred for unifocal orbital lesions. Diffuse infiltrating nature and intracranial extension make this difficult and unsafe. Intralesional steroid (4 mg/kg) is minimally invasive with excellent response.[2,3] It can be repeated every 4–6 weeks. Long-term follow-up is critical for local recurrence and consecutive multifocal LCH.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Cite this article as: Sen M, Painuly S, Mulay K, Honavar SG. They Grow (up) Too Fast. Indian J Ophthalmol 2021;69:2256.