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Conjunctival chemosis or not?

Case Report

A 7-year-old girl presented with redness of the left eye since the past 4 months [Fig. 1a]. She was diagnosed elsewhere with left eye conjunctival chemosis and prescribed topical steroids and lubricants. Her best-corrected visual acuity was 20/20 in both eyes. There was left upper eyelid ptosis and a diffuse, pale pink thickening of the inferior and nasal bulbar conjunctiva, extending from the limbus to the inferior fornix with intrinsic blood vessels and small punctate hemorrhages within the lesion [Fig. 1b]. Proptosis was absent, ocular motility was full and free, and the rest of the anterior segment and fundus examination were normal in both the eyes.

What is Your Next Step

A. Start the patient on systemic steroids
B. Consider an orbital computed tomography scan
C. Get a complete blood count done including a peripheral smear
D. Do an incision biopsy from the conjunctival thickening.

Findings

Incision biopsy from the conjunctival lesion revealed polymorphous infiltrate of lymphoid cells with large atypical lymphoblastic cells in the background [Fig. 1c]. Immunohistochemistry identified the cells to be highly positive for terminal deoxynucleotidyl transferase and CD20, CD79a, and CD34 and negative for CD3, CD117, and MPO [Fig. 1d]. Her blood investigations showed a total leucocyte count of 8000 cells/mm^3 with 27% lymphocytes and 8%–10% atypical lymphoblasts. Bone marrow biopsy showed replacement of the marrow with predominant lymphoblasts. Cerebrospinal fluid cytology was normal. Systemic examination revealed left preauricular lymphadenopathy and no organomegaly.

Diagnosis

Conjunctival extramedullary B-lymphoblastic lymphoma/leukemia.

Correct Answer: C

Discussion

Ocular involvement is an uncommon extramedullary manifestation of acute leukaemia and is usually limited to retina, choroid, vitreous, optic nerve, orbit, or sclera.[1] Leukemic infiltration of the conjunctiva is rare and appears as fleshy erythematous thickening giving the appearance of a "solid chemosis" as it was in our patient.[2,3] Acute lymphocytic leukemia (ALL), the most common acute leukemia subtype in children, commonly presents with nonspecific symptoms of anemia and generalized weakness. In our patient, "solid chemosis" was the initial manifestation of ALL. Peripheral blood smear showed immature lymphoblastic cells and it should be part of initial evaluation for an early diagnosis in such cases. Surgery can be avoided in these patients if a peripheral blood smear is performed primarily and detects the presence of blast cells.

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.

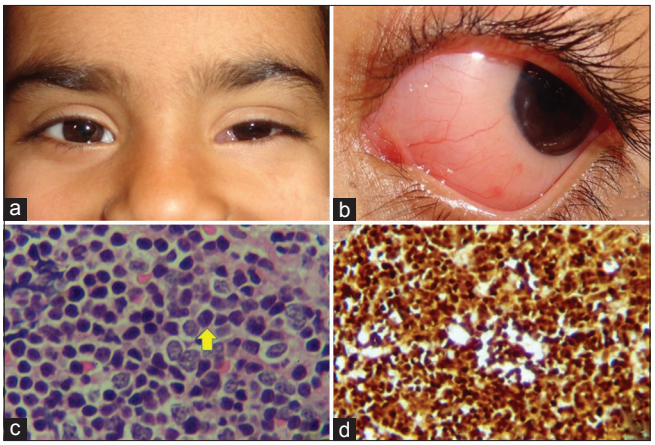

Figure 1: Clinical photograph showing left upper eyelid ptosis and conjunctival chemosis left eye (a). High magnification photograph showing pale pink thickening of the inferior and nasal bulbar conjunctiva with areas of punctate haemorrhages (b). Histopathology of the conjunctival incision biopsy specimen showing dense infiltrate of malignant cells with prominent plasmacytic features and large atypical lymphoblasts (yellow arrow) (H and E, ×40) (c). Immunohistochemistry showing positive staining with terminal deoxynucleotidyl transferase (Tdt) (d)

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Conflicts of interest

There are no conflicts of interest.

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