

To prick or to poke?

Case

A 15-year-old male presented with gross proptosis of right eye for the past 3 weeks. On presentation, visual acuity in the right eye was just perception of light while it was 20/20 in the left eye. Right eye was grossly proptosed with severe exposure keratopathy and surrounding conjunctival chemosis [Fig. 1a]. A firm mass was palpable in the superior quadrant of the orbit. Computed tomography scan was suggestive of bilateral well-defined isodense mass lesion in the superior extraconal space with dural-based masses in the frontal and parietal lobes [Fig. 1b]. Patient had been extensively investigated 2 weeks back by a local ophthalmologist and the blood investigation revealed severe microcytic hypochromic anemia (hemoglobin 6 gm/dl). However, there were no abnormal white blood cells in the peripheral smear.

What is Your Next Step?

- Refer the patient to a hematologist to build up hemoglobin and plan for incisional biopsy
- Urgent incisional biopsy under local anesthesia
- Bone marrow biopsy
- Repeat peripheral blood smear

Findings

Repeat peripheral bloods smear revealed blast cells with multiple nucleoli and Auer rods suggestive of acute myeloid leukemia [Fig. 2a and b]. The patient was referred to a hematologist for further management.

Diagnosis

Orbital granulocytic sarcoma

Correct Answer: D.

Discussion

Granulocytic sarcoma is a childhood malignancy mostly seen in less than 10 years of age. It is seen in 3–5% of patients with acute myeloid leukemia (AML).^[1] When granulocytic sarcoma occurs in the absence of AML (aleukemic leukemia), it is known as de novo granulocytic sarcoma.^[1] De novo orbital granulocytic sarcoma is rare, and in all such cases, both the peripheral blood smear and bone marrow biopsy are normal, while biopsy from the orbital mass suggests granulocytic sarcoma.^[2,3] Although the mean duration after which leukemia develops in a case of de novo granulocytic sarcoma has been reported to be 10.5 months,^[4] it is always advisable to repeat the peripheral blood smear in all cases where the index of suspicion is high even if the report had been normal in the recent past. This simple test precludes the need of an orbital biopsy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for the patient's image and other clinical information to be reported in the journal. The patient's parents understand that the patient's name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

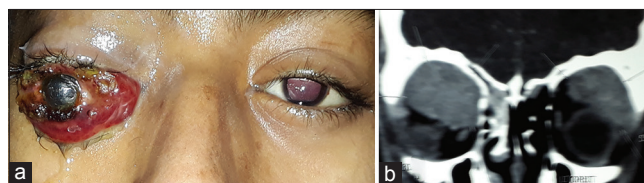


Figure 1: (a) External photograph showing gross proptosis of the right eye with severe exposure keratopathy, and (b) computed tomography scan of the orbit, coronal cuts, showing bilateral isodense orbital mass in the superior extraconal space

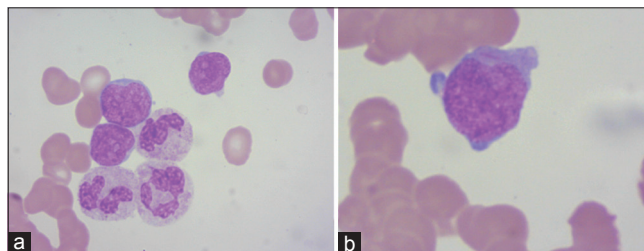


Figure 2: (a) Peripheral blood smear showing numerous blast cells with prominent nucleoli, and (b) peripheral blood smear with blast cell demonstrating Auer rod

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Conflicts of interest

There are no conflicts of interest.

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Md. Shahid Alam, Vathsalya Vijay¹

Department of Orbit, Oculoplasty, Reconstructive and Aesthetic Services, Aditya Birla Sankara Nethralaya, Kolkata, West Bengal, ¹Department of Orbit Oculoplasty Reconstructive and Aesthetic Services, Sankara Nethralaya, Medical Research Foundation, Chennai, Tamil Nadu, India

Correspondence to: Dr. Md. Shahid Alam,

Department of Orbit, Oculoplasty, Reconstructive and Aesthetic Services, Aditya Birla Sankara Nethralaya, Mukundapur, EM Bypass, Kolkata - 700 099, West Bengal, India.
E-mail: mshahidalam@gmail.com

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