Case Report:
Granulocytic Sarcoma as the First Sign of Acute Leukemia in Childhood

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Abstract: Acute myeloid leukemia (AML) may rarely involve the orbit as a solid tumor termed granulocytic sarcoma. This report describes the case of a child who presented with rapidly progressive unilateral proptosis and was diagnosed as rhabdomyosarcoma. However subsequent examination of the peripheral blood film revealed AML. Thus proptosis may present as the initial manifestation of AML.

Key Words: Granulocytic sarcoma; Acute myeloid leukemia; Proptosis.

Introduction:
The orbit, especially in children can be involved in a number of diseases either ophthalmic or non-ophthalmic. The common lesions include orbital cellulitis, dermoid cysts, rhabdomyosarcoma and hemangioma. However certain rare causes also exist, like granulocytic sarcoma. Thus the differential diagnosis should be kept in mind and a thorough clinical examination and appropriate investigations should be carried out to facilitate early treatment.

Case Report:
Eight years old child presented with proptosis of right eye with ocular pain and redness over a span of 15 days.
Vision in right eye was 20/200 (later decreased to 20/400). Proptosis of 6 mm was noted with downward and lateral displacement of globe [Fig 1]. Extra ocular movements were restricted superiorly and inferiorly. Conjunctiva was congested and chemosed, cornea showed exposure keratitis, minimal hypopyon, pupil reacted sluggishly to light, intraocular pressure was 22 mmHg, fundus examination revealed normal disc with dilated tortuous vessels. Left eye was normal.
A provisional diagnosis of rhabdomyosarcoma was made due to the acute onset of proptosis and young age, rhabdomyosarcoma being the commoner primary orbital malignancy in children. CT Scan revealed 3 x 3 cm well defined mass in superior aspect of right orbit with contrast enhancement, superior rectus was not seen separately. No bone destruction, no intracranial extension was seen [Fig 2]. Radiologists suggested hemangioma.
Subsequently peripheral blood film (PBF) was ordered which showed lymphopenia and thrombocytopenia, hypochromic macrocytes with anisocytosis, polymorphs 30%, lymphocytes 25%, >blasts 45% [Fig 3].
openia and thrombocytopenia. On account of severe thrombocytopenia, we did not proceed with orbital biopsy.

Granulocytic sarcoma or chloroma is a tumor composed of granulocytic precursor cells occurring in an extramedullary location. The term chloroma “green tumor” is derived from the greenish gross coloration of this lesion, attributable to the myeloperoxidase in the cells of granulocytic lineage. The theory for the formation of these lesions is that normal hematopoietic progenitor cells selectively bind to bone marrow stroma and then begin to proliferate and differentiate, while in vitro they bind to skin fibroblasts. Binding of these cells to the fibroblasts localize in non-hematopoietic tissues may result in the formation of extramedullary myeloid metaplasia. The predilection for bone or subperiosteal involvement in the axial skeleton is thought to be related to the active hematopoiesis in these locations. Orbital cases can be explained by their origin from adjacent bone, or less commonly, the lacrimal gland or extraocular muscles. Myeloid sarcomas are most common in certain subtypes of AML, in particular M5a (monoblastic), M5b (monocytic), M4 (myelomonocytic), and M2 (myeloblastic with maturation).

The differential diagnosis of acute childhood proptosis includes orbital cellulitis, inflammatory pseudotumor, ruptured dermoid cyst, rhabdomyosarcoma, hemangioma, chocolate cyst of lymphangioma, neuroblastoma, granulocytic sarcoma. Of all the orbital lesions only 5% are malignant and granulocytic sarcoma accounts for only 1 of 250 malignant cases with the incidence being slightly higher in Africa and Asia. In a study of 86 Indian patients with AML, 8 (9.3%) were found to have orbital deposits in one or both eyes. Majority of the cases present as bilateral proptosis. However our case showed the rare unilateral presentation.

In many cases proptosis precedes the systemic manifestations of AML. Most cases in patients without a previous diagnosis progress to AML within one year and even earlier in the case of an initial orbital disease. However, some patients do not develop hematologic evidence of the disease as long as 30 months following the presence of an orbital tumor. In our case too, proptosis preceded the systemic manifestation of AML. In the absence of systemic features it becomes difficult to diagnose granulocytic sarcoma. In such cases PBF, a simple and inexpensive investigation proves effective in diagnosis and acts as an easy tool in saving time and money spent on CT and MRI and thus initiating early treatment.

References: