

## Proptosis in a Teen

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**Abstract:** We report a case of 15 years old boy who presented with unilateral proptosis as initial manifestation of acute myeloid leukemia. This case highlights how important it is to examine the peripheral smear of such a case for early diagnosis and prompt treatment for cure.

**Keywords:** Acute Myeloid Leukemia, Myeloid Sarcoma, Proptosis.

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### INTRODUCTION

Myeloid sarcomas are rare extramedullary manifestations of Acute Myeloid Leukemia (AML) [1]. Orbit is the most common site [2] and in the case of orbital location, bilateral proptosis is more frequent than unilateral proptosis.

We report about a 15 year old boy who presented with unilateral proptosis as initial manifestation of acute myeloid leukemia. Even though it is a well described entity it is not common to encounter such a patient in general practice. This case highlights how important it is to examine the peripheral smear of such a case for early diagnosis and prompt treatment for cure.

### CASE REPORT

A 15 year old boy presented with pain, swelling and blurring of vision of the right eye since 1 month. Examination showed gross eccentric proptosis of the right eye with chemosis, lagophthalmos, restricted ocular movements and decreased vision (Fig. 1). His general examination was unremarkable, and he had mild hepatosplenomegaly. A computed tomogram of the orbits showed a well defined soft tissue density lesion 3.8x1.8x1.6 cm involving the retrobulbar region of the right orbit, confined to the extraconal compartment. MRI brain showed proptosis of the right eye with a well defined 3.4x2.8x3 cm irregular lesion superlaterally. The lesion was isointense to muscle on T1 and hyperintense on T2W and was uniformly enhancing with contrast. Right eye ball was inseparable from the mass (Fig. 2). His haemoglobin was 10.3 gm/dl, total leucocyte count was 20500/mm<sup>3</sup> with 64% atypical cells, and platelet was 63000/mm<sup>3</sup>, peripheral smear showed 34% abnormal cells, serum chemistries were normal and LDH was 1814U/L.

A bone marrow examination showed 80% blasts which were peroxidase positive. Flow cytometry showed the abnormal cells to be positive for CD13, CD33, CD64, CD117 and CD11C, CD34 and HLA DR. A diagnosis of acute myeloid leukemia (AML) with myeloid sarcoma of the right orbit was made.

He received induction chemotherapy with Ara C and Daunorubicin (7/3). The orbital mass disappeared completely after induction and his bone marrow achieved complete remission (Fig. 3). He received consolidation with high dose AraC. A repeat MRI orbit was normal and he has regained his vision. He was in remission for the past 5 months and now he relapsed.



**Fig-1: Right eye showing eccentric proptosis with chemosis**



**Fig- 2: MRI showing the mass inseparable from the right eye ball.**



**Fig. 3: The orbital mass disappeared completely after induction chemotherapy**

## DISCUSSION

Myeloid sarcoma (MS) also known as granulocytic sarcoma or chloroma is a rare extramedullary tumor composed of myeloid progenitor cells. The most common sites of involvement are the periosteum, soft tissue, bone, lymph nodes but any site may be involved like the brain, breasts, urinary bladder, and gastrointestinal tract [3]. Males and females are equally affected, with a mean age of 48 years. Myeloid sarcoma occurs in only 2 to 8% of AML mostly with FAB subtypes M2, M4 and M5 [4, 5]. It occurs either concurrently, following or rarely prior to the onset of AML. It is seen less frequently in chronic myelogenous leukemia and other myeloproliferative disorders.

Orbital involvement as an initial manifestation of AML is less common and present with unilateral proptosis. The proptosis is usually caused by leukemic infiltration, rarely by retrobulbar hemorrhage or venous blockade. Other ocular manifestations include ptosis, conjunctival involvement, lacrimal gland involvement, restricted ocular movements and decreased vision [5]. Orbital granulocytic sarcoma typically affects children and young adults. On MRI, these lesions appear isointense on T1W and hyperintense on T2W, enhancing intensely with contrast [6]. Bone marrow examination and flow cytometry would help to confirm the diagnosis of systemic disease. Treatment of myeloid sarcoma is similar to AML with induction chemotherapy with AraC and daunorubicin and consolidation chemotherapy followed by allogenic bone marrow transplantation.

## CONCLUSION

The orbital mass in our patient disappeared completely following chemotherapy. This case narrates the need for looking at the peripheral smear in a child who presents with proptosis.

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