

## A Rare Case of Myeloid Sarcoma Presenting As Bilateral Proptosis in a Patient of Acute Myeloid Leukemia

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**Abstract:** Bilateral proptosis in a case of acute myeloid leukemia is a rare and unusual presentation. The extramedullary proliferation of the myeloblasts is known as myeloid sarcoma. It is also known as chloroma. About 2% of AML cases presents as myeloid sarcoma. It has approximately the same rate of occurrence in both sexes. Children are more often affected than adults. We report a very unusual case of acute myeloid leukemia in a 3 year old patient presenting with sudden onset bilateral proptosis as its initial manifestation. Radiological investigation, fnac and bone marrow study was done for confirmation. The purpose of reporting such a rare entity is to highlight AML as a rare but important differential diagnosis of bilateral proptosis.

**Keywords:** acute myeloid leukemia, chloroma, proptosis.

### I. Introduction

Leukaemia is a disease resulting from the neoplastic proliferation of haemopoietic or lymphoid cells. It results from mutation of a single stem cell, the progeny of which form a clone of leukaemic cells. The term acute myeloid leukemia (AML) refers to a group of marrow-based neoplasms that have clinical similarities and distinct morphologic, immunophenotypic, cytogenetic, and molecular features. Myeloid sarcoma is an extramedullary tumor that occurs in 2 to 14% of cases of AML. (1). This tumor was first described by Allen Burns (2) in 1811. These tumors are called chloromas because some appear green or turn green in dilute acid secondary to expression of MPO. The tumors are usually localized; they often involve bone, periosteum, soft tissues, lymph nodes, or skin. Myeloid sarcomas may occur at diagnosis of AML or may precede the diagnosis. These sarcomas are very sensitive to focal irradiation or chemotherapy; they generally resolve completely in less than 3 months, although they recur in approximately 23% of patients. (3). AML typically presents with systemic findings secondary to derangement of the hemopoietic system. Common symptoms include fever, easy bruising, pain, weight loss and pain in the joints. Common causes of acute bilateral proptosis in pediatric age group are: orbital cellulitis, inflammatory pseudotumor, metastatic neuroblastoma, thyroid orbitopathy and rhabdomyosarcoma. Rarely, the cause of this acute bilateral proptosis in children may be due to acute myeloid leukemia. Rarely AML may present with sudden bilateral proptosis with out any systemic symptoms of leukemia. We report such a case in a 3 year old patient alongwith its characteristic findings.

### II. Case Report

A 3 year old male patient presented with a complain of sudden onset bilateral proptosis (Fig 1) along with fever. For fever the patient was initially given broad spectrum antibiotics but the fever failed to subside. After a week later the parents noticed swelling around the right eye which within a week fully became protruded followed by the protrusion of left eye. The proptoses of both eyes were painless. Eye examination revealed bilateral, non-reducible, non-axial proptosis. The right eye proptosis was more than the left eye. Movement of the globe was restricted due to myositis. The eyelids were retracted and open with chemosed conjunctiva. cornea was dry looking with exposure keratitis. Visual acuity was impaired.

The complete blood count of the patient revealed elevated WBC count ( $26.70 \times 10^3/\mu\text{l}$ ) and reduced platelet. Peripheral blood smear showed more than 30% myeloblast (Fig 2). After this a bone marrow study was advised and done to confirm the diagnosis. Bone marrow aspiration study revealed myeloblast 40%, myelocyte 16%, metamyelocyte 12%, neutrophils 16%, lymphocytes 12%, monocyte 2% and eosinophil 2% with depressed thrombopoiesis (Fig 3). The myeloblasts were strongly positive for Sudan black stain (Fig 4). The final diagnosis was given AML M2 (according to FAB classification).

CT scan of the orbits showed homogenously enhancing soft tissue density lesion in preseptal, intraorbital region involving intracoronar and extra coronal space bilaterally (Fig 5). There was extension of the mass in to the bilateral ethmoidal sinuses and bilateral maxillary sinuses with patchy destruction of floor of orbit and nasal septa. No intracranial extension was seen.

FNAC of the orbital swelling from both the sides revealed presence of isolated, non cohesive, non granular cells, with round to oval nucleus with fine, dispersed chromatin, distinct nuclear membrane, small eosinophilic nucleolus and rim of pale cytoplasm (Fig 6). Few of these myeloblastic cells showed presence of

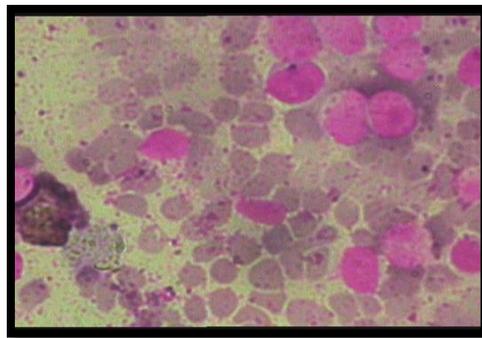
Auer rods (Fig 7). Hence the diagnosis was given as orbital infiltration of myeloblastic cells, a feature consistent with myeloid sarcoma.

Cytogenetic studies could not be done in our case. A diagnosis of acute myeloid leukaemia with myeloid sarcoma was given. The patient was referred for chemotherapy

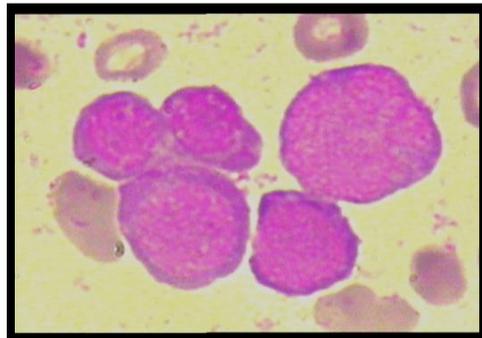
**1.1 Pictures**



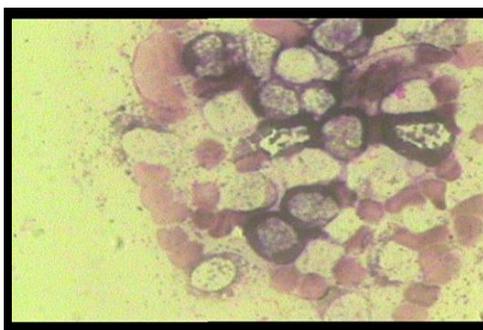
**Figure 1:** bilateral proptosis



**Figure 2:** peripheral blood smear



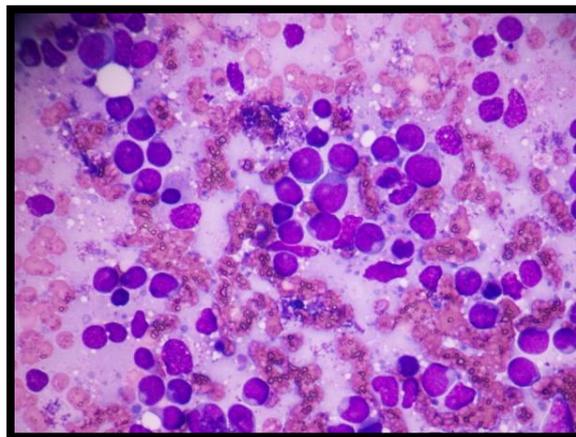
**Figure 3:** bone marrow aspiration smear



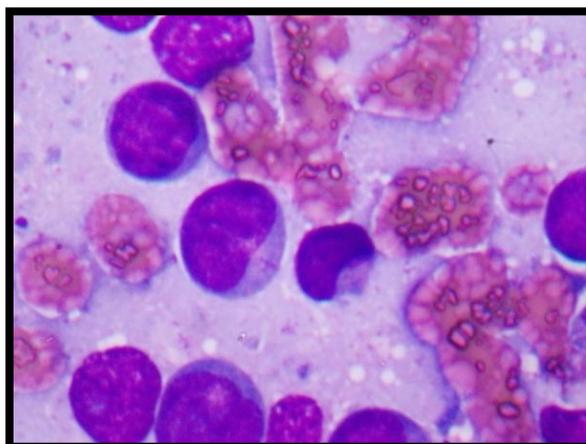
**Figure 4:** bone marrow aspiration smear stained with sudan black.



**Figure 5:** ct scan: coronal section showing bilateral soft tissue mass involving intraorbital region.



**Figure 6:** fnac smear of orbital swelling



**Figure 7:** fnac smear of orbital swelling showing auer rods in myeloblast

### **III. Discussion**

Acute myeloid leukemia (AML) accounts for nearly 15% of all leukemias in children (4). Myeloid sarcoma is an extramedullary tumor that occurs in 2 to 14% of cases of AML. Rarer sites of its presentation include the orbit, the paranasal sinuses, the gastrointestinal tract, genitourinary tract, breast, cervix, salivary glands, mediastinum, pleura, peritoneum, and bile duct (5). Myeloid sarcomas may occur at diagnosis of AML or may precede the diagnosis; they have also been seen in association with myelodysplastic syndromes or myeloproliferative disorders and usually predict transformation to acute leukemia (6). The diagnosis should be suspected if eosinophilic myelocytes are present in hematoxylin and eosin-stained biopsy sections. Imprint preparations can be helpful. The diagnosis can be made if Auer rods are detected or if the myeloid sarcoma is confirmed by cytochemical or immunohistochemical methods. Although they are radiosensitive, systemic chemotherapy is warranted in most cases. Neutropenia is common in patients with myeloid sarcoma; the marrow may be hypocellular and often reveals increased blasts.

Acute onset bilateral proptosis in children is frequently encountered in clinical day to day practice. In younger age group the acute onset proptosis is more rapid in its progression. These lesions may be ophthalmic or non-ophthalmic in origin. Some of the commoner conditions causing bilateral acute onset proptosis in children are orbital cellulitis, fungal sinusitis with extension to orbits, metastatic neuroblastoma cavernous sinus thrombosis, inflammatory pseudotumor, thyroid ophthalmopathy, rhabdomyosarcoma, bilateral optic glioma and neuroblastoma [7]. Rarely, we encounter a case of acute myeloid leukaemia showing bilateral proptosis.

This case is extremely unusual as the presenting complain was that of bilateral proptosis alone; without any other systemic symptoms of AML. Proptosis in acute myeloid leukaemia is due to diffuse leukaemic infiltration of orbital muscles

Recognition of myeloid sarcoma is important, because aggressive induction chemotherapy or radiation therapy can induce complete remission. When myeloid sarcomas are clinically symptomatic, the skeleton is the usual site affected. There is more affinity for periosteal involvement and also predilection for the orbit and epidural spaces has been described in cases of CNS and skull involvement (8). The purpose of this article is to bring in consideration of acute myeloid leukemia as a differential diagnosis in a case of childhood bilateral proptosis.

### **IV. Conclusion**

Myeloid sarcoma is an uncommon malignant neoplasm associated with myeloid leukaemia showing characteristic diagnostic feature and aggressive chemotherapy early in its course has a good prognosis. Therefore, careful history and accurate investigations accompanied by high index of suspicion are needed for proper and timely management of the patient.

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