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# BILATERAL PROPTOSIS - INITIAL AND RARE PRESENTATION: AML

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**ABSTRACT** We report bilateral proptosis as the initial presentation of Acute Myeloid Leukemia (AML) in a child. An Eight year child presented with a history of painless proptosis in the both eyes within 10 days. Radiological investigation (CT scan) showed infiltration of orbit with the metastatic tumour cell. AML was diagnosed with complete blood count, General Blood Picture (GBP) and bone marrow biopsy. The presumptive diagnosis of leukemic infiltration of the orbit is made. We report this case as AML can rarely present in child as a bilateral proptosis due to leukemic infiltration. Urgent treatment modality for this rare condition is radiation.

**KEYWORDS :** AML, proptosis

**INTRODUCTION -**

Leukemia is a malignant proliferative disorder of leukopoietic bone marrow stem cells. It is characterized by over-crowding of the bone marrow by immature neoplastic leukocytes and widespread infiltration of organs, tissues, and peripheral blood with immature leukocytes[1]

The ophthalmic manifestations of leukemia could result from primary/direct leukemic infiltration of ocular tissues or secondary/indirect ocular involvement following systemic leukemic changes. Primary leukemic infiltration can present in three patterns as anterior segment uveal infiltration; orbital infiltration including chloromas, spontaneous hyphema, orbital hemorrhages, and proptosis; and neuro-ophthalmic signs of central nervous system (CNS) leukemia that include optic nerve infiltration, cranial nerve palsies, and papilledema[2]

Acute myeloid leukemia (AML) can involve the orbit as a solid tumour termed myeloid sarcoma or chloroma[3-5]. We herein describe a case of a child who was seen with bilateral proptosis which was the initial manifestation of AML.

**CASE REPORT-**

We report the case of metastatic tumour of orbit. An eight year old child presented with sudden onset of bilateral proptosis of 10 days duration in which proptosis started in right eye and then involved left eye with fever on and off. There was difficulty in closing the eyes (lagophthalmous). Initial diagnosis was made of orbital cellulitis and child was started on broad spectrum antibiotics and anti-inflammatory drugs but not responded to treatment. Ocular examination revealed 15mm proptosis in right eye and 8mm proptosis in left eye. Visual acuity was 6/12 in both the eyes. Ocular movements were restricted bilaterally in all directions of gaze. The anterior segment and fundus examination of both the eyes was unremarkable.

**Figure 1- Bilateral proptosis (right eye > left eye)**

Complete blood count, CT Head, General Blood picture (GBP) and bone marrow biopsy was done. Based on the clinical findings and imaging study results, the differential diagnosis included leukemia, lymphoma metastatic neuroblastoma and idiopathic orbital inflammation (inflammatory pseudotumor). The initial complete blood count revealed an elevated blood cell count of 67000cells/mm3, with a differential count of 50% segmented neutrophils, 20% lymphocytes, 3% Eosinophils, 02% Monocytes, platelet count of 23000cells cumm.

In GBP – RBCS are predominantly microcytic hypochromic, TLC is markedly raised and immature forms of myeloid series predominantly myeloblasts, promyelocytes and myelocytes, very few mature cells of myeloid series are identified. Lymphocytes and monocytes show normal morphology and maturation. Platelet counts are markedly reduced and show normal morphology and maturation. No hem parasite was seen. These findings are suggestive of Acute Myeloid Leukemia. According to FAB classification it is AML-M2/M3.

Thyroid profile was done in which serum T3 was 0.70ng/ml; serum T4 was 7.11µg/dl; serum TSH 0.76µlu/ml. Thyroid profile was within normal range.

Bone Marrow Aspiration revealed 8% myeloblasts, 54% promyelocytes, 48% myelocytes and metamyelocytes, 42% band forms and mature neutrophils, 0% Eosinophils, 0% Basophils, 2% Monocytes and 28% Lymphocytes. Impression is of Acute Promyelocytic Leukemia, AML M3.

On CT scan (serial 5.0mm cuts were taken through posterior fossa and supratentorial compartment) –homogenous soft tissue is seen in superior and medial extraconal spaces of bilateral orbit, no evidence of bony erosion causing bilateral proptosis. Similar soft tissue is seen in left infratemporal fossa likely neoplastic lymphoma.

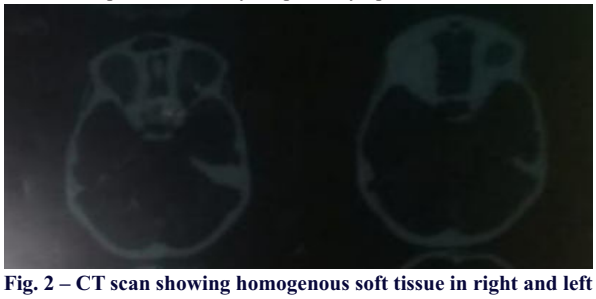

**Fig. 2 – CT scan showing homogenous soft tissue in right and left extraconal compartment superiorly placed. The lesion is limited to extraconal compartment. Bilateral proptosis is present.**

A confirmatory bone marrow biopsy was performed and diagnosis of

AML was made.

On systemic examination, there was absence of hepatomegaly and splenomegaly.

The treatment included systemic antileukemic chemotherapy, where the standard regimen for AML was given in the paediatric oncology unit. The regimen was two cycles of highly intensive chemotherapy. The first cycle of chemotherapy consisted of allopurinol, high doses of vitamin A, dexamethasone and intrathecal methotrexate; daunorubicin hydrochloride, and intrathecal vidarabine for central nervous system prophylaxis.

## DISCUSSION-

Orbital myeloid sarcoma (chloroma) as an initial symptom of acute myeloid leukemia (AML) is a rare medical condition. It is well known that AML can be seen initially with orbital involvement, before the diagnosis of the underlying leukemia.[6-9] Soft tissue accumulations of leukemic cells were previously referred to as granulocytic sarcoma or chloroma.[10]

Myeloid sarcomas are most common in certain subtypes of AML, in particular M5a (monoblastic), M5b (monocytic), M4 (myelomonocytic), and M2 (myeloblastic with maturation).[11]

The main conditions that can cause bilateral orbital masses in children are idiopathic nongranulomatous orbital inflammation, metastatic neuroblastoma, and myeloid sarcoma. Paediatric idiopathic nongranulomatous orbital inflammation is initially unilateral in 10% of cases, but it can eventually show bilateral involvement in 46%. However, involvement of the second eye is usually sequential and not simultaneous. Orbital metastasis is the initial sign of abdominal neuroblastoma in 3% to 4% of patients and is bilateral in 50%.Our patient had bilateral orbital involvement by myeloid sarcoma. Although myeloid sarcoma is a relatively uncommon paediatric orbital tumour, it becomes a major diagnostic consideration in the setting of bilateral orbital involvement. Published reports on orbital myeloid sarcoma have not always provided complete details with regard to initial features and laterality.[12,13,14]

Orbital involvement by acute myeloid sarcoma is relatively rare among orbital tumours and pseudotumors. However, in the presentation of simultaneous bilateral orbital tumours in children, myeloid sarcoma appears to be a highly likely. Any child with an orbital mass of uncertain origin should undergo prompt evaluation for underlying AML.[15]

The direct infiltration of the orbits in leukemia can present with proptosis, chemosis, lid edema, intra-retinal macular or sub-hyaloid hemorrhages, cotton wool spots, blurring of vision, diplopia, palsies of the extra-ocular muscle, or papilledema due to the increased intracranial pressure. Most cases of orbital involvement in AML present with unilateral proptosis, [16-20] and few case reports have reported bilateral orbital involvement as an initial manifestation of AML.[21,22]

**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.

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**COMPETING INTEREST STATEMENT -** There are no competing interests for any author.

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