

Case Report

Bilateral proptosis as initial sign of acute myeloid leukemia: case report and review of literature

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ABSTRACT

We report bilateral proptosis as the unusual initial presentation of Acute Myeloid Leukemia (AML) in a child. A six years child presented with a history of painless proptosis in the both eyes with fever within 15 days. Radiological investigations were showing infiltration of orbit with the metastatic tumour cell. AML was diagnosed with complete blood count and bone marrow biopsy. The presumptive diagnosis of leukemic infiltration of the orbit and EOM was made, and urgent radiotherapy, allopurinol with high doses of vitamin A, intrathecal methotrexate, and intravenous daunorubicin were instituted. Proptosis was started to decrease. We report this case as AML can rarely present in child as a bilateral proptosis due to leukemic infiltration. Radiation treatment should be considered as an urgent treatment modality for this rare condition.

Keywords: AML, Metastatic tumour, Proptosis

INTRODUCTION

Metastatic cancer probably represents the most common form of intraocular malignancy. However in practice of ocular oncology it is not encountered as frequently as uveal melanoma, possibly because many affected patients have advanced systemic cancer & do not come to the attention of ophthalmologist.

Most paediatric orbital tumours are unilateral, and little is mentioned in the literature of the frequency and differential diagnosis of bilateral paediatric orbital tumours.

Acute myeloid leukemia (AML) can involve the orbit as a solid tumour termed myeloid sarcoma or chloroma.¹⁻³ We herein described a child who was seen with bilateral proptosis that were the initial manifestation of AML. A literature review suggests that leukemia might be the most likely diagnosis in a child with bilateral soft tissue orbital tumours, a point that has not been widely recognized.

CASE REPORT

We report the case of metastatic tumour of orbit. A six year child presented with proptosis in RE with fever on and off. Initial diagnosis made was orbital cellulitis and child was started broad spectrum antibiotics and anti-inflammatory drugs but not responded to treatment and within two weeks there was development of proptosis in LE. Ocular examination revealed five mm proptosis in RE initially and then developed proptosis in the LE after two weeks (Figure 1). Visual acuity was 20/20 in both eyes. Fundus examination was normal. Ocular movements were restricted in RE.

Then complete blood count, CT head (Figure 2), MRI was done and bone marrow biopsy⁹ (Figure 3) was done. Based on the clinical findings and imaging study results, the differential diagnosis included leukaemia, lymphoma, metastatic neuroblastoma, and idiopathic orbital inflammation (inflammatory pseudotumor). The initial peripheral blood cell count revealed an elevated white

blood cell count of $24.9 \times 10^3/\mu\text{L}$, with a differential count of 3% segmented neutrophils, 52% lymphocytes, 18% monocytes, 5% promyelocytes, and 22% blast cells (Figure 4), which was strongly suggestive of leukaemia. No anaemia or thrombocytopenia was found. Serum chemistry studies disclosed a markedly elevated lactate dehydrogenase level of 1138 U/L.



Figure 1: Bilateral proptosis (Right eye > Left eye).



Figure 2: Transverse computed tomographic scan of the orbits shows bilateral superior orbital masses. The mass on the right eye was larger and more clearly seen.

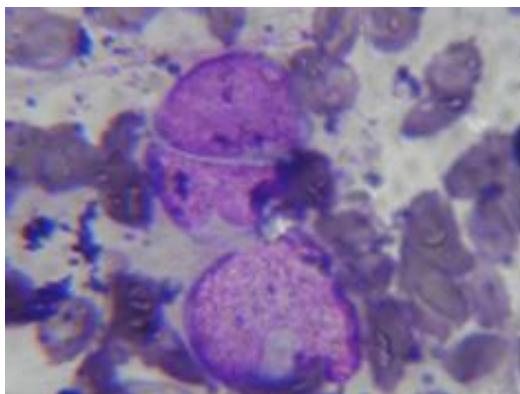


Figure 3: Bone marrow biopsy specimen shows proliferation of poorly differentiated cells with irregular nuclear contours, typical of monoblasts (hematoxylin-eosin stain).

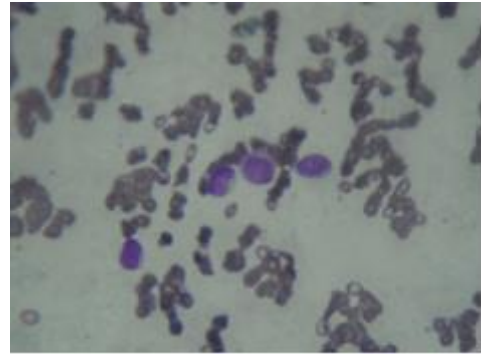


Figure 4: Peripheral blood smear.

A confirmatory bone marrow biopsy was performed and diagnosis of AML was made.

The patient was treated under the current protocol of the children's oncology group for newly diagnosed AML, which consists of two cycles of highly intensive chemotherapy, followed by an allogeneic bone marrow transplantation if a suitable donor can be found. The first round of chemotherapy was started and consisted of allopurinol with high doses of vitamin A along with intrathecal methotrexate and dexamethasone, daunorubicin hydrochloride, intrathecal vidarabine for central nervous system prophylaxis and. urgent radiotherapy was instituted. Proptosis started decreasing with the treatment of chemotherapy.

DISCUSSION

According It is well known that AML can be seen initially with orbital involvement, before the diagnosis of the underlying leukemia.^{1,2,4,5} Soft tissue accumulations of leukemic cells were previously referred to as granulocytic sarcoma or chloroma.¹⁻⁴

Myeloid sarcomas are most common in certain subtypes of AML, in particular M5a (monoblastic), M5b (monocytic), M4 (myelomonocytic), and M2 (myeloblastic with maturation).⁹

The French-American-British cooperative group defined this subtype of AML, which is also referred to as acute monocytic leukemia, as having a bone marrow biopsy specimen showing 80% or more of the nonerythroid cells demonstrating monocytic lineage (therefore, less than 20% are of granulocytic lineage). In addition, fewer than 80% of the monocytic lineage cells must be monoblasts (i.e., maturing promonocytes are clearly evident). When 80% or more of the cells are monoblasts, the lesion is classified as acute monoblastic leukemia (M5a).⁹

In most instances, orbital myeloid sarcoma occurs in young children. It is rare among the orbital tumours of childhood, accounting for only 1 of 250 cases in a previous report.⁵ The disease is relatively uncommon in the western hemisphere, but is more prevalent in the

middle East, Asia, and Africa.¹ Most of the larger reported series have come from Turkey¹⁰ and India.

When evaluating an orbital mass in a child, the ophthalmologist must consider a variety of benign and malignant conditions, particularly inflammatory, cystic, and vascular lesions such as idiopathic orbital inflammation, dermoid cyst, capillary haemangioma, lymphangioma, and others.⁴⁻⁶ About 90% to 95% of orbital masses of childhood that come to biopsy prove to be benign on histopathologic examination.⁵

Of the 5% to 10% that are malignant, rhabdomyosarcoma is the most common disease.^{5,6}

Most childhood orbital tumours are unilateral.¹⁻⁵ Most benign conditions, like dermoid cyst, capillary haemangioma, lymphangioma, and optic nerve glioma, usually affect only a single orbit. Rhabdomyosarcoma, the most common malignant orbital tumour of childhood, is invariably unilateral.⁶

The main conditions that can cause bilateral orbital masses in children are idiopathic nongranulomatous orbital inflammation,⁵ metastatic neuroblastoma,¹⁰ 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%.¹⁰

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.

However, study carried out by Jerry A. Shields, MD; Gary A. Stopyra, MD; Brian P. Marr, MD; Carol L. Shields, MD, Philadelphia, on the basis of a review of the available literature, calculated that about 88% of cases with proptosis that are seen by the ophthalmologist have no history of leukemia at the time of presentation. In addition, they estimate that about 60% of orbital myeloid sarcomas are bilateral.

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.

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