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Unilateral Orbital mass in a 3-Year-Old Boy of Acute Lymphoblastic Leukemia : A Rare Presentation

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ABSTRACT

Background & objective: We report a case of a 3-year-old boy who presented with left eye swelling, low-grade fever, pallor, and generalized lymphadenopathy. He was diagnosed with a case of B-cell acute lymphoblastic leukemia (ALL) on the basis of morphology and immunophenotyping of bone marrow. Orbital manifestation is rarely seen in ALL; rather it is more common in acute myeloid leukemia (AML). Knowledge regarding the unusual presentation of ALL will help timely diagnosis and treatment which is crucial for preserving the eye as well as the vision.

Keywords: Orbital mass; Childhood; Acute Lymphoblastic Leukemia etc.

CASE REPORT

INTRODUCTION:

Acute lymphoblastic leukemia (ALL) is the most common childhood malignancy accounting for about 25% of childhood cancer.[1] Features of leukemic infiltration of extramedullary sites are found commonly in the liver, spleen, lymph node, CNS, and testis.[2] Other sites like skin, bone, and GI tract are less common.[3]

Ocular manifestations in leukemia can occur due to direct infiltration of the orbits presenting as proptosis, lid edema, and chemosis.[4] Vascular abnormalities of the retina may also occur; presenting as intra-retinal macular or sub-hyaloid hemorrhages, cotton wool spots causing blurring of vision, and diplopia.[5] Occult ocular involvement is the third most common site of extramedullary infiltration after the CNS and the testis, which is diagnosed in up to one-third of the children with ALL by cautious investigation.[6] Proptosis is commonly seen in acute myeloid leukemia (AML), also called granulocytic sarcoma. Orbital mass is also a common finding in orbital cellulitis, orbital rhabdomyosarcoma, retinoblastoma, lymphoma, & metastatic neuroblastoma. In these circumstances, prompt treatment is of utmost importance for preserving the eye and vision by preventing ophthalmic artery and optic nerve compression or invasion. Herein we present a 3-year-old boy who came with a left-sided orbital mass, fever, and hepatosplenomegaly. He is diagnosed as a case of acute lymphoblastic leukemia (precursor B cell lineage) by bone marrow morphology and immunophenotyping.

CASE PRESENTATION:

A 3-year-old boy presented with a history of painless swelling of the left lower eyelid and low-grade intermittent fever for 4 months. During the period of illness, he was transfused with whole blood several times. He was treated with injectable antibiotics but no improvement occurred. A biopsy was done on the tissue specimen taken from the lower eyelid, but no specific diagnosis was established. Then the patient was taken to the Pediatric Hematology and Oncology Department of

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Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka for further evaluation and management.

He was ill-looking, febrile, and moderately pale. He exhibited cervical and preauricular non-tender lymphadenopathy. Eyelid swelling involved the left lower eyelid only. It was painless and vision was intact. There was neither white pupillary reflex nor bleeding in the anterior chamber. There was no history of preceding trauma to the eyelid. His vital signs were within the normal limit.

Investigation revealed a total WBC count 73x10^9/microliter, Haemoglobin 4.1 gm/dl, and platelet 20000 cells/cu-mm. On differential count, relative neutropenia was evident. PBF bshowed >90% atypical cells, mostly blast cells. Other biochemical examination (like S. Creatinine, Electrolytes, SGPT, Uric acid, S.Phosphate and Ca^2) was within the normal limit. Bone marrow morphology and immunophenotyping revealed ALL (B-cell lineage). Cytogenetics was not done. A cerebrospinal fluid (CSF) study showed CNS status.[1]

Pic A. ALL with orbital infiltration

Pic B. Lid swelling resolved following induction therapy

A) The patient was diagnosed finally with Acute Lymphoblastic Leukaemia (B-cell lineage) and stratified as an intermediate-risk group. He was treated according to the UKALL-2003 protocol.

B) The lid swelling completely resolved after 14 days of induction therapy. The patient is now on maintenance therapy and regular follow-up with our outpatient department.

DISCUSSION:

Leukemic presentation as an orbital mass is usually observed in acute myeloid leukemia.[7,8] Other etiologies include retinoblastoma, optic glioma, rhabdomyosarcoma, lymphoma, histiocytosis, and Ewing sarcoma. Orbital involvement in patients with ALL is extremely rare and is a diagnostic challenge.

Presentation of ALL as proptosis was reported in a limited number of cases.[7,9-17] The age group described in these cases ranged between 6 months to 16 years with no gender discrimination. The present case was diagnosed at 3 years of age. Most

of the cases of ALL with orbital involvement had unilateral involvement which is similar to our case. Bilateral orbital involvement is rare.[14] Leukemic infiltration of orbit possibly arises from an extension of the orbital wall; therefore, simultaneous ocular and orbital infiltration seems unusual. Only two case series reported simultaneous ocular and orbital involvement.[16,17] In most cases, proptosis is associated with other non-specific signs and symptoms of ALL, such as fever, which may help physicians think about systemic etiologies. In the present case, the initial presentation of proptosis in ALL was associated with fever, pallor, and a history of blood transfusion. Isolated proptosis in a previously healthy child was reported in only two studies.[12,16] The prognostic influence of orbital involvement in ALL is not known. However, ocular involvement in ALL is associated with a poor prognosis.[18]

CONCLUSION:

The ocular manifestations of ALL usually regress after chemotherapy similar to the present case. So, the right diagnosis at the right time in these patients is of immense significance to completely resolve the proptosis.

CASE REPORT

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