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Case Report

Conjunctival Mass as the Initial Presentation of Acute Lymphoblastic Leukemia in a Child

Abstract

Acute lymphoblastic leukemia (ALL) is the most common childhood malignancy with excellent survival. ALL has varied presentations. Presentation of ALL as a conjunctival mass is very rare. We report a child who presented to us with redness of the left eye and was found to have conjunctival mass. On biopsy, this mass was later diagnosed as ALL. The bone marrow examination supported the diagnosis. Timely diagnosis and initiation of treatment are essential to salvage the vision and to improve survival in such cases.

Keywords: *Conjunctival mass, extramedullary, leukemia*

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Introduction

Acute lymphoblastic leukemia (ALL) is the most common malignancy in children. It accounts for about one-fourth of all childhood cancers.[1] With current advances in treatment, the survival and cure rates of childhood ALL have improved significantly. Although ALL most commonly presents with medullary involvement, extramedullary sites may be involved as a part of the disease. Common extramedullary sites include the central nervous system (CNS) and testis. Other sites for extramedullary involvement include the skin and the subcutaneous tissue, the bone, the breast, the head and neck, and the gastrointestinal system.[2] Leukemia presenting with ocular involvement is usually seen in acute myeloid leukemia, as a chloroma or granulocytic sarcoma. It is very rare for ALL to have initial presentation as a conjunctival mass and has not been reported as a primary presentation. In such situations, a high index of suspicion and timely initiation of chemotherapy are very important for the preservation of vision and survival.

Case Report

A 7-year-old female child presented to us in February 2017 with complaints of swelling over the left eyelid and redness of left eye for 6 months [Figure 1]. There was no history of any discharge from the eyes, no diminution of vision, no pain in the eye, no fever, and no weight loss. Physical examination did not reveal pallor and edema. She had enlargement of left-sided preauricular lymph node (1 cm × 1 cm), firm, and nontender. There was no organomegaly. Respiratory, cardiovascular, and CNS examinations were normal.

Investigations revealed a hemoglobin of 13.2 g/dl, platelets of 258 × 10^3/cumm, and total leukocyte counts of 10,500/cumm. Her blood film showed normocytic-normochromic red cells. The conjunctival mass biopsy showed features of B-lymphoblastic lymphoma/leukemia [Figure 2], and the immunohistochemistry showed positivity for CD20, CD79a, Tdt, and CD99 and the MIB-1 proliferation index was high. Cellular bone marrow preparation showed near total replacement by blasts which were morphologically lymphoid, and the myeloperoxidase was negative. Flow cytometric analysis of the bone marrow showed approximately 50% blasts which were positive for CD19, CD10, CD38, CD22, CD34, cytoplasmic CD79a, and HLA-DR. The cerebrospinal fluid sample did not contain any leukemic blast cells.

She was started on a three-drug induction with prednisolone, L-asparaginase, and vincristine. The prednisolone response was good and she was treated on the standard risk arm. She did not take any active

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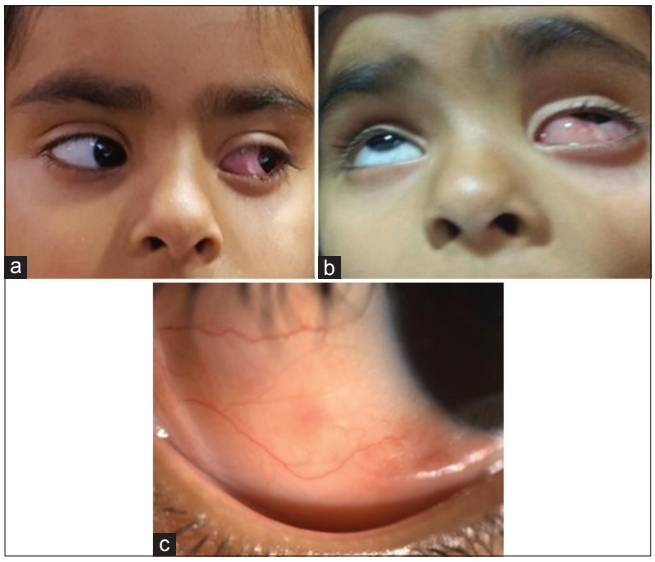

Figure 1: (a) Face photograph showing redness of the left eye; (b) showing left conjunctival mass involving inferior bulbar conjunctiva and fornix; (c) slit-lamp examination showing diffuse conjunctival mass involving inferior bulbar conjunctiva and extending into inferior fornix

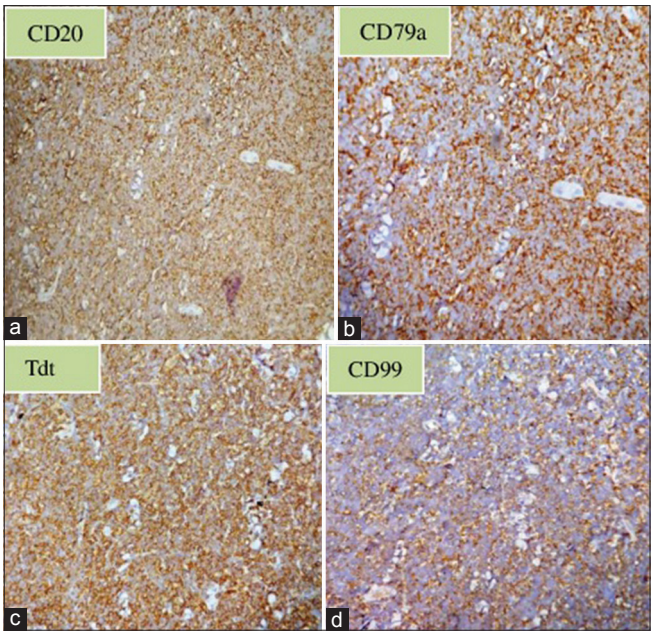

Figure 2: (a-d) The tumor cells from conjunctival mass biopsy are immunopositive for CD20 and CD79A; the tumor cells are immunopositive for Tdt and CD99

treatment for eye problem. At the end of the induction chemotherapy, the bone marrow aspirate of our patient was in morphological remission, but the minimal residual disease (MRD) was positive and the chemotherapy arm was changed to high risk. In high-risk consolidation phase, she received cyclophosphamide, L-asparaginase, cytosine arabinoside, vincristine, and 6-mercaptopurine. At the end of the consolidation phase of chemotherapy, the MRD was negative. Clinically, there was complete resolution of the conjunctival mass over a 3-month period after initiating treatment. Follow-up slit-lamp examination revealed resolution of mass lesion [Figure 3]. She is presently in the maintenance phase of chemotherapy and doing well.

Discussion

ALL arising from lymphoid precursor cells of the bone marrow can present with medullary and extramedullary involvement. Ophthalmic manifestations in acute leukemia may be due to either ocular involvement or orbital involvement. After the CNS and the testis, ocular involvement is the third most common site of extramedullary infiltration in leukemia. Primary ophthalmic involvement implies leukemic infiltration into ocular structures, orbital infiltration, or signs of CNS leukemia. Secondary involvement is due to hematological abnormalities leading to retinal or vitreous hemorrhage, infections, or ischemic changes.[3] Up to one-third of the newly diagnosed ALL may be having occult ocular involvement on vigilant ophthalmological examination.[4] Primary manifestations of conjunctiva involvement in ALL are caused by direct infiltration by blast cells.

Kincaid and Greene reported that both acute and chronic leukemia can cause ocular signs, either initially or later in the disease process. Leukemic manifestations occur most often in the retina but can also involve the choroid, vitreous, optic nerve, orbit, cornea, sclera, and anterior chamber.[5] Ophthalmic leukemic involvement can be a manifestation of primary or relapsed disease. Conjunctival infiltration is rarely encountered in acute leukemia. An autopsy study found conjunctival involvement in 4% of patients with leukemia reported by Kincaid and Greene.[5] Conjunctival mass has been reported as the presentation of ALL relapse in childhood[6] and in adults.[7] It has never been reported as the initial manifestation of ALL. Kiratli *et al*. reported simultaneous involvement of conjunctiva, uvea, and orbit as the initial sign of ALL presentation in a 9-year-old female child with unremarkable complete blood counts.[8] The index case was also a female child and had normal peripheral blood counts.

Ocular involvement is associated with a poor prognosis and increased risk of relapse.[9] Many of the ocular lesions may be asymptomatic; therefore, it is important to consider ophthalmic evaluation at the time of diagnosis of ALL as well as after completion of treatment to look for relapse. A long-term follow-up of our patient will provide more information about the disease course and survival.

Conclusion

If a patient presents with conjunctival mass, ALL should also be considered as one of the differential diagnoses and the patient should be stated on high-risk treatment arm.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

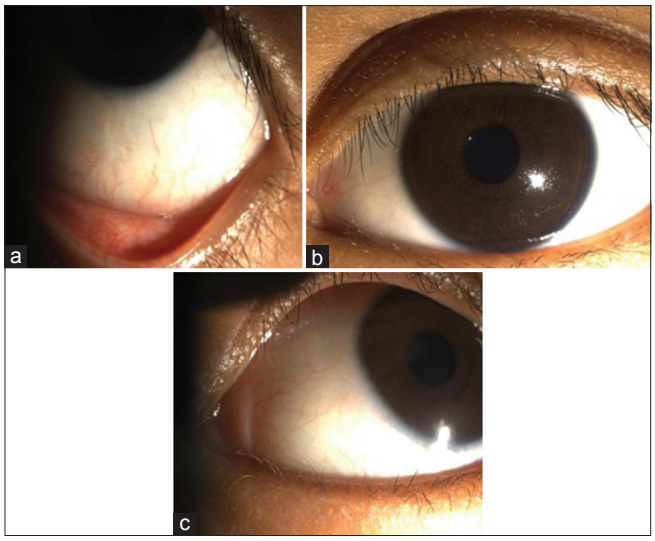

Figure 3: (a-c) Posttreatment slit-lamp examination showing resolution of the mass lesion after treatment

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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