Acute Myeloid Leukemia with Bilateral Proptosis as the Sole Presenting Sign

Dr. Anju Kochar*, Dr. Sarita Maharia**, Dr. Nabab Ali Khan***
Department of Ophthalmology,S.P.Medical College,Bikaner

Abstract

Acute myeloid leukemia (AML) accounts for nearly 15% of all leukemias in children[1]. The leukemic cells can infiltrate any extramedullary site, tumorous accumulations within soft tissues and bones being labeled as granulocytic sarcomas. Granulocytic sarcoma (GS) or extramedullary leukemic deposits is an unusual manifestation of AML, accounting for about 3% of cases of AML . Bilateral proptosis is fairly common in association with acute and chronic lymphatic leukaemia, on the other hand myelogenous leukaemia rarely give rise to proptosis. Here we present a rare case of 4year old male child presenting as bilateral proptosis with no other manifestations of systemic malignancy at presentation. Radiological investigation, peripheral blood smear, bone marrow aspiration 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 and emphasise the importance of peripheral blood smear in its diagnosis.
Keywords: Acute Myeloid Leukaemia; Proptosis, extramedullary deposits

Introduction-

Acute leukemias are the most common neoplasm seen in the paediatric population. In acute myeloid leukemia (AML),there is proliferation of malignant clones of immature myeloid cells,which replaces the bone marrow and invades other tissues of the body.[1] Primary orbital presentation without any evidence of systemic disease is only rarely seen in acute childhood leukemia and is typically due to chloroma.

Methodology

This case report highlights atypical presentation of AML in a paediatric patient . A four year old male child presented in eye department on March 2017 with proptosis in both eyes with low grade fever since 10 days. On examination, the child was irritable ,lean built weighing 7 kg and in severe agony , there was no lymphadenopathy and the vitals were stable except mild elevation in temperature.

On ocular examination the patient had severe and irreducible axial proptosis.which was tender on palpation ,there was marked fullness of both orbits,almost symmetrical with dilated vessels on upper eye lids,there was severe chemosis and cornea was exposed and dry. (Figure 1). Pupils were sluggish in reaction. Pressures were elevated in both eyes by digital tonometry and motility was extremely limited in all

*Professor , Department of Ophthalmology,S.P.Medical College,Bikaner
**Resident, Department of Ophthalmology,S.P.Medical College,Bikaner (corresponding author)
***Assistant professor, Department of Ophthalmology,S.P.Medical College,Bikaner

directions of gaze. Visual acuity could not assessed since the child was uncooperative . Fundus examination was not possible since cornea was hazy.

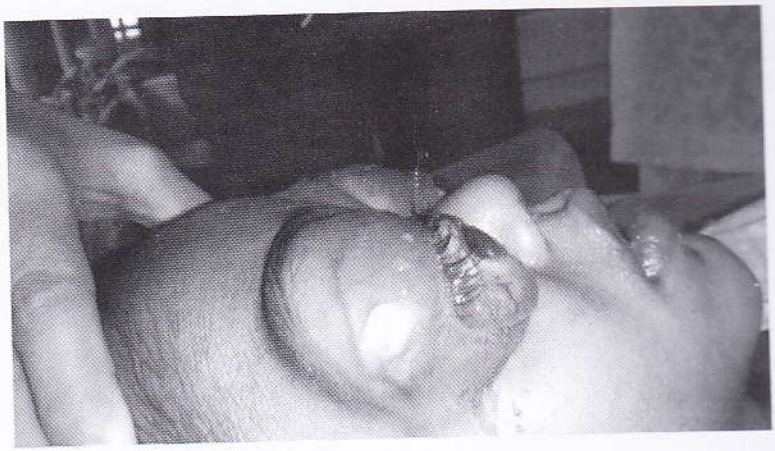

Figure-1

Provisional diagnosis was bilateral orbital cellulitis was made and child was refered to paediatric hospital and was started broad spectrum antibiotics and anti-inflammatory drugs but there was no response and proptosis continued to increase in the next few days.

Then complete blood count was done ,TLC-43,600 cell/cumm, Hb-8mg/dl,ESR-100mm/hr.MRI orbit reaveled lesion involving superior rectus,lateral rectus,bilateral lacrimal gland and the size of the lesion in right orbit was 46x37x22 mm and left orbit 39x31x20 mm and reported lymphomatous lesion/?pseudo tumour of orbits.

Based on the clinical findings and imaging study results, the differential diagnosis included lymphoma, metastatic neuroblastoma, and idiopathic orbital inflammation (inflammatory pseudotumor) and leukaemia were made.

After this peripheral blood smear for cell morphology was made and revealed moderately raised white blood cell count with a differential count of 30% segmented neutrophils, 20% lymphocytes, 6% monocytes, 4% promyelocytes, and 40%blast cells , which was strongly suggestive of leukaemia. thrombocytopenia was found. Following this peripheral blood picture a confirmatory bone marrow aspiration and biopsy was performed and bone marrow biopsy (Figure 2) showing myeloid series showing evidence of hyperplasia with presence of 40% blast cell.Erythroid series showing normal reaction (M:E=40:1) and diagnosis of AML was made.

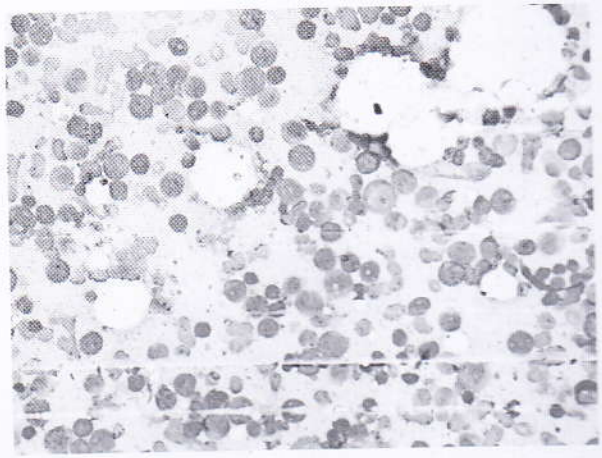

Figure -2

Since the diagnosis was confirmed by PBS and B.M biop sy,orbital tissue biopsy was foregone.

Once the diagnosis was established patients were referred to oncologist for further management and were not followed up by us.

Discussion

AML accounts for approximately15% of all leukemias in children.[1]Leukemic cells may infiltrate any extramedullary site. Granulocytic sarcoma is thought to originate in the bone marrow and the cells are believed to spread via the Haversian canals to collect in the subperiosteum and form a soft tissue mass[3].They more commonly affect the skeletal system, commonly the ligaments or periosteum. In cases with head and neck involvement they commonly affect the orbit or epidural space [6]. These tumors most commonly affect the skull,orbit, paranasal sinuses, spine, ribs, sacrum and sternum, involvement being related to the active hematopoeisis at these sites[3]. It can also involve the lymph nodes, skin and kidney [7]. This tumor can present prior to, concomitantly or even during remission of systemic leukemia[8].The presence of unilateral and bilateral proptosis has been reported with AML.[3,4] . the diagnosis of such tumor can be challenging especially when there are no signs of systemic leukemia. In the presence of systemic malignancy, a peripheral blood smear or a bone marrow biopsy may provide useful clues to the diagnosis.

Children with bilateral proptosis should always undergo systemic evaluation and blood investigations to rule out not only AML but other diseases as well apart from imaging studies because it is difficult to differentiate infective conditions and other malignancies even on CECT and MRI[2]

Peripheral smear is an invaluable ,non invasive,un expensive,very reliable tool in diagnosing of grave systemic form of AML.It shows immature blast cells with a high total leukocyte count and relative neutropenia. Leukemic proptosis, however, may not always be associated with leukocytosis or immature cells in the peripheral smear.

Doing a peripheral smear along with bone marrow aspirate and biopsy in all patients of AML manifesting with proptosis in the pediatric age is, therefore, justified.[5]
Although rare, in a child with the sudden onset of proptosis without any other systemic findings, the diagnosis of acute leukemia must be considered.

Conclusion-

Granulocytic sarcoma is a rare cause of childhood proptosis.When child present with rapidly growing orbital mass or orbilateral proptosis, AML should be kept in mind in differential diagnosis. For early diagnosis of AML, radiological imaging, peripheral blood smear along with bone marrow aspirate should be performed in all cases.If the diagnosis can be establish by a non invasive test like peripheral blood smear and one can avoid surgical intervention.
We report this case to increase the awareness of the pediatricians and oncologists regarding the unusual presentation of this rare neoplasm.

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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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