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Case Report

Unilateral Ptosis - An Unusual Initial Presentation of Acute Myeloid Leukaemia in an Adolescent Girl

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I N F O

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A B S T R A C T

In case of acute myeloid leukaemia (AML), there is increased multiplication of blast cells forming the malignant clones of immature myeloid cells in the bone marrow in different body tissues and also enormously carried by blood. Here, we report an 18-year-old adolescent girl who presented with unilateral ptosis. Magnetic resonance imaging (MRI) of brain showed right orbital anteromedial mass. Bone marrow examination showed myeloblasts and metamyelocytes suggestive of AML. Patient was started on chemotherapy but could not survive the infections during the hospital stay.

Keywords: AML, Chloroma, Ptosis, Orbital Mass

Background

Incidence of acute myeloid leukaemia (AML) is 15% among all types of leukaemia in children.[1] It is more common in older age groups. Immature hematopoietic precursor cells form a tumour on orbits called granulocytic sarcoma (GS) and it is considered as a complication of AML.[2] AML primarily affects children less than 10-year age and they are more frequently having granulocytic sarcoma in orbit, like so many other tumours of orbit. It causes difficulty in making the diagnosis. Here, we report a case of young girl presenting with unilateral ptosis without any other systemic complaints.

Case Presentation

An 18-year-old girl presented to Medicine outdoor with complaints of gradually progressive drooping of right eye lid and diplopia from 15 days. On examination, she was conscious and oriented. There was drooping of right eyelid without any sign of inflammation (Figure 1).

There was no history of fever or focal neurological deficit in any other part of body. On the side of ptosis inter palpebral fissure was 7mm from upper eye lid margin and on left side it was 10mm. Ocular motility was normal on both the sides. Her blood pressure was 100/ 70 mmHg, pulse rate was 88 beats/minute, regular. Pallor was present.

Left eye was completely normal. Rest of the systemic examination was within normal limits. Lab investigations showed pancytopenia with haemoglobin (Hb) of 4.4 gm%, total leukocyte count (TLC) of 2700/ µl, polymorphs of 56% and lymphocytes 39% with many atypical lymphocytes and blasts in peripheral smear. Platelet count was 51,000/ µl. As she was a vegetarian, we did vitamin B_{12} and folic acid level to look for the aetiology and found it within normal limits. Bone marrow biopsy showed myeloblasts (>35%) and metamyelocytes in the bone marrow suggestive of acute myeloid leukaemia (Figure 2 A & B).

Figure 1.Right sided ptosis

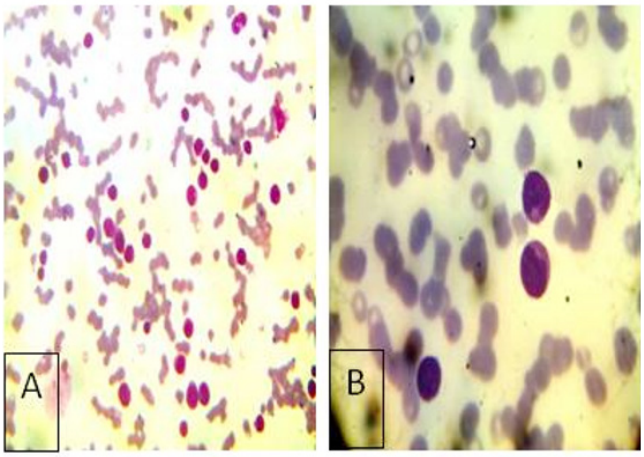

Figure 2(A&B).Blood smear of acute myeloid leukemia showing myeloblasts and metamyelocytes [Leishman stain, X20&X40]

The diagnosis was further confirmed by flow cytometric analysis. We decided to do MRI of orbits in view of ptosis, which showed a mass lesion of 11mm diameter anteromedial of right orbit. There was no involvement of brain, extra ocular muscles, lacrimal gland and optic nerve. Removal of orbital mass by excisional surgery was suggested. Patient was referred to department of haematology where chemotherapy with daunorubicin and cytosine arabinoside was started. But due to secondary infections she expired after five days of starting chemotherapy.

Discussion

Granulocytic sarcoma (GS) was previously known as 'Chloroma' which is collection of leukemic cells in soft tissues and is an unusual leukemic manifestation.[3] Only 5% of Caucasians cases of AML have unique presentation of GS in the form of leukemic infiltration.[4] Excess amount of greenish hue of myelo-peroxidase in GS, named it 'chloroma' which also may be of other so many colours like white, gray, or brown, on the basis of oxidative state of myelo-peroxidase enzyme. Hence, a more general terminology 'granulocytic sarcoma' (or occasionally 'myeloid sarcoma') is now in use. GS has significant geographic and racial variation. In child age group, favourite site for GS is orbit. Association of OGS (Orbital GS) with those AML cases demonstrating at (8; 21) translocation is very strong and have better prognosis. Pathogenesis of making soft tissue mass by GS starts from bone marrow and spread via haversian canals to penetrate the periosteum. This is the reason for typical bony location of the mass like skull, sinuses, orbits, spine, ribs, sacrum, and sternum. There are varied presentations of GS and the most common being proptosis. Others are lacrimal gland involvement, eyelid tumor, ptosis, iris tumor, uveitis, conjunctival mass, and scleral mass.[5] In case of absence of systemic manifestations of AML, making the diagnosis of GS is not easy. Bone marrow, muscles and white matter at T1-weighted MRI, are slightly hypersignal while at T2-weighted imaging, these are isosignal in case of OGS. OGS presents as isodense to muscle on CT scan. Suspicion of orbital myeloid sarcoma may be raised due to clinical and lab investigations but biopsy remains confirmatory. Increased TLC along with >5% peripheral and >30% medullar blasts cells and Auer rod presence is pathognomic of AML. In comparison to many other subtypes of AML, prognosis for patients with GS is better. It becomes even better if the blasts cells in marrow are less than 5%.[6] Early diagnosis and initiation of therapy in case of OGS is important and rewarding.

Differential Diagnosis

Retinoblastoma the commonest orbital tumor in children, occurs within the globe, rhabdomyo-sarcoma, Ewig's sarcoma, orbital lymphoma, neuro-blastoma, orbital hemangioma, and inflammatory pseudotumor of optic nerve are the various differentials of OGS which can present in similar manner and should be ruled out.

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Conflict of Interest: None

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