

Chloroma of the orbit as a presenting feature of acute myeloid leukemia in a four year old female child

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Abstract

A four year old female child presented with a painless swelling of the right lower lid for 25days, which was progressively increasing in size. On palpation, a firm nodular mass just above the inferior orbital margin was noted. MRI showed homogenous enhancing mass in the orbit. Progressive axial proptosis developed in 8 days, with inability to close the eye. Bone marrow aspiration at this stage revealed Acute Myeloid Leukemia. Chemotherapy was started. Bone marrow picture after 10days showed remission phase. Symptoms and signs began to regress. The child was discharged on maintenance chemotherapy; but was admitted 3weeks later with fever; the child expired after admission 2 days due to secondary infection.

Key words: Acute myeloid leukemia, Chloroma, painless swelling, Eye

Introduction

Among the malignancies seen in childhood, myeloid leukemia is very common, and has a prevalence of 15-20% [1]. Acute myeloid leukemia (AML) is a condition characterized by an abnormal proliferation of malignant clones of immature myeloid cells, which replaces the normal bone marrow and begins to invade other tissues in the body.

Chloroma is a localized tumor composed of malignant cells of myeloid origin which occurs in patients with AML, and appears as a greenish yellow tumour mass which is seen to involve the bones of the skull, orbit, skin and abdomen [2].

Most commonly chloroma is seen to occur as a manifestation of established myeloid leukemia; but very rarely it precedes the diagnosis of leukemia. It has been reported that the incidence of leukemic deposits in the orbit with bilateral proptosis is 2% [3]. Among individuals in whom AML is already diagnosed, the development of chloromas indicate poor prognosis [4,5]. The presence of chloroma in a child with AML is rare; and such a case where the development of chloroma preceded the diagnosis of AML is even more rare. Hence we find the need to publish such a case in a scientific journal.

Case Report

A four year old female child presented to us with a painless swelling involving the right lower lid since 25 days, which has been progressively increasing in size.

On examination a hard nodular mass was noted under the inferior orbital margin, with edema and pigmentation of the overlying skin. Rest of the anterior and posterior segment was normal.

Urgent MRI was ordered, which revealed a homogenous enhancing mass involving the right lateral rectus, and extended infero-nasally displacing the globe supero-medially. It was seen to also involve the superior, medial and inferior recti, and extended posteriorly to indent the optic nerve head for 90 degrees. There was no obvious extension in the globe.

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Figure-1: At presentation (Day 1)

Blood investigations at this stage revealed severe anemia with a haemoglobin of 6.6% and a platelet count of 20,000.

Over the next 8 days the child developed progressively increasing axial proptosis with associated chemosis, and inability to close the eye. The child soon developed exposure keratopathy, and began to bleed from the exposed chemosed conjunctiva. Ocular movements were restricted completely. The child was pyrexix at this stage.



Figure-2: Increase in proptosis and chemosis (Day 9)

Though we considered doing a biopsy of the lesion, it was not attempted in view of severe thrombocytopenia and highly vascular nature of the lesion. Hence a Bone marrow aspiration was done at this stage, which revealed AML.

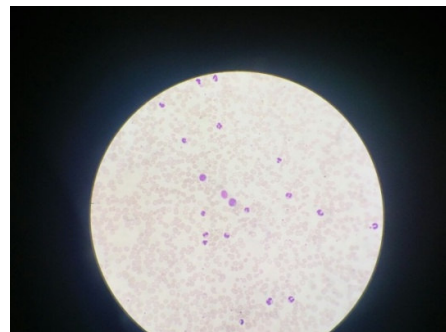


Figure-3: Bone marrow picture showing Acute Myeloid leukemia

The child was soon started on chemotherapy using cytarabine and daunomycin.

After 10 days of chemotherapy, the bone marrow aspiration was repeated, showing the bone marrow in remission phase. Also the signs and symptoms had begun to regress. There was a decrease in the axial proptosis, the chemosis was decreasing, ocular movements were returning and eye closure was possible. The platelet counts also increased to 2,60,000.



Figure- 4: Improvement in signs and symptoms after chemotherapy

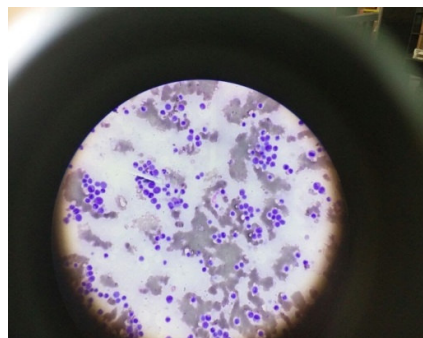


Figure-5: Bone marrow in the remission phase

The child was discharged and was kept on maintenance chemotherapy. After 3 weeks the child was brought back to the hospital with fever and cytarabine induced neutropenia. In her last few days the child was markedly toxemic, anaemic and semi-comatose. The relatives took the child home against medical advice and within 3 days we received the information of her death.

Discussion

Chloroma, also known as granulocytic sarcoma is the accumulation of leukemic cells at various sites such as abdomen, skull, periosteum, skin, paranasal sinuses, spine, ribs and orbit. It is composed of primitive granulocyte precursors namely myeloblasts, promyelocytes, and myelocytes along with supporting connective tissue and vascular stroma [4]. It was first described by Allen Burns in the year 1811. The first presentation of AML is usually fever, anemia, bleeding from various sites or enlarged lymph nodes. Chloroma is a rare presentation of AML seen in only 3% cases of AML, and chloroma of the orbit being the first presentation of AML is even rarer.

Common causes of proptosis in the first decade are dermoid, rhabdomyosarcoma, hemangioma, orbital cellulitis or lymphoma.

Orbital chloroma in children may present as a periorbital edema, or mass lesion in the region of the lacrimal gland or lids. It may be unilateral or bilateral [6,7,8,9,10]. Patients will usually have restricted ocular movements and decreased vision, as seen in our case. Bone marrow aspiration and peripheral smear are the most important diagnostic tools which help in reaching the diagnosis of AML [10]. Peripheral smear shows high total WBC count with an increase in the number of primitive blast cells and relative neutropenia.

Pathology: on gross examination the tumor is seen to have a predilection towards skeletal system. The bone marrow becomes hyperplastic and there is an increase in the primitive blast cells. In our case the blast cells were more than 60%, which subsequently reduced following

chemotherapy. Such tumors respond very well to chemotherapy and show signs of regression soon after starting therapy. However bone marrow transplant still remains the treatment of choice [11]. Similar studies have been reported by Gupta et al and by Ansari S et al in a 2 year old and 6 year old child respectively, who presented with bilateral proptosis as a first presentation of AML, where the diagnosis of AML was made after peripheral smear, bone marrow aspiration study and radiological imaging [11,12].

Conclusion

Hence we may conclude that AML has to be kept in mind as a differential diagnosis in a child presenting with unilateral or bilateral proptosis, and all such children should be subjected to peripheral smear examination and radiological imaging, along with bone marrow aspiration study where ever necessary. Also it is important to diagnose such cases as early as possible as they are highly responsive to chemotherapy.

It should be noted that the definitive treatment still remains bone marrow transplant.

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