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ORBITAL CHLOROMA IN ACUTE MYELOID LEUKAEMIA PRESENTING AS UNILATERAL PROPTOSIS – A RARE CASE REPORT

Ophthalmology

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ABSTRACT

Aim: To document a case presenting with ocular manifestations in acute myeloid leukaemia(AML).

Materials and methods: A 4 year old male child presented with severe pain, watering, bulging of right eye.On clinical examination of right eye vision was no perception of light, axial proptosis, necrotic cornea due to exposure keratitis. CT Scan and MRI revealed retro-orbital mass encasing right optic nerve and extending upto right cavernous sinus and paranasal sinuses and intra-cranially into anterior temporal region. Peripheral blood smear showed atypical blast cells suggestive of acute leukaemia. Bone marrow smear showing 90% myeloblasts and flow-cytometry confirmed diagnosis as AML-M2.

Result: patient was put on BFM-87 chemotherapy protocol and after 3 months the proptosis has reduced.

Conclusion: This case emphasizes that childhood proptosis can rarely be initial manifestation of AML. It shows the aggresive nature of the disease and if presented early, with vigorous treatment one can salvage both eye and visual acuity.

KEYWORDS

Acute Myeloid Leukaemia, Chloroma, Chemotherapy

INTRODUCTION:

Chloroma or Myeloid Sarcoma or Granulocytic Sarcoma is an extramedullary manifestation of Acute Myeloid Leukaemia (AML), so they are solid collection of leukaemic cells occurring outside bone marrow .[1]About 3-8% of AML present with myeloid sarcoma .[1] Chloromas may be found in variety of locations like – lungs, lymph nodes, small intestine and also orbit . Generally chloromas are found unilaterally with sequential involvement of second eye .[2]Aim of the study was to document the clinical course of the orbital chloroma.

CASE REPORT:

A four year old male child presented with unilateral protrusion of right eye of three months duration. The protrusion was gradually progressive and at the time of presentation patient was unable to close his eyes. Visual acuity in right eye was no perception of light (PL Negative) and left eye was 20/20(6/6) Ocular examination revealed in right eye - axial, irreducible ,non pulsatile, tender proptosis with ulceration. Eyelids were everted. Eye movements were totally restricted. and in left eye– anterior and posterior segments were within normal limits with normal intraocular pressure.(fig- 1,2).On systemic examination– fever, bleeding gums and pallor were noted.

CT SCAN and MRI orbit revealed -- retro orbital soft tissue mass encasing right optic nerve and extending upto right cavernous sinus, supra-zygomatic space, right paranasal sinuses and intra cranially into anterior temporal region. (Fig –3,4) Routine blood investigations revealed :RBC count – 2.08 million/cu.mm, haemoglobin – 6.3gm% (anaemia) and platelet count to be 33 thousand/cu.mm signifying thrombocytopenia .

Peripheral blood smear also showed bicytopenia (anaemia and thrombocytopenia). Blast cells were found (27%), which was suggestive of acute leukaemia. (Fig -5) Bone marrow smear showed - hypocellular marrow with predominant myeloblasts(90%). Increased myeloid : erythroid ratio. Hence the impression was acute myeloid leukaemia (Fig -6,7,8)

Other investigations were done like - CSF fluid cytology :paucicellular smear with occasional lymphocytes. No malignant cells found. And to find out about the spread of leukaemia-X Ray chest, USG whole abdomen, liver function test, renal function test were done and all were found within normal limits.

Flow Cytometry of T-Cell , B- Cell Markers clinched the detailed diagnosis.it showed- bright expression of CD 34,moderate expression of CD 117, CD 13, HLA-DR, MPO,dim expression of CD 45,Aberrancy detected in CD 19. Impression was Acute Myeloblastic Leukaemia with maturation (AML – M2).

Patient was put on BFM-87(Berlin Frankfurt Munich) chemotherapy protocol Currently patient has completed three months of treatment.

Induction Phase:

Cytarabine, Daunorubicine, Etoposide.Consolidation Phase - Prednisolone, Vincristine, Cyclophosphamide, Cytarabine, Methotrexate.Now he is awaiting the cranial irradiation phase. Delayed presentation has led to loss of vision in right eye.(fig -9)

DISCUSSION:

Myeloid sarcoma or chloroma or granulocytic sarcoma is a solid tumour composed of immature white blood cells (WBC) called myeloblasts.[1] The term chloroma came from Greek word chloros meaning green as these tumours, due to presence of myeloperoxidase have a green tint and was first described by British physician Allen Burns in 1811.[5] In 1967, Rappaport coined the more correct term 'granulocytic sarcoma' as 30% of these tumours can be brown, white or grey.[6] More recently, the term 'myeloid sarcoma' has been favoured.[7] Orbital chloroma is a disease of children and young adults, with vast range of age from infancy to 61 years , with mean age being 7years. [8]

Chloromas are more common in patients with **AML-M2** (French-American –British (FAB) classification) , with myeloblasts expressing CD13 or CD14 and with high peripheral WBC count [3]. Definative diagnosis of chloroma is the biopsy of the lesion. But nowadays immunohistochemical staining using monoclonal antibodies against CD34 and CD117 are mainstay of diagnosis. Systemic chemotherapy for leukaemia is considered first line of treatment for chloromas. These features have been noted in this case .Presence of chloromas augur poor prognosis, with poorer response to treatment and worse survival .[4]

CONCLUSION:

This case emphasizes that childhood proptosis can rarely be initial presentation of AML.It is challenging for an ophthalmologist as chloromas can occur without a known pre-existing or concomitant diagnosis of leukaemia. Since it is associated with poor prognosis, hence careful history and correct investigations with degree of suspicion are needed for diagnosis. This case also highlights the aggressive nature of the disease and with prompt diagnosis and vigorous treatment can lead to good ocular remission.

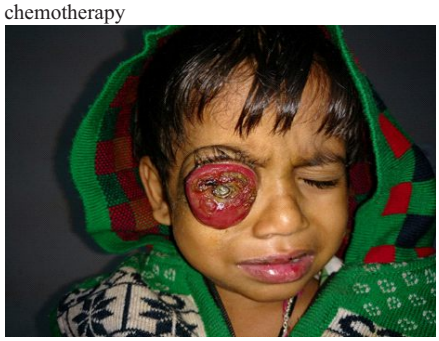
Legends for pictures:

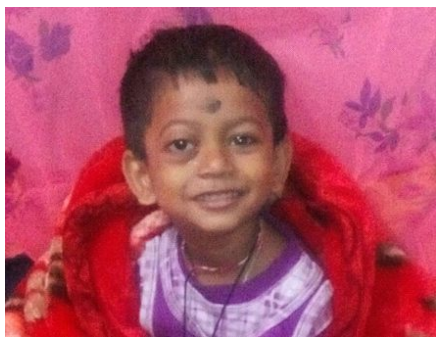
1. At presentation
2. 2 months before presentation, swelling of right eye noted by parents

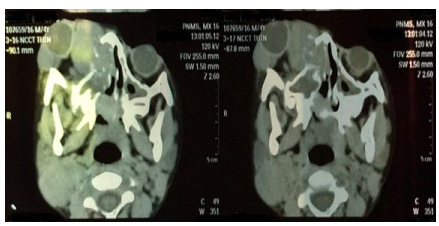
3,4. NCCT findings – show the soft tissue mass and its extension

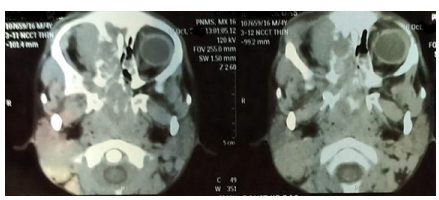
5. Peripheral blood smear – showing pancytopenia

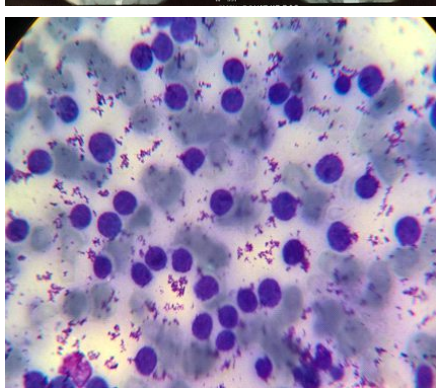
6. bone marrow smear showing myeloblasts
7. bone marrow smear showing myeloblasts
8. bone marrow smear showing myeloblasts
9. post chemotherapy

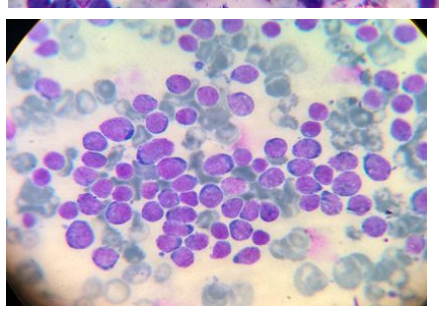


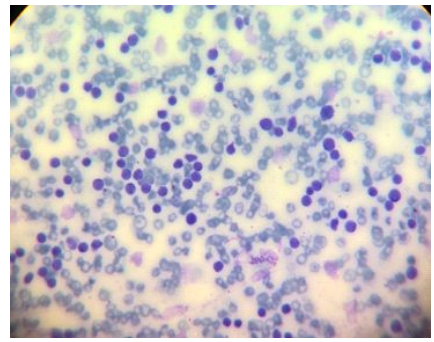


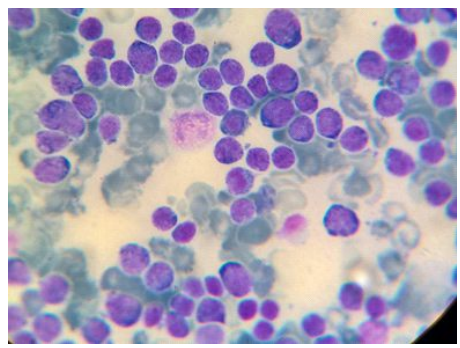












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