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Proptosis and Facial Nerve Palsy as a Sole Manifestation of Acute Myeloid Leukemia in Infant

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Abstract: *Proptosis and Facial nerve palsy as presenting features of Acute myeloid leukemia is very rare (1) and more so in infants. We report a case of five month old female infant presented with simultaneous proptosis and facial nerve palsy and diagnosed to be a case of acute myeloid leukemia as per her Peripheral blood smear (PBS) and bone marrow reports. Granulocytic sarcoma (GS) or chloroma is rare manifestation in acute myeloid leukemia occurring about 3% of childhood acute myeloid leukemias.(2,3,4) Orbital Granulocytic sarcoma may cause proptosis while leukemic deposits in temporal bone or CNS infiltration can lead to facial nerve palsy in child with acute myeloid leukemia.(5) The purpose of this case report is to keep high index of suspicion of underlying malignancy like leukemia while evaluating infants and young children presenting with proptosis and / or facial nerve palsy and highlight the importance of PBS examination & bone marrow study in these children.*

Key words: *Granulocytic sarcoma, Acute myeloid leukemia, chloroma, Proptosis, Facial nerve palsy, PBS.*

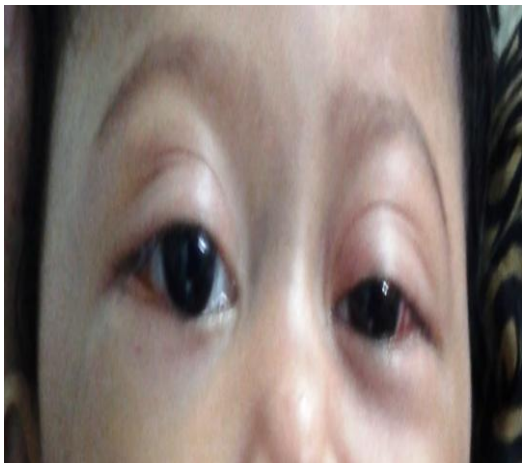
I. Introduction:

Leukemia is a most common malignancy in childhood (6) Acute myeloid leukemia accounts for 15% of all leukemia in children(7). Granulocytic sarcoma (GS) can occur uncommonly in acute myeloid leukemia. It is focal masses of immature myeloid cells from granulocytic linkage. GS can occur at any location especially skin, orbit, Para nasal sinuses, bone, respiratory tract, gastrointestinal tract, genitourinary tract, liver, lymph nodes. Leukemic deposits in temporal bone or CNS infiltration can lead to facial nerve palsy in child with acute myeloid leukemia. It can occur at any sight in course of disease or during remission or relapse. But concurrent occurrence of facial nerve palsy and proptosis is very rare presentation of acute myeloid leukemia.

II. Case History:

A 5 month old female child presented to our clinic with complaints of ipsilatral right sided facial nerve palsy and bilateral proptosis since 15 days. On examination child was anaemic. There were no signs of raised intra cranial tension. There is bilateral non reducible proptosis, no subconjuctival hemorrhage, extra ocular movements and pupillary light reflex was normal. On neurologic examination patient had right LMN type facial nerve palsy. There was no organomegaly or other systemic involvement. On investigation patient had Hemoglobin 7.7gm/dl. PBS examination showed leucocytosis with total leucocyte count of 1,18,000/cmm with 10% neutrophils, 32% lymphocytes and 22% blast cells. Thrombocytopenia with Platelet count of 58,000/cmm.

CT brain suggestive of bilateral frontoparietal subdural hygroma without significant mass effect with a soft tissue swelling in orbital region. Bone marrow aspiration showed hypercellularity with increased M:E ratio, with 25% blasts showing cytoplasmic vacuolations, 40% promyelocytes , 8% myelocytes, 5% band cells confirming our diagnosis of acute myeloid leukemia. Patient was referred to hematologist for induction chemotherapy regime.



III. Discussion

Acute myeloid leukemia can occur at any age group. Acute myeloid leukemia in children presents with hepatospleenomegaly, pancytopenia weight loss prolonged fever and many other symptoms (9). Cranial nerve involvement occasionally observed with acute myeloid leukemia in any part of disease process due to bone and nerve infiltration(5), but the involvement as a presenting feature of this serious disease is striking one. Proptosis may be unilateral or bilateral, may be seen with orbital chloroma (granulocytic sarcoma) as a unusual variant in acute myeloid leukemia as a presenting feature(9).

In our study both bilateral proptosis and right facial nerve palsy occurred simultaneously as an alarm resulting in diagnosis of acute myeloid leukemia. Age of the patient is 5 month which is also at the very extreme as very few cases are reported in infantile age group. There is no hepatospleenomegally, Lymphadenopathy or other systemic manifestations. Thrombocytopenia was noted. Peripheral blood smear and bone marrow confirmed our diagnosis. Chemotherapy is mainstay of treatment and prognosis depends on underlying systemic malignancy although early bone marrow transplant may be reassuring.

Hence I like to conclude with -

1. Acute myeloid leukemia can occur at any age.
2. Proptosis and cranial nerve involvement in the form of facial nerve palsy may be the rare presenting features of underlying malignancy hence high index of suspicion is needed more so when there is no systemic signs.
3. Simple tests like hemogram and peripheral blood smear may be helpful at times.
4. Bone marrow examination will confirm our diagnosis.
5. Chemotherapy is a mainstay of treatment.
6. We suggest complete physical examination (including liver, spleen and lymph nodes), complete hemogram with peripheral blood smear should be routinely done in all patients presenting with cranial nerve palsy and/or proptosis. At times bone marrow examination can be performed in doubtful cases
7. Keeping high index of suspicion of underlying malignancy in such cases may be worth rewarding in diagnosing such condition.

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