

Proptosis and Facial Palsy as an Unusual Clinical Presentation of Acute Myeloid Leukemia

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Abstract

Simultaneous proptosis and facial palsy as the clinical presentation of childhood Acute Myeloid Leukaemia (AML) is very rare. To date, no case have been reported anywhere to the best of our knowledge. Extra medullary leukemic deposits or Granulocytic Sarcoma (GS) is a rare manifestation in about 3% of childhood AML, 9.3% of GS manifested as orbit deposits causing proptosis in one or both eyes. CNS infiltration or acute otomastoiditis subsequent to leukemic infiltration of the temporal bone may be implicated with facial paralysis. We are here with reporting the case in a 3-year-old boy who presented with proptosis and facial palsy in a case of AML. The purpose of reporting this case is to emphasize the need of examining the peripheral blood and bone marrow in children presenting as proptosis and facial palsy for early diagnosis of childhood AML.

Keywords: Acute myeloid leukemia; Proptosis; Facial palsy; Granulocytic sarcoma

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Introduction

In children, Acute Myeloid Leukemia (AML) accounts for 15% of all leukemias. Extramedullary leukemia deposits or Granulocytic Sarcoma (GS) is a rare manifestation in about 3% of cases and 9.3% of GS manifested as orbit deposit causing proptosis in one or both eyes [1]. Facial palsy in such cases as a clinical presenting symptom is also very rare. Moreover, especially in childhood AML, the simultaneous presence of both proptosis and facial palsy have not been previously reported anywhere to the best of our knowledge. In this report, such unusual presentation is being reported.

Case Report

A 3-year old boy presented to our Outpatient Department (OPD) with progressive proptosis of left eye for the last 3 months and left facial palsy for last 20 days. On examination, he was found to be moderately anemic without petechial patches and had loss of appetite and weight. There were no symptoms of raised intracranial tension. His systemic neurological examination showed left facial nerve palsy. There was no palpable abdominal organomegaly and no lymphadenopathy.

Ophthalmological examination revealed non reducible axial proptosis of left eye without subconjunctival hemorrhage. Ocular movements were not restricted in all directions. Pupillary reflexes were positive in both eyes. His hemoglobin was 7.0gm% and the total leukocyte count was 8000/mm³, differentiated leukocyte count showed 28% neutrophils, 48% lymphocytes, 12% blast and platelet count was 20,000/mm³. The contrast CT scan/MRI of orbits showed polypoidal soft tissue mass in the left maxillary antrum showing pressure erosions of smooth thinning of the medial, posterolateral walls and boxy orbital floor at places extending into the inferior extraconal space of the left orbit with mild proptosis. The bone marrow aspiration showed hypercellular marrow smears; 15% blast, 45% promyelocytes with increased myeloid erythroid (M: E) ratio conforming to the diagnosis of acute promyelocytic leukemia (AML, M3). Incisional biopsy of the orbital swelling was avoided in view of the low platelet count and imaging report correlating with leukaemic proptosis. The brain MRI and CSF analysis were normal. Thus, the patient was diagnosed as AML, M3 with extra-medullary disease involving left orbit and left facial



Figure 1. A 3-yr-old male child showing proptosis and facial palsy



Figure 2. Axial Contrast Enhanced Computed Tomography (CECT) showing enhancing mass occupying left orbit.

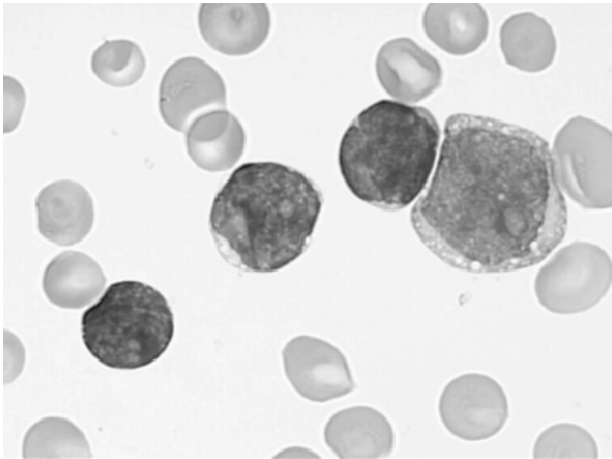


Figure 3. Microphotograph of peripheral smear showing myeloblast, (Leishman's stain, 1000X)

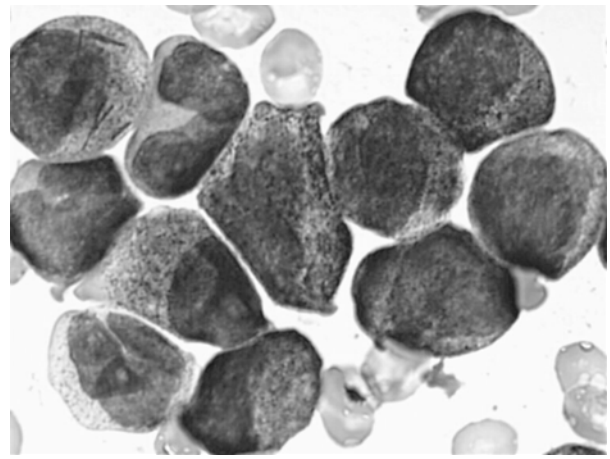


Figure 4. Microphotograph of bone marrow smear showing promyelocytes with Auer rods, (Leishman's stain, 1000X)

nerve palsy. After second cycle of chemotherapy induction cycle comprising of injection Daunorubicin 45g/m² and injection Cytarabine 200mg/m² there was partial remission of proptosis and improvement of the facial palsy. Patient is doing well up to last follow up.

Discussion

Granulocytic sarcoma even though a rare presentation can present from infancy to old age affecting commonly children and young adults.

These tumors can present prior to or concomitantly or even during remission of systemic leukemia showing its variable natural history [2, 3]. In cases with head and neck involvement, they commonly affect the orbit or epidural space and proptosis being the most common presenting feature [4].

Murthy et al [5] has reported 12 cases of extra medullary leukaemia in children who presented with only proptosis. In our case the patient was noted to have orbital disease, proptosis, facial palsy and systemic involvement concurrently.

Most of the reported cases have decreased visual acuity and restricted extra-ocular movements unlike our case. Early detection of the leukemia as well as limited orbital extension of the leukemia infiltration may be the reason.

When there are no signs of systemic leukemia, the diagnosis of this tumor can be challenging. This tumor can also be confused with rhabdomyosarcoma, neuroblastoma and lymphoma.

In case where the orbital tumor is the initial manifestation, peripheral blood and bone marrow

involvement usually occurs within a year of the occurrence of orbital disease [1, 3].

Cranial neuropathies, especially of the seventh nerve, occur in children with central nervous systemic leukemia caused by leukemic infiltration; however, they are extremely rare as the presenting sign of the disease [6]. Other than CNS infiltration, acute otomastoiditis subsequent to leukemic infiltration of the temporal bone may be implicated with facial and acoustic nerve paralysis.

Our patient had both rare clinical presentations of proptosis and facial palsy without systemic sign and symptom of AML.

Even though peripheral smear is an invaluable tool in diagnosing the systemic form of AML, smear may not always be associated with a leukocytosis or immature cells in the leukaemic proptosis. Therefore, advising for bone marrow aspirate or biopsy in all patients manifesting with proptosis in the pediatric age is very much justifiable [7]. Previous reports have highlighted the role of non invasive test to diagnose such cases of childhood proptosis [8, 9], complete hemogram and bone marrow examination were advised before any local invasive test to confirm the case of proptosis in our case. The extra- medullary leukemia on CT scan may appear as well defined lesion, isodense to muscles within the orbits and rarely extension to the paranasal sinuses as seen in our patient.

The prognosis depends on the course of underlying systemic malignancy. The presence of extramedullary leukemia does not alter the survival of patients with AML.

Chemotherapy is the mainstay of treatment but allogenic bone marrow transplantation from a matched family donor still remains the best long-term option that provides remission-free survival for most patients.

In conclusion, simultaneous presence of both proptosis and facial palsy in childhood AML, is very rare clinical presentation and possibility of granulocytic sarcoma as one of the causes should be kept in mind. Non- invasive investigations like complete hemogram along with bone marrow aspirate even if the complete hemogram is within normal limits should be performed for early detection and early treatment of this disease.

Acknowledgment

The authors would like to acknowledge the patient for his contribution.

Conflict of interest

The authors have no conflict of interest in this article.

Authors' Contribution

Dhaneshor Sharma Takhenchangbam designed and wrote the case. Rajesh Singh Laishram contributed the pathological findings and photomicrograph of the case. Akoijam Sunita contributed in the review of literature. Lanu Tiameren Imchen helped in writing and image of the manuscript. Tomcha Singh Thoudem helped in review of literature and overall correction of the manuscript.

References

1. Stockl FA, Dolmetsch AM, Saornil MA, Font RL, Burnier MN Jr. Orbital granulocytic sarcoma. *Br J Ophthalmol.* 1997; 81:1084-8.
2. Davis JL, Park DW II, Font RL. Granulocytic sarcoma of the orbit. A histopathologic study. *Ophthalmology.* 1985; 92:1758-62.
3. Zimmerman LE, Font RL. Ophthalmic manifestation of granulocytic sarcoma (myeloid sarcoma or chloroma). The Third Pan American Association of Ophthalmology and Journal of Ophthalmology Lecture. *Am J Ophthalmol.* 1975; 80: 975-90.
4. Porto L, Kieslich M, Schwabe B, Zanella FW, Lanfermann H. Granulocytic sarcoma in children. *Neuroradiology.* 2004; 46(5):374-7.
5. Murthy R, Vemuganti GK, HonavarSG, Naik M, Reddy V. Extramedullary leukemia in children presenting with proptosis. *Journal of Hematology & Oncology.* 2009, 2:4.
6. Bilavsky E, Scheuerman O, Marcus N, Hoffer V, Garty BZ. Facial paralysis as a presenting symptom of leukaemia. *Pediatr Neurol.* 2006; 34: 502-4.
7. Ghose S, Kumar R, Chaudhuri S, Jain Y, Gujral S, Singh H, et al. Importance of bone marrow examination in childhood proptosis . In *Indian Ophthalmology Year Book 1997, Proceedings of 55th All India Ophthalmology Society Conference, New Delhi, Pasricha JK (ed).* 1997: 414-16.
8. Sethi A, Ghose S, Jain P, Kumar R. Childhood proptosis: the invaluable but overlooked peripheral smear. *Indian J Ophthalmol.* 2001; 49(2):121-3.
9. Panda A, Dayal Y. Acute proptosis in myeloid leukaemia. *Indian J Ophthalmol.* 1984; 34:239-41.