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Bilateral Proptosis: An Atypical Presentation of Acute Myeloid Leukaemia

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Abstract

Acute myeloid leukaemia accounts for nearly 15% of all leukaemias in children out of which only about 3% cases present with extramedullary deposits in the other sites like the orbit. Bilateral proptosis is fairly common in association with acute and chronic lymphatic leukaemia, on the other hand myelogenous leukaemia rarely give rise to proptosis. Here we present one case of 6 year old presenting as bilateral proptosis with no other manifestations of systemic malignancy at presentation. Radiological investigation, FNAC, bone marrow study and flowcytometry was done for confirmation. The purpose of reporting such a rare entity is to highlight AML as a rare but important differential diagnosis of bilateral proptosis.

Keywords: Acute Myeloid Leukaemia; Orbital Myeloid Sarcoma; Proptosis.

Introduction

Acute myeloid leukaemia (AML) accounts for nearly 15% of all leukaemias in children [1]. The leukemic cells can infiltrate any extramedullary site, tumorous accumulations within soft tissues and bones are known as myeloid sarcomas. Myeloid sarcoma or extramedullary leukemic deposits is an unusual manifestation of AML, accounting for about 3% of cases of AML [2]. Allen Burns was the first to describe it in 1811 as a green tumour involving the orbit [3]. Because these tumours can exhibit a characteristic green colour they were named chloroma. Exposure of the myeloperoxidase enzyme present in these tumour cells to ultraviolet light is responsible for this green colour. Orbital location is the most common [4]. In the case of orbital location, bilateral proptosis is slightly more frequent than unilateral proptosis [5].

We herein described a case with bilateral proptosis as the initial manifestation of AML. A literature review suggests that leukaemia might be the most likely diagnosis in a child with bilateral soft tissue orbital tumours, a point that has not been widely recognized.

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Case Report

A 6 year old male child presented with bilateral proptosis with fever on and off. Eye examination revealed a palpable, ill-defined orbital mass of 3x3 cm in both right and left eye below the eyebrow. Patient was unable to close right eye. The conjunctiva was chemosed and cornea was dry looking. Visual acquity was normal in both the eyes. MRI scan of the orbits show extensive soft tissue infiltration affecting bilateral orbits, eyelid, bilateral masticator spaces, infratemporal fossa, maxillary sinuses, nasopharynx, premaxillary and prezygomatic soft tissue (Figure 1). Both lesions showed enhancement with contrast agents. There is no bone erosion or sinus involvement. The initial peripheral blood cell count revealed a normal white blood cell count of 6.4 x 10^9/L, with a differential count of 14% segmented neutrophils, 42% lymphocytes, 2% eosinophils and 42% blast cells (Figure 2), which was strongly suggestive of acute leukaemia.

Haemoglobin was 6.3 gm/L and platelet was 50

x 10^9/L. Bone marrow study revealed increased cellularity with depressed erythroid and megakaryocytic series. The myeloid series is increased with around 40% Myeloblast (Figure 3). Cytochemistry by MPO was positive in around 10% of the blasts cells (Figure 4).

FNAC of the orbital swelling from both the sides revealed a cellular smear with cells having round to oval nucleus with fine, dispersed chromatin, distinct nuclear membrane, conspicuous nucleoli and moderate amount of cytoplasm (Figure 5).

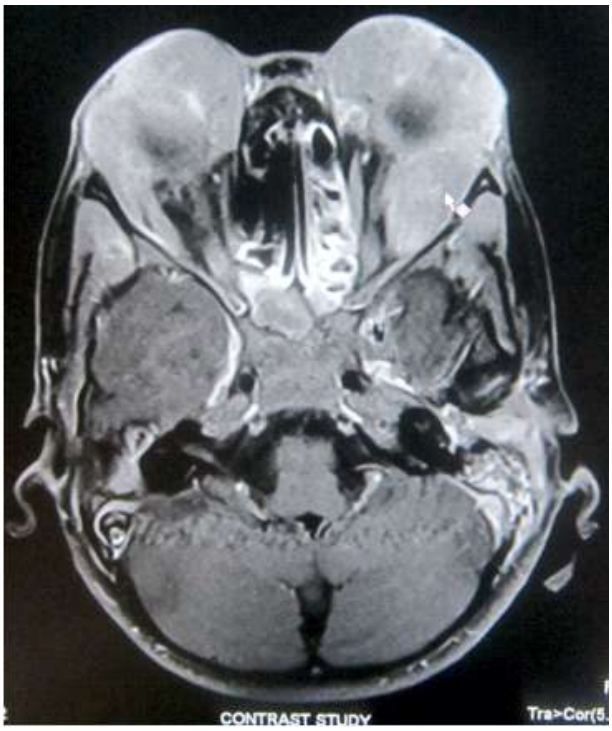

Fig. 1: MRI orbit showing infiltration around orbit

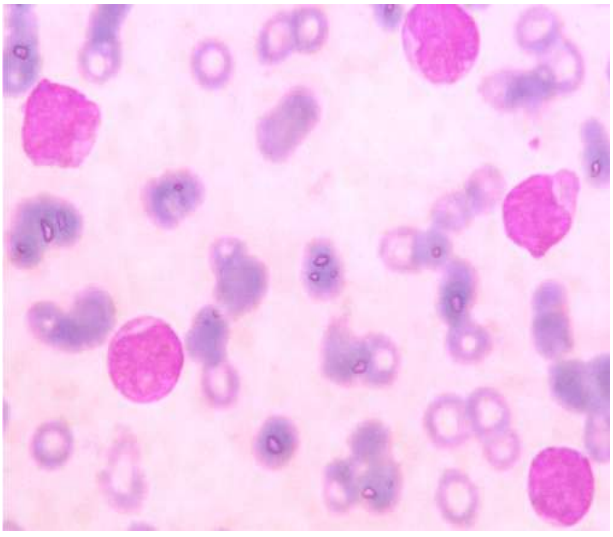

Fig. 2: Showing blasts in PBS

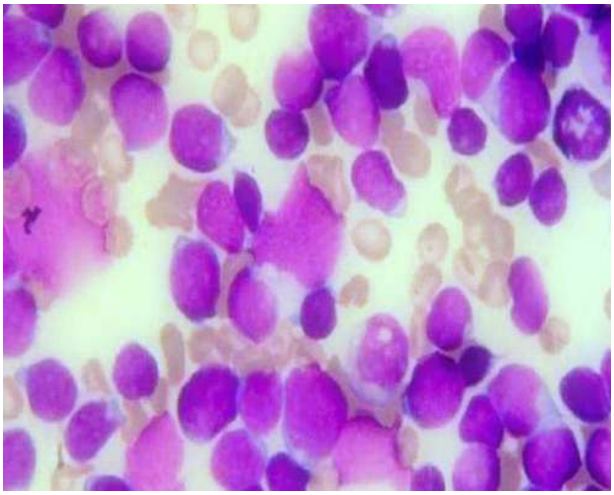

Fig. 3: Bone marrow showing blasts

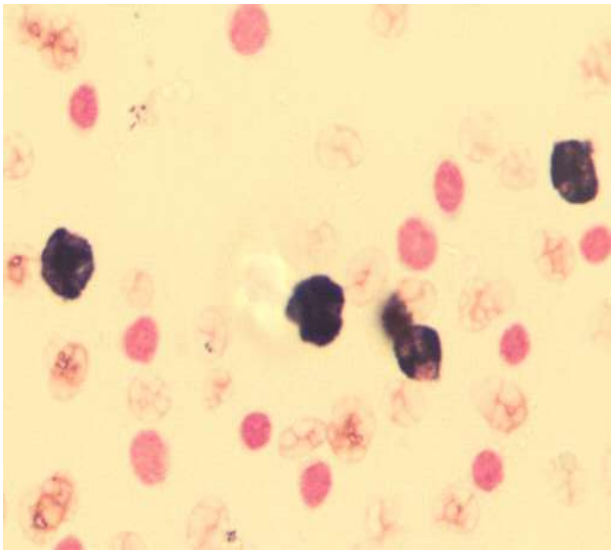

Fig. 4: MPO positivity of blasts

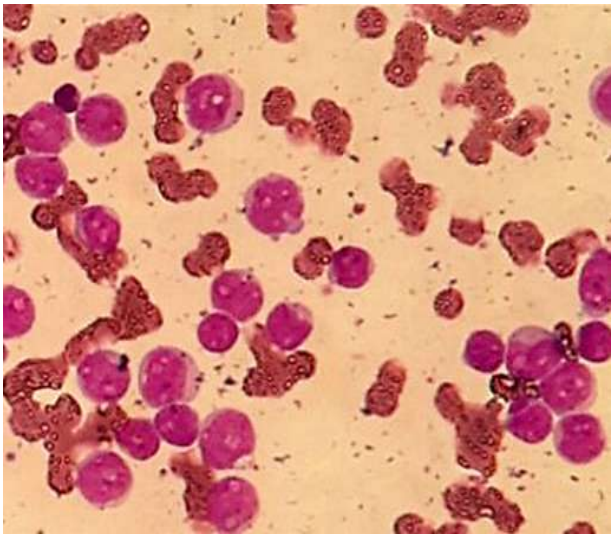

Fig. 5: FNAC from the orbital swelling

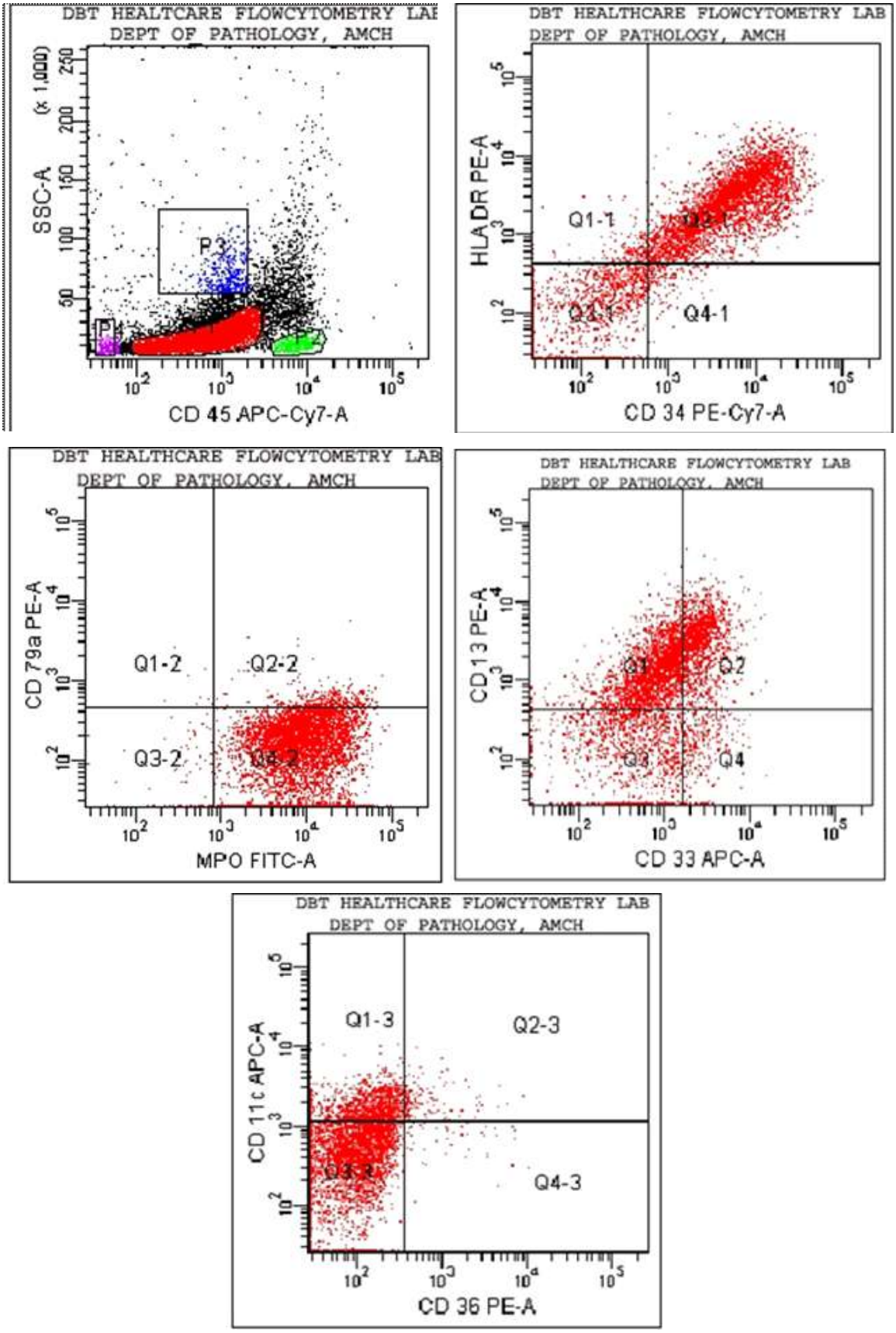

Fig. 6: IPT showing positivity for myeloid markers

Flowcytometric evaluation of the blast cells revealed IPT as MPO, CD13, CD 33, CD34, HLADR, CD11c, CD64 positive cells (Figure 6). Hence diagnosis of acute myeloid leukaemia is made.

Discussion

Acute myeloid leukaemia (AML) accounts for nearly 15% of all leukaemias in children. Rarer sites of its presentation include the orbit, the paranasal sinuses, the gastrointestinal tract, genitourinary tract, breast, cervix, salivary glands, mediastinum, pleura, peritoneum, and bile duct [6].

Acute onset bilateral proptosis in children is frequently encountered in clinical day to day practice. In younger age group the acute onset proptosis is more rapid in its progression. These lesions may be ophthalmic or non-ophthalmic in origin. Some of the commoner conditions causing bilateral acute onset proptosis in children are orbital cellulitis, fungal sinusitis with extension to orbits, metastatic neuroblastoma cavernous sinus thrombosis, inflammatory pseudotumor, thyroid opthalmopathy, rhabdomyosarcoma, bilateral optic glioma and neuroblastoma [7].

Rarely, we encounter a case of acute myeloid leukaemia showing bilateral proptosis. This case is extremely unusual as the presenting complaint was that of bilateral proptosis alone; without any other systemic symptoms of AML. Proptosis in acute myeloid leukaemia is due to diffuse leukemic infiltration of orbital muscles. Recognition of myeloid sarcoma is important, because aggressive induction chemotherapy or radiation therapy can induce complete remission.

Conclusion

Leukemic proptosis is an uncommon manifestation of acute myeloid leukaemia in children. It presents as a diagnostic dilemma when it precedes the prior diagnosis or development of other haematological or systemic manifestation. A peripheral smear examination with bone marrow aspiration in all cases of bilateral proptosis in paediatric age group must therefore be performed. The use of special stains and immuno-phenotyping can help further in establishing the diagnosis. Thus, the purpose of this article is to bring in consideration of acute myeloid leukaemia as a differential diagnosis in a case of childhood bilateral proptosis.

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Conflict of Interest: None.

Ethical Approval: Ethical approval was given by the institutional ethical committee for conducting the present study.

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