

## Acute myeloid leukaemia presenting as bilateral proptosis in a young child

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### BRIEF REPORT

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### ABSTRACT

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#### Background

Myeloid sarcoma is an extramedullary manifestation of acute myeloid leukaemia (AML).

#### Aims

We are reporting a paediatric case presenting with bilateral proptosis, which we were able to diagnose with peripheral blood smear (PBS) examination.

#### Methods

Case Report

#### Results

This case highlights the utility of simple routinely available PBS test in diagnosing this rare disease.

#### Conclusion

Our case highlights the importance of haemogram and peripheral blood smear in the initial evaluation of proptosis. Correct diagnosis of this rare entity is vital especially in cases where (myeloid sarcoma) MS is the presenting feature of AML.

#### Key Words

Myeloid sarcoma, orbit, peripheral blood smear, acute myeloid leukaemia, paediatric

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#### Implications for Practice:

##### 1. What is known about this subject?

The orbit is a known site of extramedullary manifestation of Acute Myeloid Leukaemia.

##### 2. What new information is offered in this report?

Routinely available tests like haemogram and peripheral blood smear can help in early diagnosis of AML presenting as orbital myeloid sarcoma in paediatric patients.

##### 3. What are the implications for research, policy, or practice?

Haemogram and peripheral blood smear should be performed for paediatric patients presenting with proptosis.

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#### Background

Proptosis in children could be due to diverse causes ranging from infections, trauma to life threatening malignancies.<sup>1</sup> An early and timely diagnosis is needed as prompt diagnosis and treatment could be lifesaving. Often the workup of proptosis involves costly and specialized investigations including Contrast Enhanced Computed Tomography (CECT), Magnetic Resonance Imaging (MRI), ocular ultrasonography, and histopathology, to name a few. Haemogram and peripheral blood smear are useful but frequently overlooked investigations in the evaluation of proptosis in children. We present a young child with bilateral proptosis who was referred to us for specialized workup. Diagnosis could be established easily with a thorough physical examination and careful evaluation of the peripheral blood smear.

#### Case details

A 4-year-old female child presented with rapidly progressing bilateral proptosis and fever for 15 days (Figure 1). On examination, best corrected visual acuity (BCVA) was 6/6, n.6 in both eyes with normal ocular motility in all gazes.

There was severe conjunctival congestion. Rest anterior segment evaluation was normal. Optic disc, macula and peripheral retina were also normal. Systemic examination was unremarkable. Differential diagnoses considered were orbital cellulitis, neuroblastomas, lymphoma, rhabdomyosarcoma, and Langerhans cell histiocytosis. Blood investigation revealed haemoglobin 9.2gm/dl (11.5-14.5gm/dl), total leucocyte count 15,000cells/mm<sup>3</sup> (4,000-12,000/mm<sup>3</sup>) and platelet count 252,000cells/mm<sup>3</sup> (150,000-400,000cells/mm<sup>3</sup>). Differential Leucocyte Count (DLC) showed 17 per cent (54-67 per cent) neutrophils, 40 per cent (25-33 per cent) lymphocytes, 4 per cent (1-3 per cent) eosinophils, 1 per cent (3-7 per cent) monocytes, 38 per cent myeloblasts, which was suggestive of AML. On peripheral blood smear (PBS), Myeloblasts demonstrated Auer rods (Figure 2). Kidney and liver function tests were within normal range.

Bone marrow biopsy was done and it demonstrated hypercellular marrow with blast percentage of 25-30 per cent which was suggestive of acute leukaemia probably myelogenous. Contrast enhanced computed tomography (CECT) demonstrated homogeneously enhancing soft tissue mass lesion in extraconal lateral space of both orbits. On both sides, there was extension of mass into pterygopalatine fissure and maxillary sinus (Figures 3 and 4).

Oncologist advised flow cytometer which confirmed the diagnosis of AML. Based on these findings, diagnosis of Myeloid Sarcoma (MS) of orbit as an extra-medullary manifestation of AML was made.

Patient was started on chemotherapy regimen consisting of Daunorubicin with Cytarabine by oncologist. After completing 6 cycles of chemotherapy, the patient continues to be monitored by the oncologist and has been periodically following up with us.

## Discussion

Proptosis is a relatively uncommon clinical entity with patients often attending outpatient services of ophthalmology, paediatrics or neurosurgery department. The differential diagnosis in children is vast including congenital diseases, infections, trauma, and tumours. Often patients are first seen at primary care and subsequently referred to specialized ophthalmology centres for further workup.

Proptosis in AML is due to an extra-medullary proliferation of myeloid blast cells in the extraconal orbital space. These sites of extra-medullary deposits of myeloblast are referred

to as myeloid sarcoma. The common locations include skin, bone, soft tissues and lymph nodes.<sup>2</sup> It was earlier known as Chloroma (because of its green colour due to enzyme myeloperoxidase) or Granulocytic Sarcoma. Apart from AML, MS may occur with myelodysplastic disorders and myeloproliferative disorders. MS may precede, occur concurrently or appear after diagnosis of AML. As in our patient, MS can be the presenting feature appearing before full blown AML and hence early diagnosis of this rare entity is important.<sup>3-9</sup>

In orbit, apart from proptosis, they may present as conjunctival masses, ptosis, and lacrimal gland involvement, iridic and diffuse uveal involvement.<sup>10,11</sup> In paediatric age group, orbit is one of the most common extra-medullary locations, whereas in adults, involvement of central nervous system, pleura, mucosa and skin are relatively more common.<sup>12,13</sup>

In our case, we were able to confirm the diagnosis without an ocular biopsy or fine needle aspiration cytology (FNAC). This case highlights the importance of blood investigations and peripheral blood smear in evaluation of proptosis and other paediatric orbital tumours.<sup>14,15</sup> These investigations can easily be done at most centres. This is especially important in paediatric cases where any specialized investigation (CT, MRI, biopsy or FNAC) needs sedation or general anaesthesia and may not be possible in emergencies.

Early diagnosis of this uncommon condition is essential as timely chemotherapy helps in preservation of vision and improves survival.<sup>16</sup> It is difficult to ascertain the effect of MS on the overall prognosis of AML but recently there is some data to suggest that presence of extra-medullary disease does not alter the prognosis of AML.<sup>17-19</sup>

Children with bilateral proptosis should always undergo systemic evaluation and blood investigations to rule out not only AML but other diseases as well apart from imaging studies because it is difficult to differentiate infective conditions and other malignancies even on CECT and MRI.<sup>20</sup>

## Conclusion

Our case highlights the importance of haemogram and peripheral blood smear in the initial evaluation of proptosis. Correct diagnosis of this rare entity is vital especially in cases where MS is the presenting feature of AML.

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## PEER REVIEW

Not commissioned. Externally peer reviewed.

## CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

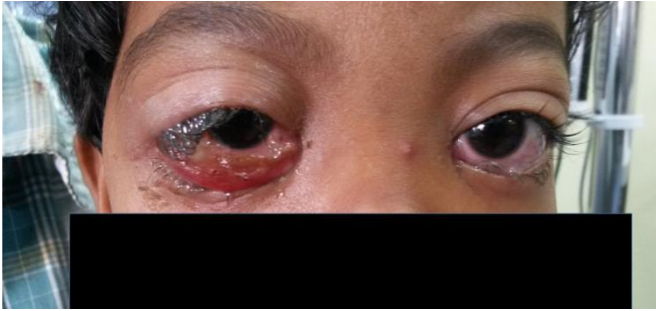
## FUNDING

None

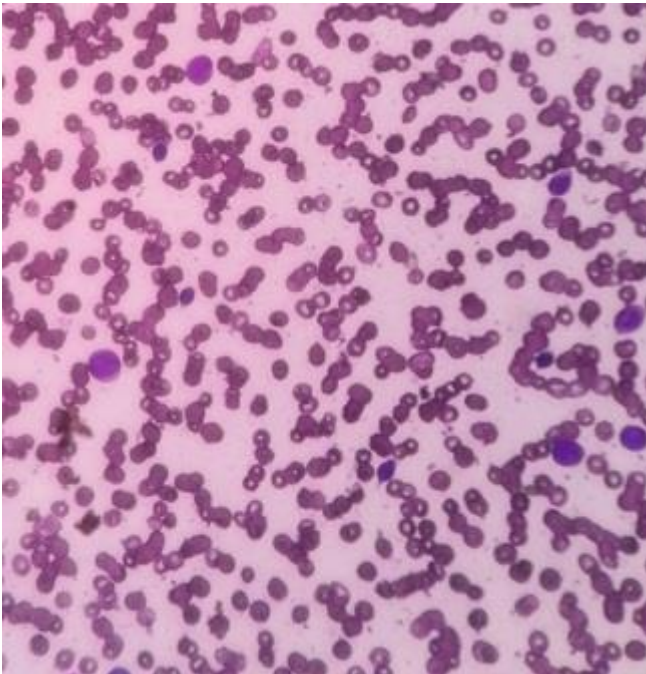
## ETHICS COMMITTEE APPROVAL

Institute ethics committee approval taken. Letter no. SGVEH/EC/ 21/2016.

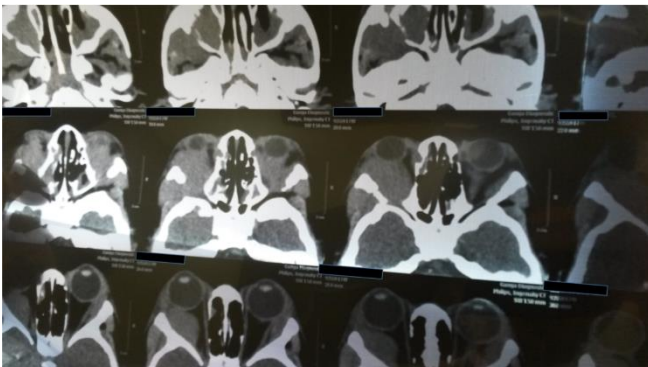
**Figure 1: Clinical photograph of patient with bilateral proptosis**



**Figure 2: Peripheral blood smear demonstrating Myeloblasts with Auer rods**



**Figure 3: Contrast enhanced computed tomography (CECT) demonstrating homogeneously enhancing soft tissue mass lesion in extraconal lateral space of both orbits and extension into pterygo-palatine fissure and maxillary sinus**



**Figure 4: Bone marrow with hypercellular marrow with blast percentage of 25-30%**

