

Granulocytic Sarcoma of the Lids with Endophthalmitis-A Rare Case Report

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Abstract: Orbital and adnexal involvement in leukemia can occur as granulocytic sarcoma which is an infiltration of the soft tissues with myeloid leukemic cells in children. We investigated a 7-year old male child with mild proptosis, large masses in each of his upper lids and the fronto-temporal area and clinical features of endophthalmitis left eye. Histopathology showed blasts cells arranged in scattered singles and diffusely infiltrating fragments of skeletal and fibroconnective tissue with occasional eosinophilic myelocytes. Peripheral smear examination showed numerous type II myeloblasts. A diagnosis of granulocytic sarcoma both lids in a case of acute myelogenous leukemia with endophthalmitis left eye was given.

Keywords: Acute myeloid leukemia, Endophthalmitis, Granulocytic sarcoma, Lid involvement

I. Introduction

Leukemic involvement of the ocular adnexal tissues usually involves the orbit and less often extends to involve the conjunctiva and eyelid. Orbital and adnexal involvement in leukemia can occur as granulocytic sarcoma which is an infiltration of the soft tissues with myeloid leukemic cells.[1] Ocular involvement in leukemia is common although not always clinically evident but orbital and adnexal involvement is quite infrequent. In a large autopsy study it was found that ocular involvement was present in 80% and orbital involvement in only 14% of chronic leukemia and 3-19% in acute leukemia.[2] About 3-8% cases of AML present with granulocytic sarcoma especially AML M2. Other variants are also known to cause extramedullary manifestations but to a lesser extent. [3]

Previously known as "chloroma" or green tumor the terminology presently in use is granulocytic sarcoma as per WHO nomenclature. [4] Granulocytic sarcoma can precede or follow or occur concurrently with myeloid leukaemia which can be acute or chronic. It can even be seen in association with other myeloproliferative disorders and myelodysplastic syndromes. A mean interval of 10.5 months has been observed for the development of leukaemia in non-leukemic cases from the time of diagnosis [4].

In this report of our case, a 7 yr old male child presented with swelling both upper lids, severe visual loss in his left eye, a mild degree of proptosis and clinical features of endophthalmitis left eye without any prior history of leukaemia. He was diagnosed as a case of AML later during the haematological investigations.

II. Case Report

2.1 Case history

A 7 year old male child presented in Eye OPD with swelling around his eyes since the past 2 months. The swelling first started in his right eye which gradually enlarged in size. The same kind of swelling started to develop around his left eye, temple and forehead later on. Since the past two weeks he started to have progressive and rapid loss of vision in his left eye. The associated visual loss was associated with pain, thick discharge and fever. There was no history of trauma to his left eye. He also developed progressive hearing loss more so in his left side. Past ocular and medical history was unremarkable. He did not have a premature birth or family history related to ocular disease. There was no developmental delay. General and systemic examinations were within normal limits (WNL).

2.2 Clinical examination

A firm solid swelling measuring 4cms diameter was present on the glabella and nasal bridge. It was mobile, non-tender and not fixed to the skin. Superficial venous engorgement was also seen. No pigmentary or colour changes were observed. No localized increase in temperature and no bruit found. Similar swelling of 6cms diameter was seen on the right temple. Both eyes showed eye lid involvement but clinical appearance of active inflammatory involvement was seen only in the left eye as evidenced by the full chamber hypopyon and a very tender globe (TABLE1).

2.3 Investigation

On B-Scan numerous hyperechoic shadows seen in the left eye. No evidence of RD.

Haemogram reports showed the presence of numerous type II myeloblasts (64%) with cup like morphology, marked leucocytosis and Auer rods.

FNAC reports of the periorbital swellings showed blasts cells arranged in scattered singles and diffusely infiltrating fragments of skeletal and connective tissue. Majority of cells display cytoplasmic granules. The report showed features of leukaemic infiltration.

Radiodiagnostic study through CECT orbits described the swellings as homogenously enhanced soft tissue lesion without cystic changes and calcification in the upper eyelids, glabella and right fronto-temporal regions. Bilateral superior rectii were compressed however intraconal compartments were normal and so were the optic nerves.

2.4 Diagnoses

Taking into account the clinical features and the investigation reports a diagnosis of AML granulocytic sarcoma both upper eyelids with endophthalmitis left eye was made.

III. Discussion

The accumulation of leukemic cells infiltrating the extramedullary sites are known as granulocytic sarcomas; first described as “Green tumor” by Allen Burns in 1811. [5] AML accounts for about 15% of all leukemias in children and granulocytic sarcoma is a rare manifestation of AML which is seen in approximately 3% of all AML cases. Moreover granulocytic sarcoma presenting as an orbital mass in AML is very rare. [6, 7] In our patient there was bilateral lid infiltration with involvement of the nasion/glabella and the right fronto-temporal region but there was minimal orbital involvement. We believe the endophthalmitis in the left eye was due to endogenous spread since there was no history of ocular trauma.

Ideally we wanted to perform an AC tap and send for microbiological investigations but the patient refused any surgical management and was subsequently referred to a pediatric oncologist at higher centre for further management. We wanted to corroborate our findings with a recent study conducted in Japan where it was found that Invasive fungal infections (IFI) were an important complication in hematologic malignancies and fungal endogenous endophthalmitis was more frequently seen than bacterial endogenous endophthalmitis.[8]

The development of orbital granulocytic sarcoma may be the first clinical feature of the underlying systemic leukaemia as in our case and radiodiagnostic studies such as CT scan can help to distinguish between granulocytic sarcomas from haematomas and abscesses, which are all possible complications of leukaemia.

IV. Figures And Tables

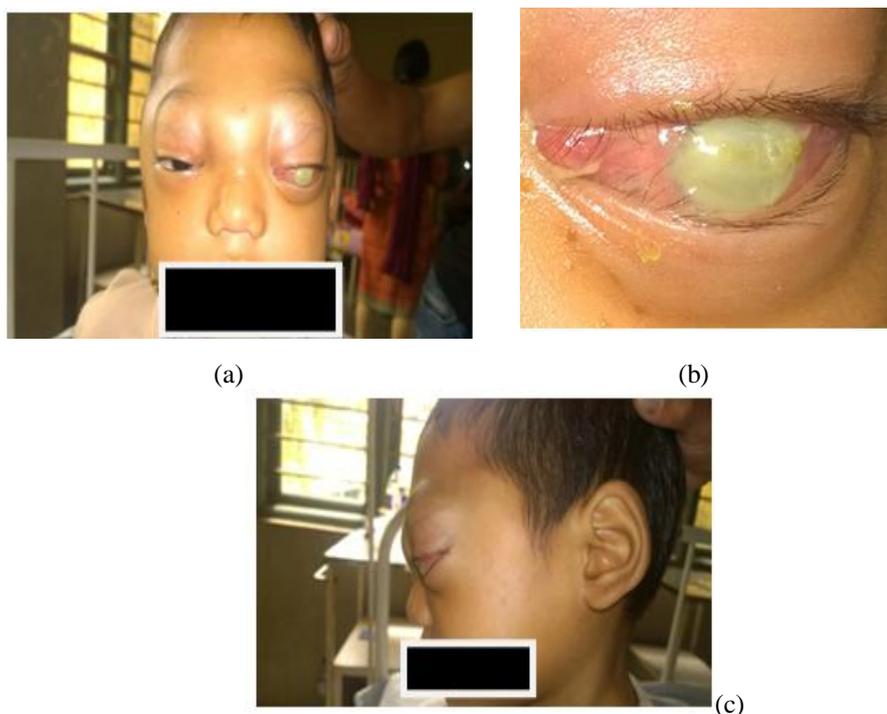


Fig 1(a)upper eyelids with large masses bilateral along with a mass in the glabellar region. (b) left eye with endophthalmitis and (c) proptosis.



Fig.2 CECT orbits showing the bilateral infiltration of the lids by leukemic cells intraconal spaces spared.

	Right eye	Left eye
BCVA	6/60, N6.	No PL.
Exophthalmometry	18mm.	Proptosis of 20mm with a downward displacement of the globe.
Eye movements	Full and free in all gaze directions.	Restricted in all gaze directions
Lids	A firm solid diffuse swelling involving the upper lid and brow, measuring 5 cms at its greatest diameter, fixed to underlying structures and mildly tender. Associated venous engorgement seen but no pigmentary changes observed. A small degree of lagophthalmos and pseudoptosis was also seen.	A firm solid diffuse swelling involving the upper lid and brow, measuring 6.5 cms at its greatest diameter, fixed to underlying structures and mildly tender. Associated venous engorgement seen but no pigmentary changes observed. A small degree of lagophthalmos was also seen.
Conjunctiva-	Normal	Injection both superficial and deep, chemosis and thick mucopurulent discharge.
Cornea	Normal	Generalized haziness and oedematous.
Anterior chamber	Normal	A full chamber hypopyon was present. Rest of the details could not be assessed.
Pupils	Normal size with normal reaction	
Fundus	Clear media Optic disc was normal neither disc edema nor features of atrophy seen. Other fundus findings were normal.	No glow.
IOT	11mmHg.	Soft on digital palpitation

Table1. Clinical findings

V. Conclusion

Ocular involvement in leukemia is common although not always clinically evident but orbital and adnexal involvement is quite infrequent, more so the involvement of the eyelids. Endophthalmitis as evidenced by this case may be a rare presenting feature. Thus in children presenting with features of endophthalmitis without any significant history like ocular trauma, leukemia should be kept in mind as a differential diagnosis.

References

Journal Papers:

- [1] J.A Shields, Eyelid, conjunctival, and orbital tumors: an atlas and textbook (Philadelphia: Lippincott Williams & Wilkins, 2008)
- [2] C.W Taylor, Leukemic infiltration of the orbit: report of three cases and literature review, *Pediatr Hematol Oncol*, 22 (4),2005, 15-22.

- [3] RS Neiman, M Barcos, C Berard, H Bonner, R Mann, et al, Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases, *Cancer*, 48,1988, 1426- 37.
- [4] H Alkatan, I Chaudhry, Myeloid sarcoma of the orbit, *Ann Saudi Med*, 28, 2008, 461-65.
- [5] A. Burns, Observation on the surgical anatomy of the head and neck (Thomas Royce & Co, Edinburg, United Kingdom, 1811).
- [6] F.A Stockl, A.M Dolmetsch, M.A Saornil, R.L Font, and M.N Jr Burnier, Orbital granulocytic sarcoma, *Br J Ophthalmol*, 81(12),1997, 1084-88.
- [7] R. Murthy, G.K Vemuganti, S.G Honavar, M Naik, and V Reddy, Extramedullary leukemia in children presenting with proptosis, *J Hematol*, 2(4),2009.
- [8] R. M Kobayashi, M. M Kaneda, T. M Sato, M. M Ichikawa, D.Suzuki, and T. M Ariga, The Clinical Feature of Invasive Fungal Infection in Pediatric Patients With Hematologic and Malignant Diseases: A 10-year Analysis at a Single Institution at Japan. *Journal of Pediatric Hematology/Oncology*, 30(12), 2008, 886-90.