

# Bilateral Extra Ocular Muscle Involvement with Proptosis as the Ophthalmic Manifestation and Primary Presentation of Acute Leukemia in an Adult

Sivakami A Pai\*, Afra M Iootah and Moza A Dekhain

Department of Ophthalmology, Dubai Health Authority, UAE

## Purpose

Leukaemic infiltration of extra ocular muscle without other evidence of orbital disease is extremely rare and late manifestation of adult onset acute Leukaemia. We here in report the clinical and imaging feature of acute lymphoblastic Leukaemia with bilateral orbital infiltration, presenting with primary ocular manifestation in an adult male.

## Materials and Design

An observational case report of a 32 year Asian seen in Rashid hospital emergency. He presented with bilateral proptosis and bloody eye discharge of two weeks duration. Fever, headache and general weakness of one week duration. There was no prior history of leukaemia or any systemic illness. Clinical examination revealed asymmetrical axial proptosis with restricted extra ocular movements. Diffuse subconjunctival haemorrhage with bloody eye discharge, Figure 1. Anterior segment and fundus examination was unremarkable. A complete blood counts, flow cytometry studies and computerized tomography (CT) were done.

## Results

Complete blood counts and differential counts are shown in Table 1. Flow cytometry showed, 77% of the cells were blasts. The blasts were positive for cytoplasmic CD3 (85%), CD7 (95%), CD 10 (82%), CD2 (35%) and CD38. The blasts were double negative for CD8 and CD4 with a sub clone (almost 12 % of the cells ) showing expression of CD8. The blasts also showed aberrant expression of CD13 (98%). The blasts were negative for surface CD3, CD5, CD34 and TDT. They were also negative for myeloid markers CD117, cytoplasmic MPO and CD33. The blasts were negative for HLA DR, CD20, CD22, CD19, CD79a, and for both CD16 and CD56. These results were consistent with the

Result date time	Description	Value	Unit	Range
07/07/10 16:31	WBC Count L	0.6	10 <sup>3</sup> /ul	3.6 - 11.0
07/07/10 16:31	RBC Count L	2.66	10 <sup>6</sup> /ul	4.50 - 6.00
07/07/10 16:31	HGB L	8.5	g/dL	13.0 - 18.0
07/07/10 16:31	HCT L	23.9	%	40.0 - 52.0
07/07/10 16:31	MCV	90.0	fL	77.0 - 92.0
07/07/10 16:31	MCH	32.0	pg	26.0 - 34.0
07/07/10 16:31	MCHC	35.6	g/dL	32.0 - 36.0
07/07/10 16:31	RDW H	16.2	%	11.5 - 14.0
07/07/10 16:31	Platelet Count L	30	10 <sup>3</sup> /uL	150 - 400
07/07/10 16:31	MPV	7.5	fL	7.4 - 10.4
07/07/10 16:31	DIFF TYPE MANUAL			
07/07/10 16:31	ABS NEUTRO	0.3	10 <sup>3</sup> /uL	1.4 - 8.3
07/07/10 16:31	ABS LYMPH	0.3	10 <sup>3</sup> uL	0.7 - 5.0
07/07/10 16:31	ABS MONO	0.0	10 <sup>3</sup> uL	0.1 - 1.1
07/07/10 16:31	NEUTRO	51	%	54 - 62
07/07/10 16:31	LYMPHNO	46	%	20 - 40
07/07/10 16:31	MONO	3	%	4.10
07/07/10 16:31	WBC MORPHO			

Table 1: Complete blood counts and Differential counts.

diagnosis of Precursor T Cell lymphoblastic Leukemia. CT scan axial view, contrast enhanced showed enlargement of all the extra ocular muscles, due to leukaemic infiltration of all extraocular muscles, Figure 2A and Figure 2B. Despite multiple blood product transfusions and chemotherapy the patients condition rapidly deteriorated and expired after ten days.

## Discussion

Rapid onset of bilateral proptosis without any other systemic findings is unusual presentation of adult onset leukaemia. Leukaemic infiltration of extra ocular muscle without other evidence of orbital disease is extremely rare and late manifestation of the advanced disease



Figure1: Picture of a 32 year old Asian showing bilateral asymmetrical axial proptosis with bloody eye discharge.

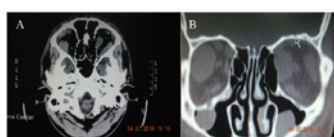


Figure 2: A. CT scan axial view contrast enhanced. Shows enlargement of all the extraocular muscles, due to leukaemic infiltration. B. CT brain and Orbits, sagittal section also showing diffuse enlargement of extra ocular muscles.

\*Corresponding author: Dr. Sivakami A Pai, Vitreo- retinal Surgeon, DHA, UAE, E-mail: [sushirshiv@yahoo.com](mailto:sushirshiv@yahoo.com)

Received February 16, 2011; Accepted May 30, 2011; Published June 01, 2011

Citation: Pai SA, Iootah AM, Dekhain MA (2011) Bilateral Extra Ocular Muscle Involvement with Proptosis as the Ophthalmic Manifestation and Primary Presentation of Acute Leukemia in an Adult. J Clin Exp Ophthalmol 2:169. doi:10.4172/2155-9570.1000169

Copyright: © 2011 Pai SA, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

[1,2]. Medline search revealed only few reports of extra orbital muscle involvement in leukaemia. A rapid onset (within 5 days) of bilateral exophthalmos in a low-grade and early stage chronic lymphocytic leukemia (CLL) was reported by Ramkissoon et al. [2]. Orbital involvement in CLL is also highly unusual and most commonly involves hemorrhage or soft tissue infiltration in advanced disease [3,4], and a primary leukaemic infiltrate of the orbit in CLL is especially rare [5-7].

Chen et al have described a 16 month old child with acute myeloid leukemia presenting as bilateral proptosis secondary to leukaemic infiltration of all extraocular muscles in both eyes, without any other orbital involvement [8]. Orbital involvement is uncommon even in childhood leukaemia, however; primary orbital presentation without any evidence of systemic disease is only rarely seen in acute childhood leukemia and is typically due to chloroma [9]. Although rare, in a child with the sudden onset of proptosis without any other systemic findings, the diagnosis of acute leukemia must be considered [8].

Ideally in this case a muscle biopsy and bone marrow biopsy should have been done to confirm leukaemic infiltration of the extraocular muscles. Confirmation of diagnosis by blood investigations and rapidly deteriorating general condition of the patient did not warrant further investigations. Despite intensive chemotherapy and supportive treatment the prognosis in such cases in adults is dismal and one of the main reasons being the late and rare presentation of the disease. Although an unusual presentation even in adults, bilateral acute proptosis without systemic illness, the possibility of leukaemia must be kept in mind.

## References

1. Kiratli H, Balci KE, Himmetoğlu C, Uner A (2010) Isolated extraocular muscle involvement as the ophthalmic manifestation of leukaemia. *Clin Experiment Ophthalmol* 38: 651.
2. Ramkissoon YD, Lee RW, Malik R, Hsuan JD, Potts MJ, et al. (2008) Bilateral infiltrative disease of the extraocular muscles: a rare clinical presentation of early stage chronic lymphocytic leukemia. *Orbit* 27: 293-295.
3. Rootman J, White VA, Connors JM (2003) Lymphoproliferative, leukemic, and histiocytic lesions of the orbit. Rootman J, ed. *Diseases of the orbit: A multidisciplinary approach*. Philadelphia: Lippincott Williams and Wilkins-Raven. 385-416.
4. Rosenthal AR (1983) Ocular manifestations of leukemia. A review. *Ophthalmology* 90: 899-905.
5. Kincaid MC, Green WR (1983) Ocular and orbital involvement in leukemia. *Surv Ophthalmol* 27: 211-232.
6. McKelvie PA, McNab A, Francis IC, Fox R, O'Day J (2001) Ocular adnexal lymphoproliferative disease: A series of 73 cases. *Clin Exp Ophthalmol* 29: 387-393.
7. Hatton MP, Rubin PA (2002) Chronic lymphocytic leukemia of the orbit. *Arch Ophthalmol* 120: 990-991.
8. Chen E, Morrison DG, Donahue SP (2004) Acute myeloid leukemia presenting as bilateral proptosis from diffuse extraocular muscle infiltration. *Am J Ophthalmol* 137: 948-950.
9. Duke-Elder S (1974) Orbital involvement in leukemia. In: Duke-Elder S, editor. *System of ophthalmology*. Volume 13. Part 2. London: Henry Kimpton 13: 1073-1076.