

Rare primary presentation of chronic lymphocytic leukemia as chronic orbital space occupying lesion

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Dear Sir,

I am Dr. Vaibhav Kumar Jain from the Department of Ophthalmology, Post Graduate Institute of Medical Education and Research, Chandigarh, India. I write to present a case report about the rare primary presentation of chronic lymphocytic leukemia (CLL) as chronic orbital space occupying lesion.

The ophthalmic acute presentation of CLL is extremely rare and its chronic presentation has not been reported till now to our best knowledge [1-2]. CLL can involve optic nerve, orbit, conjunctiva, choroid, retina and vitreous [1-4]. CLL presenting as orbital space occupying lesion is exceedingly rare. Here, we report a case of CLL presenting as orbital space occupying lesion of chronic duration in an elderly patient.

A 72-year-old male presented with 4mo history of left eye proptosis which was insidious in onset and gradually progressive, associated with ptosis of upper eyelid for 1mo. It was painless in nature and was not associated with diminution of vision or watering. There was no history of trauma, constitutional symptoms and systemic illness. He gave history of cataract surgery with lens implantation done in both eyes 6mo ago.

Ocular examination revealed best corrected visual acuity 6/6 both eyes with normal color vision. Right eye revealed compensatory upper lid retraction and rest of the examination

being normal. Left eye had 5 mm of proptosis and mechanical ptosis. Extra-ocular movements were restricted on superior, medial, and inferior gaze. There was fullness of superior sulcus and mild conjunctival congestion over temporal bulbar conjunctiva (Figure 1A). Palpation revealed a firm, multilobulated, ill defined mass superior to globe, free from superior orbital margin, and overlying skin. Its posterior extent could not be palpated. Retrobulbar resistance was raised. Intra-ocular pressure was 18 mm Hg. Anterior segment and posterior segment were within normal limits.

Magnetic resonance imaging of the orbit revealed lobulated heterogeneous lesion, iso-to hypo-intense on T1W/T2W in extraconal compartment of left orbit, encasing superior rectus muscle and extending into intraconal compartment (Figure 1B). The lesion was abutting medial wall of orbit, medial rectus, optic nerve and globe. It was also molding along the superior wall of orbit and extending into the preseptal region.

Patient's general physical examination was within normal limits. His total leukocyte counts were significantly raised at 36.1×10^3 per microliter with absolute lymphocyte count of 32.5×10^3 per microliter. Hypochromic microcytic anaemia (Haemoglobin 10 g/dL, mean corpuscular volume 62 fl, mean corpuscular haemoglobin concentration 19.8 pg, red cell distribution width 14.1) with high erythrocyte sedimentation rate (32 mm) was also noted. Peripheral blood film showed small, mature lymphocytes and normocytic red blood cells. Orbital mass biopsy revealed nodular aggregates of mature lymphocytes with high nucleoli to cytoplasm ratio, cleaved nuclear membrane, hyperchromatic nuclei, infrequent mitotic figures and scanty cytoplasm (Figure 1C). Bone marrow showed hypercellular marrow spaces with interstitial excess as well as nodular aggregates of lymphoid cells. Immuno-phenotyping verified CD19, CD20, CD23, CD22, CD5 positivity consistent with diagnosis of CLL.

Based on clinical, imaging, peripheral blood film, bone marrow and immuno-phenotyping studies, diagnosis of CLL with extranodal orbital mass, Rai stage III, Binet B was made. Patient was started on systemic chemotherapy with injection bendamustine 90 mg/m² for 2d and rituximab 375 mg/m² for 1d. Total 6 courses of chemotherapy were completed. At the end of therapy, positron emission tomography scan was repeated which showed complete resolution of extranodal deposition in the orbit. Other

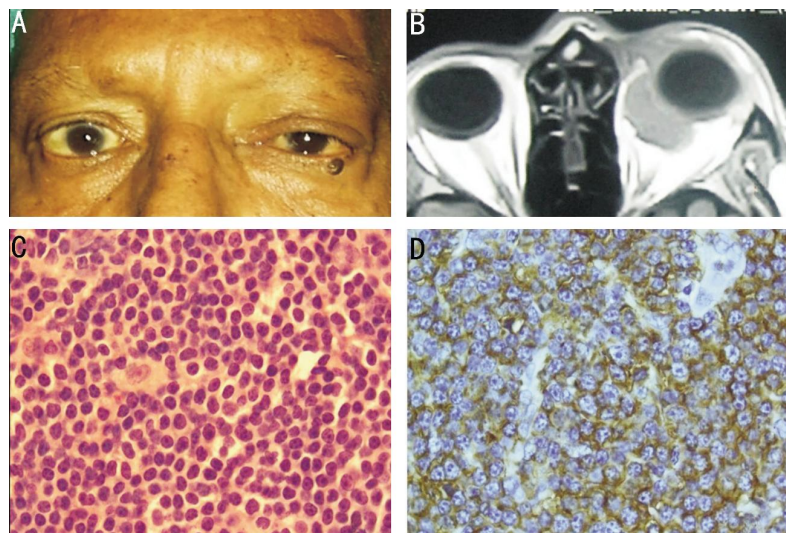


Figure 1 Clinico–pathological photographs of patient with chronic lymphocytic leukaemia A: Clinical photograph at presentation showing left proptosis, pseudoptosis, fullness of superior sulcus and mild temporal conjunctival congestion; B: Magnetic resonance imaging T1W axial scan showing retrobulbar hypointense lesion in the intraconal space and extending anteriorly into preseptal space; C: Hematoxylin and eosin-stained section from orbital mass biopsy shows diffuse sheets of small lymphocytes (×400); D: Small lymphocytes with diffuse membranous immunopositivity for CD20 (Pan B cell marker) (×400).

hematological parameters reverted to normal and currently patient is in complete remission state.

Lymphoma is the most common malignant orbital tumor representing 12% of cases in adults [5] and 24% of cases in elderly population [6]. Overall incidence of systemic disease associated with orbital lymphoid proliferation is 30%-35%[7]. Though lymphoma is most common malignant tumor in the orbit, involvement in leukemia is very rare [8]. Except for myeloid leukemia which presents as an orbital soft tissue mass (granulocytic sarcoma or chloroma), leukemia mostly has ocular involvement with fundus and optic nerve predominantly. Leukemia presenting as a chronic orbital mass is very rare[4].

Though orbit and adnexal structures are known to involve by leukemia cells but the association is more common with acute leukemia [5,7]. Orbit involvement in CLL is exceedingly rare, with only few cases reported [5]. CLL presenting as chronic orbital space occupying lesion is not reported till now to our best knowledge which we described. Ramkissoon *et al*[9] reported bilateral infiltrative disease of extraocular muscle in early stage of CLL in a 72-year-old man at presentation. Burton *et al*[10] reported eye involvement mimicking scleritis in the form of mass in inferior orbit attached to sclera in a 87-year-old man with CLL. The orbital mass developed in two patients of CLL on chemotherapy reported by Kiratli *et al* [4] in the form of orbital Richter's syndrome. Another case presented as profound bilateral ptosis due to tumor mass primarily in the extraocular muscles[11].

Our patient had this unusual presentation of CLL in the form of chronic orbital space occupying lesion and proptosis. Patient responded remarkably to combination of chemotherapy and anti CD20 targeted therapy.

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