Unilateral Conjunctival Chemosis as a Presenting Sign of Plasmablastic Lymphoma (PBL) - a case report

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Background

Plasmablastic lymphoma (PBL) was first described in 1997 by Delecluse et al.,[1] with a total of sixteen cases, exclusively involving the oral cavity and predominantly in human immunodeficiency virus (HIV) positive patients. Since then, approximately 120 cases of PBL in oral and extra-oral sites in HIV-negative patients have been reported in the literature. PBL is distinguished as a subtype of diffuse large B-cell lymphoma (DLBCL) due to the presence of neoplastic cells resembling B immunoblasts but with immunophenotypic features of plasma cells. Although there have been reported cases of extra-oral PBL, orbital involvement in an immunocompetent patient is extremely rare. To date, there are only eight published cases of orbital PBL and of those, only two have been reported in HIV-negative patients.[2-6] We report a third case of an HIV-negative patient with ocular adnexal involvement as the initial presenting sign of PBL.

Case Report

A 79-year-old Caucasian male presented to our clinic as a new patient for an evaluation of a swollen, mildly painful right eye x two weeks. He also complains of intermittent tenderness in his right frontal and paranasal sinuses. He states having been evaluated by an ENT specialist one week prior to his visit with us and reports the results of that exam to be unremarkable.

BCVA: 20/40 in the right eye and 20/30 in the left eye

PERRL (-)APD; EOM: full and smooth in all directions of gaze.

External evaluation: Mild proptosis of the right eye; Digital palpation of the right paranasal, maxillary sinuses and periorbital areas revealed a diffuse mass.

SLE: moderate conjunctival chemosis, greatest in the temporal quadrant; diffuse inferior eyelid edema, and moderate keratitis in the right eye. Mild nuclear sclerotic cataracts in each eye. Posterior segment: unremarkable in each eye.

MRI of the brain and orbits without contrast revealed an expansile lesion of the right maxillary, ethmoid, and paranasal sinuses (Figures 1a and 1b).

CT scan of the paranasal sinuses revealed a large lobulated mass of the right paranasal sinus, with multiple areas of bony erosion with extension into the anterior cranial fossa and into the right orbit (Figure 2). Needle biopsy and pathology analysis revealed a very rare form of plasmablastic lymphoma (Figure 3).

Figures

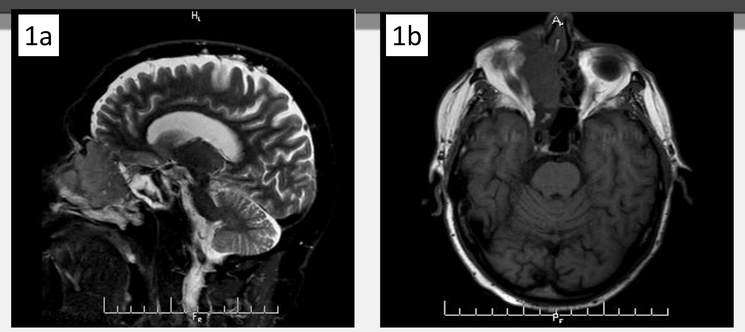

Figure 1a and 1b: MRI of Brain/Orbits without contrast shows a lobulated mass in the right maxillary and ethmoidal sinuses with invasion into the right orbit with compression of the inferior medial rectus muscle (sagittal and axial views)

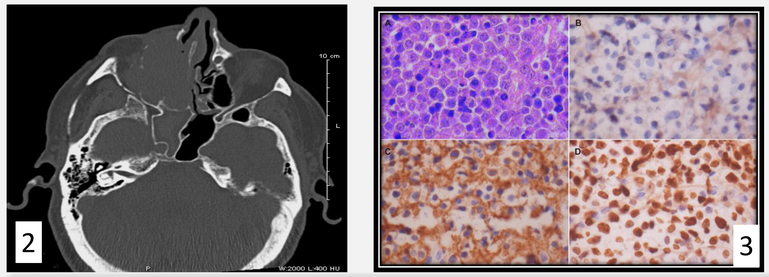

Figure 2: CT scan of sinuses shows mass extending into the right maxillary and ethmoidal sinuses with multiple areas of bony erosion. The mass also extends through the cribriform plate into the anterior cranial fossa and the right orbit causing right globe proptosis (axial view)
Figure 3[5]: Microphotograph and immunohistochemical staining. A. Atypical plasmacytoid tumor cells with abundant cytoplasm and a large eccentric vesicular nucleus with a prominent nucleolus. B. Immunohistochemical staining showing lack of immunoreactivity for CD 20. C. Strong membranous CD138 immunoreactivity. D. Strong Ki-67 immunoreactivity in almost all tumor cells

Table 1: Clinicopathologic Characteristics of all Orbital PBL cases reported in current literature versus our case

	Degnan et al (n=1)	Morley et al (n=2)	Barthuysen et al (n=1)	Mulay et al (n=3)	Valenzuela et al (n=1)	Our Case (n=1)
Age	43	40, 49	50	45,45,48	41	79
Gender (Male vs Female)	M	M,M	F	F,M,M	F	M
Race	Caucasian (C)	West African, C	C	All Indian	C	C
Presentation	L pain/proptosis; L-sided HA; Jaw abscess	Nasal congestion; swelling over right cheek	Acute proptosis; vision loss; ophthalmoplegia	1: severe proptosis and decreased vision 2: conjunctival chemosis with severe proptosis 3: proptosis with lid edema and complete ptosis with restricted globe motility	Painless RUL induration	moderate conjunctival chemosis; mild globe proptosis
OD/OS	OS	OD, OS	OS	OD, OS, OS	OD	OD
HIV status	+	+, -	+	-, +, +	+	-
BCVA	Not indicated	NLP, Normal	Decreased	CF, LP, CF	Not indicated	20/40
Proptosis	yes	yes	yes	yes	yes	Yes
Treatment	EPOCH/Pegfilgrastim with recurrence at 10 months	CHOP/HAART CHOP/DHAP	HAART, RCHOP, radiotherapy to skull	1: No Treatment 2: HAART/CHOP 3: HAART/CHOP	HAART/CHOP Relapse at 6 months; hepatotoxicity to chemotherapy led to cessation	CHOP – patient declined after initial treatment; radiation treatment thereafter
Dead/Alive	Alive and on treatment	Both died within 3-mo of initial presentation	Alive 13 months after diagnosis	1: Died 1 week after diagnosis before treatment initiated 2: Died 6 months following initial presentation 3: alive 3 months post diagnosis with improvement	Died of massive GI hemorrhage due to invasive duodenal lymphoma	Alive 11 months after diagnosis

Discussion

PBL involving the ocular adnexa in an immunocompetent person is extremely rare. Liu et al.,[7] performed a comprehensive Pubmed literature search which identified a total of 114 cases of HIV-negative PBL described in either case reports or small sample size case analyses between February 1997-2014. We performed a thorough literature search of all reported cases of orbital PBL via Pubmed and found only eight cases, and only two of those in HIV-negative individuals. Our patient will be the third reported case of orbital PBL in an HIV-negative individual.

Key Discussion Points about PBL:

- It is a rare subtype of diffuse large B-cell lymphoma – ocular involvement is extremely rare
- It has a male predominance with mean age of 39 years at presentation in HIV+ patients and 54 years in HIV- patients
- It accounts for only 2.6% of all AIDS-related lymphomas but is highly aggressive
- Its pathogenesis is still not very clear – most PBL patients (including our patient) are positive for the Epstein Barr virus, perhaps confirming its role in pathogenesis of PBL
- The more common presenting signs of orbital PBL are conjunctival chemosis, eyelid swelling, ptosis, decreased vision, and proptosis
- No clear guidelines are available for treating PBL – most are treated with CHOP and CHOP-like regimens
- No long term follow up data are available due to relapse and early death, despite treatment.

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