Case report

Retinal necrosis as the initial presentation of primary intraocular lymphoma

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Abstract

A 72 years old female presented with bilateral painless progressive loss of vision over one year. She was diagnosed as non-resolving bilateral panuveitis. Her visual acuity in right eye was hand movement close to face and left eye was perception of light with inaccurate projection of rays. Bilateral anterior chamber had 1+ cells and flares. Vitreous cells had 1+ cells and haze in right eye but the left eye had 3+ vitreous cells and haze. Right eye fundus had multiple, discrete sub retinal yellowish deposits with subretinal haemorrhage and macular edema with perivascular infiltrates. In left eye, disc was just visible. The patient underwent diagnostic vitrectomy in left eye and undiluted vitreous sample on cytology showed reactive large lymphoid cells with necrotic background pattern suggestive of intraocular lymphoma. Patient underwent external beam radiotherapy and chemotherapy.

Keywords: Lymphoma, Retinitis, Uveitis, Vitrectomy

Introduction

Intraocular lymphoma though uncommon, is highly malignant and lethal disease. It is an important masquerade syndrome usually affecting the adults and can be easily mistaken for uveitis. Intraocular lymphoma can be broadly divided into primary intraocular lymphoma (PIOL) arising primarily in the eye and Central Nervous System (CNS) or secondary lymphoma disseminated from the systemic diseases. It is estimated to represent 4-6% of all the intracranial tumors and 1-2% of all extranodal non-Hodgkin's lymphoma (Freeman et al., 1987). Primary intraocular lymphoma can present with a wide variety of manifestations. Typically, it presents with vitreous opacification due to dense infiltration of sheets of lymphoma cells. It can also present with subretinal infiltration but infrequently it can mimic as diffuse uveitis that is refractory to corticosteroids (Gill and Jampol, 2001). Herein, we report a case bilateral primary intraocular

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lymphoma masquerading as necrotizing viral retinitis which was mistaken and treated as panuveitis.

Case report

A 72-year-old- female presented with diminution of vision in right eye for 3 months and decreased vision with floaters in both eyes for one year. The patient was diagnosed as bilateral panuveitis elsewhere and was under oral corticosteroid therapy since 3 months but with no improvement. On ocular examination, her visual acuity in right eye was hand movement close to face (HMCF) and left eye had perception of light with inaccurate projection of rays. Anterior segment examination of both eyes revealed presence of 1+ cells and flares and pigments on anterior lens surface. The right eye had grade II nuclear sclerosis and left eye had grade III nuclear sclerosis grade. The vitreous cells and haze was 1+ in right eye but the left eye had 3+ vitreous cells and haze. Right eye funduscopy showed multiple, discrete as well as coalesced sub retinal yellowish deposits with subretinal haemorrhage and macular edema (Figure 1). There were areas of perivascular infiltrates (Figure 2) as well as peripheral sclerosed vessels. In left eye, disc was just visible and other details could not be ascertained due to media haze and lenticular opacity (Figure 3). Spectral domain optical coherence tomography (SDOCT) of both the eyes showed presence of lumpy-bumpy RPE-choroidal alterations suggestive of infiltration of Retinal Pigment Epithelium (RPE) by subretinal deposits (Figure 4). The patient was subjected to phacoemulsification with diagnostic vitrectomy in left eye and undiluted vitreous sample was sent for cytology study. Vitreous cytology revealed reactive large lymphoid cells with high nucleocytoplasmic ratio with numerous necrotic cells with necrotic background pattern suggestive of intraocular lymphoma (Figure 5) suggestive of intraocular lymphoma. Following surgery, the detailed fundus view was possible and revealed pale disc with multiple areas of atrophic scars involving macula so the vision remain static to perception of light with inaccurate projection of rays. Patient was referred to an oncologist where she underwent external beam radiotherapy for the both eyes and chemotherapy.

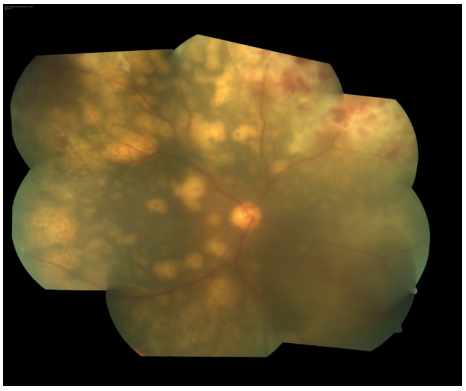

Figure 1: Fundus montage of right eye showing multiple areas of subretinal infiltrates with peripheral haemorrhage and macular edema

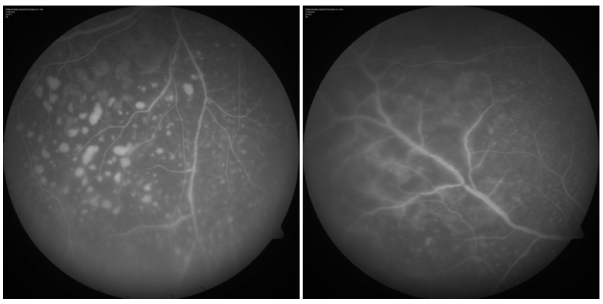

Figure 2: The red free fundus photograph of right eye showing multiple subretinal deposits (left) and inflamed vessel with perivascular cuffing (right)

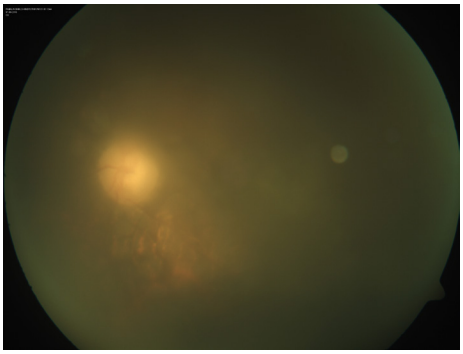

Figure 3: Fundus photograph of left eye with dense vitritis and just visible disc

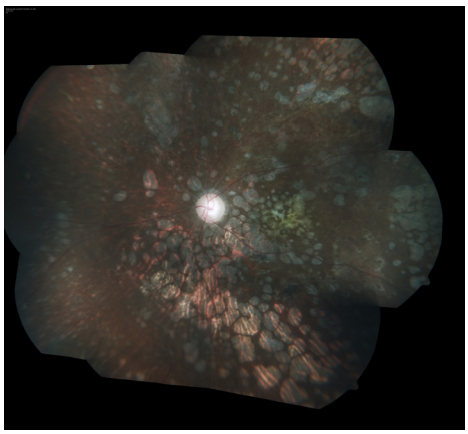

Figure 4: Post operative fundus photo of left eye showing pale disc with extensive retinal scarring and atrophic areas

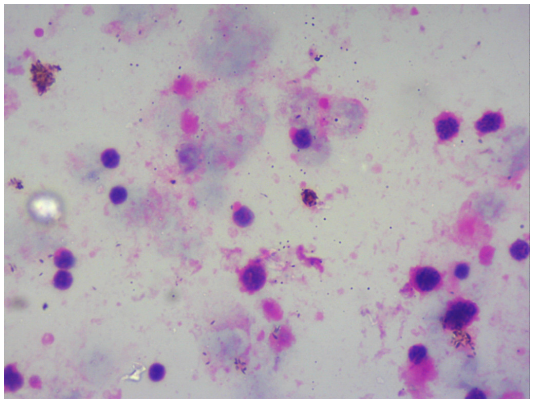

Figure 5: Vitreous cytology showing large lymphoid cells with necrotic background (H & E x 400).

Discussion

PIOL usually affects the elderly females above 60 years. Eighty to ninety percentage of them can develop bilateral involvement over few years of life (Kimura et al., 2012). Due to the rarity of PIOL and varied clinical presentations, there are chances of misdiagnosis and delays in management and prognosis (Goto, 2016) which has its inverse effect in visual outcome like in our case. The disease is not only sight threatening but also life threatening with 5-year survival rate of 30-60% (Kimura et al., 2012), (Chan, 2007). Though, it can masquerade in any form but initial presentation in form of necrotizing viral retinitis is considered rare thus chances of misdiagnosis is high and this holds true for our case where delayed diagnosis lead to permanent loss of vision. Literature review shows evidence of only one isolated case report of PIOL presenting as retinal necrosis (Whitcup et al., 1993).

Ophthalmologist plays an important role in diagnosis of intraocular lymphoma where it can be the first presenting feature without any systemic signs. A high degree of suspicion is very essential in the diagnosis and should always be considered in the differential diagnosis of non resolving inflammation of long duration especially in patients in older age group refractory to steroid therapy. Although characteristic vitreous sheets and leopard spots in the fundus gives a clue to diagnosis but this may not be the usual scenario. Vitreous biopsy is important for the diagnosis and clinched the diagnosis in our case. Char and colleagues reported the need of multiple vitreous samples to correctly establish the diagnosis due to the fragileness of the lymphoma cells in the sample. (Char et al., 1988).Vitrectomy has both therapeutic and diagnostic significance. The clearance of dense vitreous helped us to better evaluate retinal lesions but the late presentation and long standing disease could not revert the functional outcome. The undiluted vitreous sample also helps in diagnosis of cytological study of infiltrative cells, measurement of cytokine levels (interleukins 10 & 6), detection of clonal rearrangement of heavy chain gene and for detection of chromosomal abnormalities (Goto, 2016). Intraocular lymphoma often has a fatal outcome, but recognition of its modes of presentation facilitates early diagnosis and treatment that may improve prognosis.

Conclusion

High index of suspicion, early clinical diagnosis and demonstration of lymphoma cells in the vitreous are to be borne in mind to identify intraocular lymphoma.

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