Case Report

Choroidal Detachment in Idiopathic CD4 Lymphocytopenia

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Abstract

Usually reduction in CD4 cell count is seen with HIV infection, however CD4 lymphocytopenia in absence of HIV infection is called as Idiopathic CD4 Lymphocytopenia (ICL). The patients usually present with features of immunosuppression and opportunistic infections. Ophthalmic manifestations of this rare entity are not well known and very few cases are reported till date. We present a case of a 69 year old male patient who reported with profound reduction in his visual acuity in his left eye for 15 days. On examination of his left eye the visual acuity was hand motion close to face and he had 360 degree choroidal detachment which was confirmed on a B scan. He had evidence of subretinal fluid on optical coherence tomography (OCT). His detailed uveitis workup was unremarkable and HIV test was negative twice repeated at two occasions six weeks apart. His CD4 count was 57 cells/ mm3 with all other investigations normal. This report highlights that iIdiopathic CD4 lymphocytopenia is a rare entity which usually presents as opportunistic infections as reported in the literature. This case of ICL had choroidal detachment which is unusual presentation of this rare entity.

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Introduction

CD4 cells have important role in the immune mechanism of the body and reduction in CD4 cell count may lead to opportunistic infections and reduced immunity. Low CD4 counts are commonly reported in the setting of HIV infection, however few patients without HIV who present with opportunistic infections may have low CD4 cell counts on extensive investigations. This entity was later reported as idiopathic CD4 lymphocytopenia (ICL), which is a rare condition with low CD4 counts in absence of HIV infection. The CD4 cell counts are usually less than 300 cell/mm3 or 20% of total T cells on two separate occasions1. Most of the cases of ICL have been described with systemic and ophthalmic opportunistic infections like viral retinitis. We present a rather rare case of elderly male reporting with reduced vision and choroidal detachment (CD). He was investigated thoroughly and was found to have low CD4 counts with a HIV negative status.

Case report

A 69 year old patient presented with profound reduction of vision in his left eye of 15 days duration. He gave a history of intermittent fever preceding the episode. He denied any history of pain, redness, flashes floaters, trauma or surgery to the fellow eye and unexplained weight loss . He was a known case of diabetes mellitus and hypertension for the past 10 years, optimized on medication.

At presentation, the visual acuity was 20/20 in right eye (RE) and hand motions in left eye (LE). RE anterior segment and fundus was normal. He had relative afferent pupillary defect in the LE. Rest of the anterior segment was normal except posterior chamber intraocular lens in the capsular bag. Anterior chamber was quiet with no evidence of cells or flare. Fundus evaluation revealed elevated brownish mounds 360 degree in LE suggestive of peripheral choroidal detachment (CD). The retina was attached without any evidence of vasculitis, retinal hemorrhages or disc edema. (Figure 1 A) Extra ocular movements were full and free. Intraocular pressure was 14 mm Hg by non-contact tonometry in both the eyes. He was subjected to multimodal imaging.

On fundus fluorescein angiography (FFA) multiple pin point areas of hyper fluorescence were noted with late staining of the disc. (Figure 1 B,C) Optical coherence tomography (OCT) showed a neuro-sensory detachment with sub retinal precipitates without any evidence of intraretinal fluid. (Figure 1 D) Ultrasound B scan in the left eye showed 360 degrees choroidal detachment. There was no intra-ocular mass noted on B-scan. No subtenon fluid was evident. (Figure 2 A-C) UBM confirmed a ciliochoroidal detachment. No gross pathological choroidal or orbital mass lesion was noted in the orbit on MRI however orbital inflammation was suspected.

Baseline blood investigations including a complete hemogram, peripheral blood smear, lipid profile, renal and liver function test and thyroid profile were normal. Detailed uveitis workup including ESR, CRP, Mantoux test, serum angiotensin converting enzyme levels, chest x ray, syphilis serology, TORCH titers and HIV serology were normal.

LE Orbital inflammation was suspected on MRI, and a trial of oral steroids along with antibiotic cover was given. Over a period of next 2 weeks, he showed slight improvement in vision in left eye to counting fingers close to face (CFCF). (Figure 3) Over a period of 1 month his CD were persistent but reduced in height, however his poor vision was unexplained. He was further investigated for CD4 counts, and a repeat HIV test. The HIV was repeated with Tridot and ELISA which was negative. His CD4 count was 57 cells/ mm3 and was reconfirmed twice. During his last follow-up after 4 months his CDs had resolved with reduction in subretinal fluid on OCT but the vision remained stable. (Figure 3 A-D)

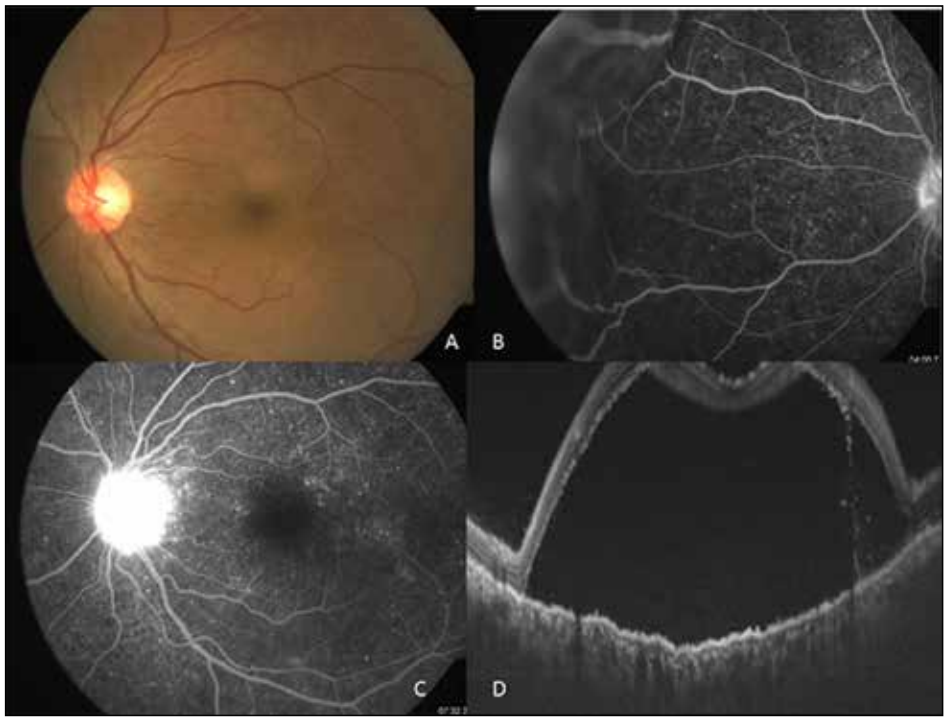

Figure 1: *(A) LE Fundus photograph at presentation (B,C) LE FFA showing peripheral CD and multiple hyper florescent dots (D) LE OCT showing neurosensory detachment and choroidal granularity*

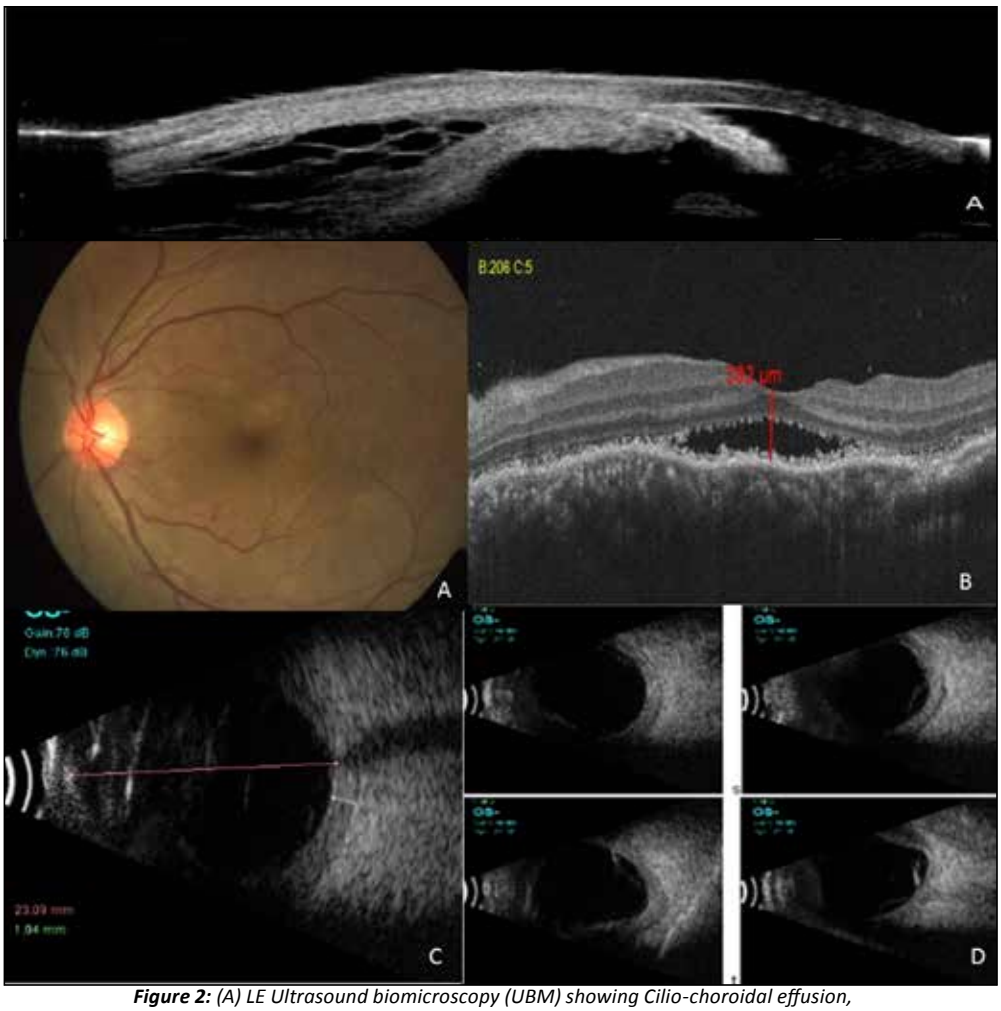

Figure 2: *(A) LE Ultrasound biomicroscopy (UBM) showing Cilio-choroidal effusion, (B, C) Ultra sound B scan LE showing thickened choroid and peripheral choroidal detachment*

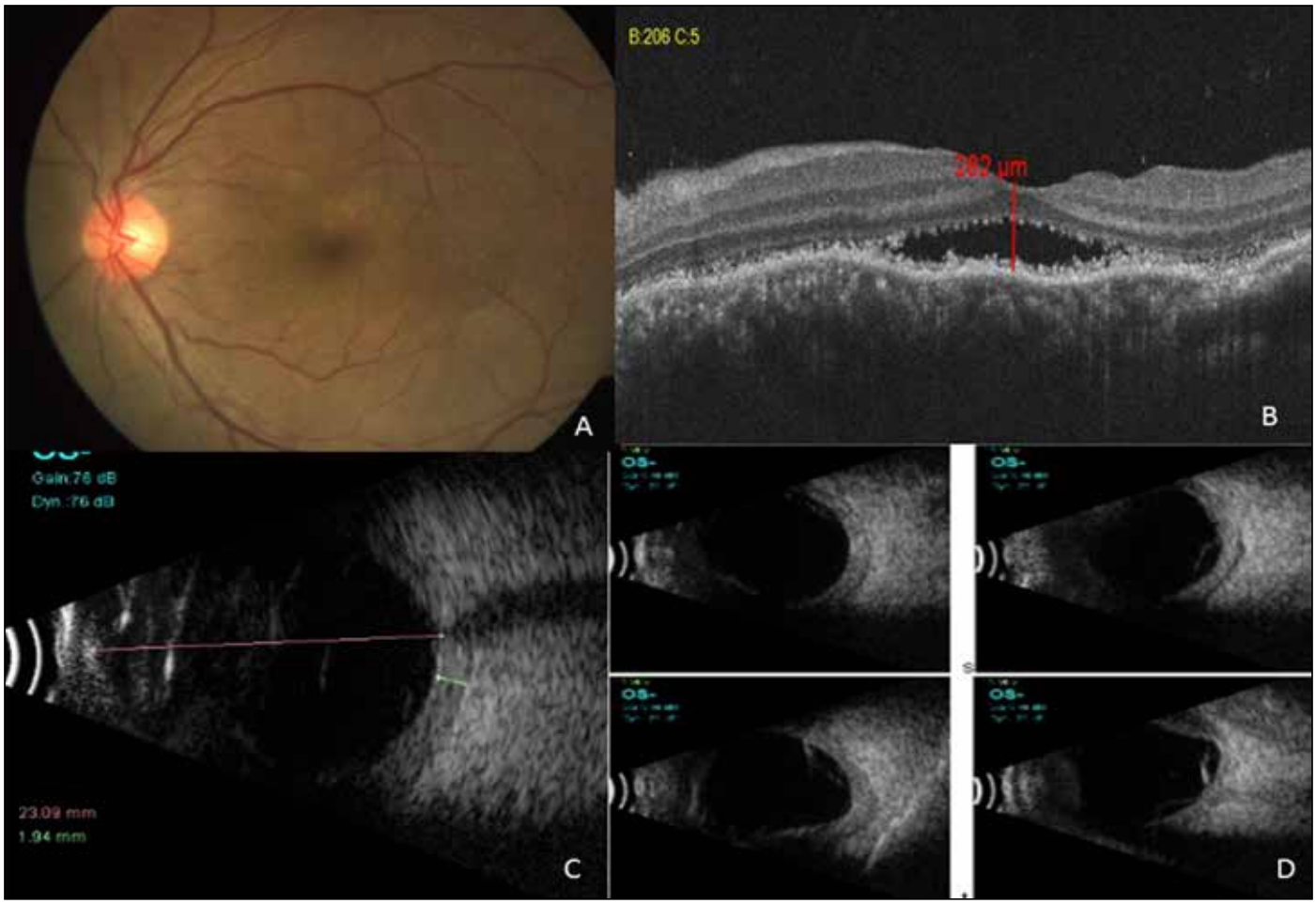

Figure 3: *(A) LE fundus picture at 4 months follow up*
(B) LE OCT showing reduced neurosensory detachment and choroidal granularity at 4 months
(C, D) Ultra sound B scan LE resolved peripheral CDs seen with reduction of choroidal thickening.

Discussion

Idiopathic CD4 lymphopenia (ICL) is a rare disorder characterized by low CD4 counts in the absence of HIV infection. A cut off value of 300 cells/ mm3 or count less than 20% of total WBC on two separate occasions has been used in various studies.[2] The common manifestations of ICL are opportunistic infections like cryptococcal meningitis, tuberculosis, human papilloma virus infection. The ophthalmic manifestations are very rare. On peer reviewed literature search, very limited data is available on patients of ICL presenting with only ophthalmic manifestations. Progressive outer retinal necrosis (PORN) has been reported by Gupta et al in 2 cases of ICL.[3] They concluded that Varicella zoster necrotizing retinitis, which is classic for PORN can occur in HIV negative patients. Idiopathic CD4 lymphocytopenia should be considered in healthy patients who develop ocular infections as seen in the immunocompromised patients. Hoang et al reported a case of recurrent cytomegalovirus (CMV) retinitis in an HIV-negative patient with CD4+ T lymphocytopenia.[4] A similar case of CMV retinitis was presented by Sloan et al in HIV negative patient.[5]

This case of choroidal detachment in ICL is unique because of its rarity. CD in this case occurred most probably due to unexplained inflammation in the choroid. The exact cause of CD in ICL is not known but the disc leak on FFA and resolution of effusion on oral steroids point towards inflammatory pathology. This case highlights an unreported infrequent presentation of a rather rare disease. The ophthalmologist should have a high index of suspicion to identify other life-threatening diseases and opportunistic infections associated with ICL. The challenge in our case lies in avoiding the similar complications in the fellow eye, in contrast to HIV where in drugs can help to improve the CD4 cell count.

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