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CRVO IN PEDIATRIC AGE GROUP: A RARE MANIFESTATION IN KIKUCHI DISEASE

Ophthalomology

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ABSTRACT

Introduction: Kikuchi Fujimoto is a rare necrotizing lymphadenitis disease. We report a case of a 17-year-old boy, known case of Kikuchi Fujimoto disease (KFD), who developed left eye central retinal vein occlusion (CRVO). **Case Presentation:** A 17-year-old boy presented with left eye sudden diminution of vision. He was diagnosed with Kikuchi Fujimoto necrotizing lymphadenitis disease 2 years back. He had left eye central retinal vein occlusion with disc and macular edema. Optical coherence tomography (OCT) showed subretinal fluid, cystoid macular edema along with disc elevation. He was on s Hydroxychloroquine since 6 months. After systemic evaluation he was started on oral prednisolone 40mg on weekly 10mg tapering dose. After a month his macular edema as well as disc edema had significantly resolved. **Conclusion:** KFD is itself is a rare entity and additional manifestation of CRVO in them, makes it rarest of rare presentation. If not diagnosed on time and treated can lead to permanent vision loss.

KEYWORDS

Kikuchi Fujimoto Disease; Central retinal vein occlusion

INTRODUCTION:

Kikuchi–Fujimoto disease (KFD) is a known cause of necrotizing lymphadenitis and is usually a self-limited cause of fever and lymph node enlargement. Patients are generally females with female/male ratio of 4:1 with a mean age of 21 years(1). Disease usually presents with fever and nodules appearing on the neck. Immunohistochemical analysis shows the histocytes in the biopsy were both MPO (myeloperoxidase)-positive and CD68-positive pointing to histiocytic necrotizing lymphadenitis(2). Ocular manifestations of KFD include uveitis, Vasculitis, lid edema, and proptosis(3). We present here a case of 17-year-old boy, known case of Kikuchi disease, with left central retinal vein occlusion with macular edema.

Case Presentation:

A 17-year-old boy presented with sudden onset diminution of vision in his left eye for 2 days. Right eye vision is 6/6 and left eye was 6/36 with an Intra-ocular pressure in both eyes was 16 mmHg. Pupils were regular round and reactive, clear lens and anterior segment examination was within normal limits. Dilated fundus evaluation right eye appeared normal while the left eye showed disc edema (figure 1A) with tortuous vessels and flame shaped hemorrhages and also associated macular edema as seen in figure 1B. Optical coherence tomography (OCT) shows presence of subretinal fluid (SRF) below the macula with an intact foveal contour (Figure 1C).

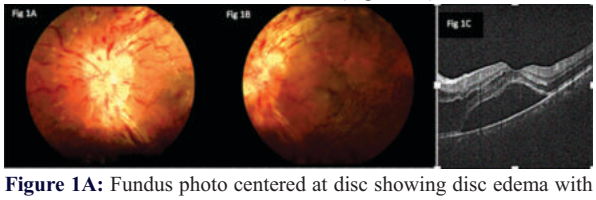

Figure 1A: Fundus photo centered at disc showing disc edema with tortuous vessels and flame shaped hemorrhages
Figure 1B: Fundus photo centered at macula showing macular edema
Figure 1C: OCT macula shows sub retinal fluid

He had history of fever 2 years back which lasted for almost 6 months. Swelling over the left side of his neck was persistent since then. A thorough investigations showed, normal blood counts and normal liver function test (LFT). His Mantoux test and Quantiferon gold tests were negative. High-resolution computed tomography (HRCT) chest and neck were within normal limits. Rheumatological investigations like Rheumatoid factor (RA), Anti-nuclear antibody (ANA), anti-ds DNA, anti-neutrophilic cytoplasmic antibodies (ANCA) were negative. No abnormality was detected in his USG abdomen. Cervical node biopsy showed changes corresponding KFD disease like histiocytic necrotizing lymphadenitis. He was started on Hydroxychloroquine 5mg/kg/day therapy for the same.

From ophthalmic point of view, we investigated him for his coagulation profile. His APLA profile (Anti phospholipid antibody) showed normal IgG (0.237) but a raised IgM (11.795). Hence with collaborations with the rheumatologists we started him on oral prednisolone 40mg in 7 days tapering manner, and topical NSAIDS Nepafenac 0.1% eyedrops three times a day for 3months. At one month follow up, his vision had improved to 6/9. Macular edema (Fig 2A) and disc edema (Fig 2B) was significantly reduced. OCT showed significant reduction in amount of SRF (Fig 2C). Patient showed significant symptomatic improvement.

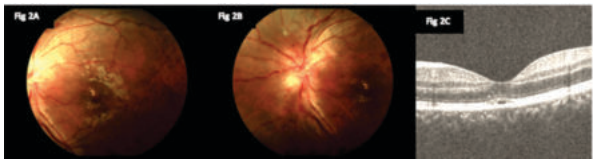

Figure 2A: Fundus photo centered at macula showing macular edema and tortuous vessels and flame shaped hemorrhages had significantly decreased
Figure 2A: Fundus photo centered at disc showed decreased edema
Figure 2C: OCT macula showing substantial reduction in sub retinal fluid

DISCUSSION:

Central retinal vein occlusion (CRVO) is most commonly seen in the age group of above 45 years of age. The most commonly associated risk factors include hypertension, diabetes, hyperlipidemia, smoking etc. (3). But our patient is a 16 year old boy with none of the risk factors. Hence, we got his coagulation profile done which indicated an increase in APLA and aided us in our diagnosis.

In KFD histopathological report shows irregular paracortical areas of coagulative necrosis with abundant karyorrhectic debris. This necrosis and debris can distort the nodal architecture. Large numbers of different types of histiocytes and thrombosed vessels are seen at the margin of the necrotic areas. Electron microscopic studies have also identified tubular reticular structures in the cytoplasm of lymphocytes and histiocytes. These changes are noted in the endothelial cells, leading to hypercoagulable state leading to thrombotic complication like vein occlusion. Probably this is the reason for the hypercoagulable state of our patient, which lead to central vein occlusion. A similar case has been reported by Zou in 2007, wherein a 22 year old female presented with bilateral occlusive retinal vasculitis(4)

CONCLUSION:

KFD is a rare, underdiagnosed, self-limiting disease of unknown cause. It is often misdiagnosed as tuberculosis or malignancy. Correct

and accurate diagnosis of the disease leading to CRVO prompted us to accurate management, which in turn helped in saving the vision of the patient. Close long-term follow-up is essential for such patients.

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