Recurrent neovascularization of the disc in sympathetic ophthalmia

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Sympathetic ophthalmia following parsplana vitrectomy is a known complication. We describe here a case of recurrent disc neovascularization in a patient of sympathetic ophthalmia. It promptly responded to steroids initially but later recurred with inflammation.

Key words: Methotrexate, neovascularization of the disc, sympathetic ophthalmia, triamcinalone

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Sympathetic ophthalmia (SO) is a well-known cause of chronic granulomatous uveitis. Neovascularization of the disc (NVD) may occur in such chronic uveitis, however, it has not been reported in SO to the best of our knowledge. Neovascularization of the disc in this case was also of a recurrent nature.

Case Report

A 25-year-old female patient presented with 2 months history of gradual painless progressive diminution of vision accompanied by floaters in the left eye. She had been treated with systemic and topical steroids with a presumptive diagnosis of pan uveitis with disc edema at a primary health care center and was referred. Her history revealed that 8 months earlier she had undergone right eye parsplana lensectomy and vitrectomy (PPL + PPV) with intravitreal antibiotics at a different center, for suspected metastatic endophthalmitis secondary to post partum abscess. Culture reports were, however, not available. There was no visual improvement following surgery and she had developed phthisical changes.

On examination best corrected visual acuity (BCVA) was no light perception in right eye while left eye had counting fingers (CF) close to face with accurate light projection. In the left eye apart from fine keratic precipitates, a 3+ cellular reaction was noted in both the anterior chamber (AC) and vitreous. Lens and intraocular pressure were normal. Fundus examination showed the presence of hyperemic disc with blurred disc margins and tortuous dilated vessels [Fig. 1A]. It was associated with serous retinal detachment with shifting fluid. Fundus fluorescein angiography (FFA) showed multiple tiny pinhead-sized hyperfluorescent spots in the superior half of the retina [Fig. 1C and D] with an inferior serous retinal detachment. Late phase showed disc hyperfluorescence with blurring of margins [Fig. 1B]. Systemic examination was normal which included hearing tests and dermatological examination. A clinical diagnosis of SO was made and intravenous pulse steroids (Dexamethasone 100 mg in 150 ml of 5% dextrose) for three days along with topical steroids and cycloplegics were started. Visual acuity improved to 13/200 on day four. Patient was continued on once daily oral steroids at 1 mg/kg body weight. However, 2 weeks after therapy patient started to develop side-effects to steroids and hence methotrexate 15 mg/week and folic acid 5 mg were added while steroid dose was reduced gradually by 10 mg/week.

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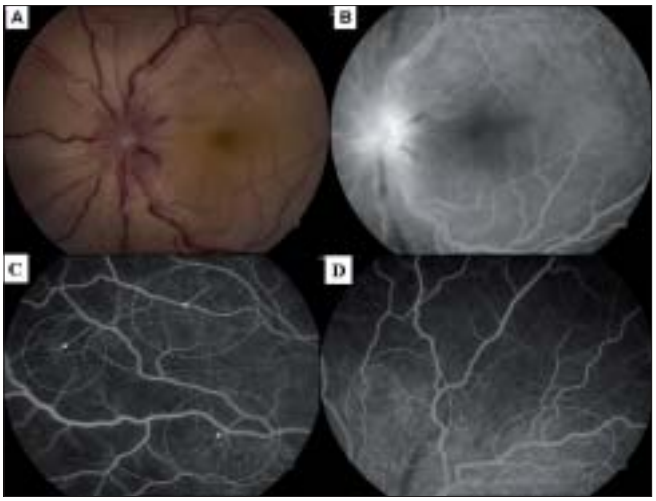

Figure 1: (A) Fundus at presentation showing hyperemic disc with blurred disc margins and tortuous dilated vessels. (B) Fundus uorescein angiography (FFA) showing disc hyper uorescence with blurring of margins in late phase. (C and D) FFA showing multiple areas (encircled) of pinhead-sized leaks (arrow)

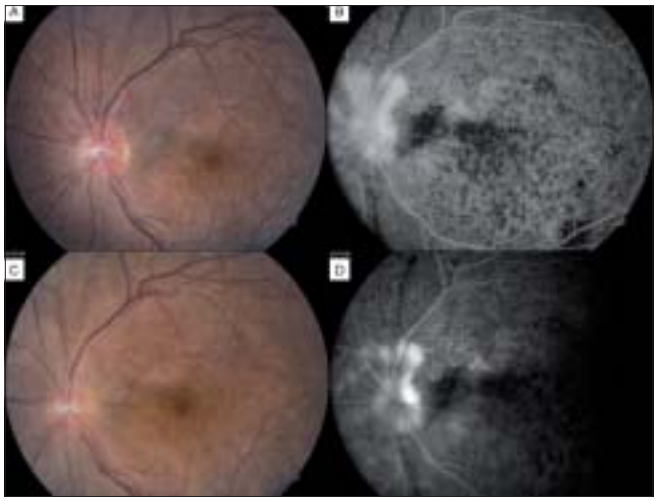

Figure 2: (A) Change after 14 weeks of methotrexate, abnormal ne branching vessels seen on the disc. (B) Neovascularization of disc (NVD) was con rmed on the fundus uorescein angiography (FFA). (C) Three months after the pan retinal photocoagulation, the vessels appeared to have regressed clinically. (D) Reduced leakage on FFA compared to initial presentation

Six weeks after starting methotrexate, cellular reaction reduced to 1+ in vitreous and no cells in AC with complete resolution of retinal detachment and the BCVA was 20/60. Patient was continued on these medications and was put on a monthly follow-up with close monitoring of the blood counts and liver function. After 14 weeks of starting methotrexate, patient was noted to have developed abnormal fine branching vessels on the disc suggestive of neovascularization and this was confirmed on the FFA [Fig. 2A and B]. Vitreous examination revealed 1+ cell, however, there was no activity in the AC. Patient was treated with pan retinal photocoagulation (PRP) and a posterior subtenon (PST) injection 0.5 ml of 40 mg/ml triamcinalone acetonide while methotrexate was continued. Three months after the PRP, the vessels appeared to have regressed clinically as well as on FFA compared to the initial presentation [Fig. 2C and D].

Two months later the patient presented with diminution of vision. On examination her BCVA was 10/200, which rapidly deteriorated to CF close to face in two days. There was no activity in the anterior segment while there were 2+ cells in the vitreous. Fundus examination revealed recurrence of NVD and multifocal serous detachments involving the macula, however, there was no peripheral serous detachment [Fig. 3A and B]. The patient was given a repeat PST injection of triamcinalone acetonide and the oral steroid was restarted at 1 mg/kg while continuing with methotrexate. Ten days after starting the steroids serous detachments resolved and the vision improved to 20/200. Six weeks later the vision recovered to 20/60. The steroid dose was gradually tapered over a period of 12 weeks and maintained at 20 mg/ day with methotrexate reduced to 7.5 mg/week.

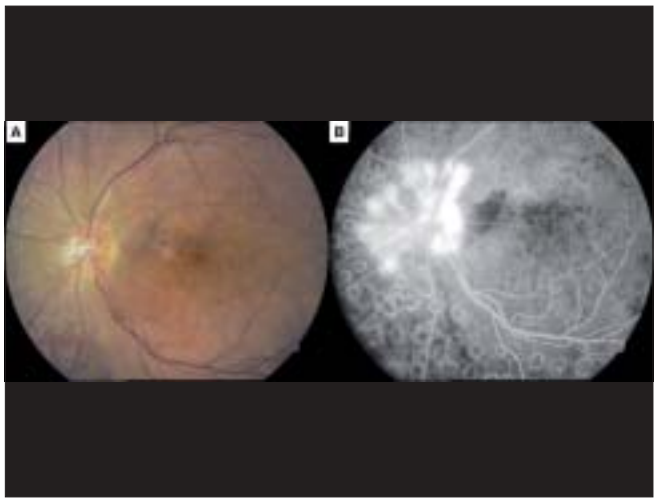

Figure 3: (A) Recurrence of NVD with reactivation of in ammation. (B) Fundus uorescein angiography showing increased leakage from NVD with diffuse leak in the macular area

Discussion

Sympathetic ophthalmia is a rare sight-threatening bilateral panuveitis with an incidence of 0.03/100000.[1] In our case, SO occurred after parsplana vitrectomy which is a rare occurrence. The reported incidence of SO following vitreoretinal surgeries is 1 in 800[1] and has shown an increasing trend, especially with repeated surgeries. A corresponding risk following external retinal detachment repair is 1 in 1357.[1]

Kilmartin *et al.*,[1] in their study have noted that the current SO risk following vitrectomy is more than twice that previously reported by Gass (0.06%).

Based on their study[1] the same authors in a commentary on SO[2] have suggested that it would be appropriate to counsel patients regarding the risk of SO before the vitrectomy procedure. Pollack *et al*.[3] in the largest case series of SO following vitrectomy without a previous trauma have noted that SO following such surgical procedure may have diverse presentations and any atypical or persistent uveitis following vitreous surgery should alert the treating surgeon of possible SO.

Another important manifestation in this case was NVD. Although occurrence of NVD is known in Behcet's disease, chronic uveitis[4] and Vogt Koyanagi Harada disease,[5] it has not been reported in SO to the best of our knowledge. NVD was of recurrent nature with the recurrence of inflammation.

In our patient the clinical setting of previous ocular surgery and findings in the left eye were consistent with a diagnosis of SO. Early and prompt use of immunosuppressive therapy with systemic steroids and steroid-sparing agents such as cyclosporin A,[6] azathioprine,[6] chlorambucil have improved the prognosis in patients with SO. In our case we were able to use methotrexate to successfully control the inflammation after initial treatment with a combination of intravenous, oral and topical steroids.

After the initial control of inflammation the patient developed NVD after 14 weeks of immunosuppressive treatment. Considering that this was the only seeing eye of the patient and also the lack of a well-defined protocol for treating such NVD in chronic uveitis, we decided to treat the patient with PST injection of triamcinalone acetonide. A PRP was also done considering the one-eyed status although there was no ischemia on FFA. The combined therapeutic approach was initially effective and resulted in regression of the NVD after 12 weeks. However, the NVD recurred within 8 weeks and was associated with multifocal serous detachments and inflammation in the vitreous cavity. On restarting steroids with another PST injection of triamcinalone acetonide the serous detachments resolved within four days.

Although the initial occurrence of NVD was not associated with increased activity in vitreous its prompt response to steroids (systemic + PST) and its later recurrence with inflammation is suggestive of an inflammatory pathomechanism. The role of PRP in this case is unclear, however, the recurrence of NVD with flare-up of inflammation suggests that PRP may not be useful in this setting. We suggest that systemic immunosuppression along with repeated PST injection of triamcinalone acetonide may be useful in treating such NVD in chronic uveitis.

References

1. Kilmartin DJ, Dick AD, Forrester JV. Prospective surveillance of sympathetic ophthalmia in the United Kingdom and Republic of Ireland. Br J Ophthalmol 2000;84:259-63.
2. Kilmartin DJ, Dick AD, Forrester JV. Sympathetic ophthalmia risk following vitrectomy: Should we counsel patients? Br J Ophthalmol 2000;84:448-9.
3. Pollack AL, McDonald HR, Ai E, Green WR, Halpern LS, Jampol LM, *et al*. Sympathetic ophthalmia associated with parsplana vitrectomy without antecedent penetrating trauma. Retina 2001;21:146-54.
4. Sanislo SR, Lowder CY, Kaiser PK, Gutman FA, Zegarra H, Dodds EM, *et al*. Corticosteroid therapy for optic disc neovascularization secondary to chronic uveitis. Am J Ophthalmol 2000;130:724-31.
5. To KW, Nadel AJ, Brockhurst RJ. Optic disc neovascularization in association with Vogt-Koyanagi-Harada syndrome. Arch Ophthalmol. 1990;108:918-9.
6. Vote BJ, Hall A, Cairns J, Buttery R. Changing trends in sympathetic ophthalmia. Clin Exp Ophthalmol 2004;32:542-5.