Idiopathic thrombocytopenic purpura and its fundus features in a patient with diabetes mellitus

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In this case report, we present a patient with thrombocytopenia secondary to idiopathic thrombocytopenic purpura (ITP), whose fundus appearance had features of diabetic retinopathy with macular edema. The macular edema did not respond to multiple intravitreal Anti-Vascular endothelial growth factor (Anti-VEGF), contrary to diabetic cystoid macular edema (CME). He was systemically investigated and was found to have ITP, and its management resulted in complete regression of the hemorrhages and CME.

Key words: Diabetic retinopathy, idiopathic thrombocytopenic purpura, macular edema

Macular edema is the leading cause of vision loss in patients with diabetic retinopathy.

Timely and appropriate management, which not only includes diagnosing but also identifying the confounding factors and mimickers goes a long way in preventing redundant treatment.

We present a case of idiopathic thrombocytopenic purpura (ITP) with fundus picture akin to moderate nonproliferative diabetic retinopathy (NPDR) and posed a diagnostic dilemma but responded to unconventional therapy consisting of systemic steroid therapy for thrombocytopenia.

Case Report

A 63-year gentleman presented with complaints of decreased vision in both eyes left more than the right. He was a known diabetic with moderate control (HbA1C of 8.6%). He was diagnosed elsewhere with moderate NPDR with macular edema for which he had already received multiple anti-VEGF injection to both eyes without much response (persisting cystoid spaces at macula on OCT). He had received the last injection to both eyes, 2 weeks before presenting to us.

His BCVA was 6/18 in right eye, while the left had 6/36. Ocular evaluation disclosed grade 1 nuclear sclerosis in both eyes with posterior subcapsular cataract only in left. Dilated fundus showed multiple large dot and blot retinal hemorrhages, while the OCT confirmed persistent macular edema with CMT of 346 µ in right eye and 338 µ in his left [Fig. 1a-d]. Since his fundus picture consisting of large retinal hemorrhages did not resemble to DR, we asked for systemic evaluation which included complete haemogram, renal and lipid profile. His platelet count was significantly less at 75,000/mm3, while the rest of the blood parameters was normal. He was referred to a haemato-pathologist where he was diagnosed with ITP (with bone marrow biopsy). Oral steroid was started, and the fundus picture improved despite altered glycaemic control albeit for a short duration. No other intervention was planned or given.

The patient was followed up periodically and was noted to have betterment in his fundus picture. At 7 months of follow-up most of the retinal hemorrhages had decreased with complete resolution of the macular edema [Fig. 2a and b]. Right eye showed conglomeration of hard exudate in the subfoveal region. Despite this, he was followed up without any active intervention.

At 16 months of follow-up, the patient complained of sudden onset painless vision loss in his left eye. He had developed vitreous hemorrhage in his left eye without any changes in the fundus. Fundus fluorescein angiography did not reveal any neovascularization [Fig. 3a-d] and there was no posterior vitreous detachment (PVD). His platelet count correlated with this incident was noticed to have decreased (68,000 cells/mm3). He was managed conservatively and at the end of three years his fundus was near normal with significant regression of retinal and pre-retinal hemorrhages, hard exudates and edema [Fig. 4a and b]. Both eyes had few residual retinal hemorrhages and micro-anuerysms. Left fundus view was hazy due to progression of the cataract.

Discussion

ITP is a relatively uncommon autoimmune haematologic condition, wherein platelets are destroyed leading to thrombocytopenia. Ophthalmic involvement in ITP is rare and mostly limited to subconjunctival hemorrhage,[11] preretinal, intraretinal, subretinal and/or vitreous hemorrhage,[2-6] which may be associated with intracranial bleeding in a Terson type phenomenon,[7] hemorrhage within the optic tract[8] and non-arteritic anterior ischaemic optic neuropathy.[9]

Our patient presented to us with persistent edema and hemorrhages, akin to fundus manifestation of diabetic
Indian Journal of Ophthalmology
Volume 68 Issue 11

Though ocular hemorrhages are common in ITP, large retinal hemorrhages as presented in our patient without underlying anemia has never been reported earlier. This fundus picture leads to the diagnostic dilemma, and probably the reason for the previous ophthalmologists to consider injecting intravitreal anti-VEGF.

Nonresponse to multiple anti-VEGF and the presence of “Large dot and blot retinal hemorrhages” prompted us to re-evaluate the patient for systemic status. This revealed thrombocytopenia (secondary to ITP) and its pathogenesis. The patient was treated with systemic steroids as a standard protocol for ITP. He responded favorably with an increase in the platelet count as well as clearing of the retinal hemorrhages and macular edema. This improvement was despite the fact that the patient’s glycaemic control was altered due to systemic steroids. This gives us an indirect evidence that the manifestation was due to the underlying thrombocytopenia rather than diabetes.

The patient developed spontaneous vitreous hemorrhage at the end of 16 months from the presentation. FFA was also done which did not reveal any neovascularisation, nor any posterior vitreous detachment. Spontaneous vitreous and sub-retinal hemorrhage is associated with ITP and well documented. This prompted us to follow conservative management. Three years after the initial presentation, fundus picture had completely normalized with stable platelet count. The patient had been completely weaned off from medications.

Conclusion
Retinal and vitreous hemorrhages may be indicative of an underlying blood dyscrasia and their presence without any known underlying etiology warrants a thorough systemic evaluation. ITP may present with subretinal, preretinal, and vitreous hemorrhages and a high index of suspicion along with a comprehensive systemic examination and appropriate laboratory work-up aids in diagnosing this grave disease. The ophthalmic manifestations respond adequately to correction of hematologic parameters and no additional therapy is warranted.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.
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