Case Report

Bilateral simultaneous multilayered retinal hemorrhages in a young adult with Idiopathic Thrombocytopenic Purpura (ITP): A rare case report

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Abstract

Introduction: Idiopathic thrombocytopenic purpura is an autoimmune hematological disorder characterized by isolated low platelet count without any known clinically apparent etiology. Although usually most cases are asymptomatic, it may present with bleeding manifestations in forms of bruises, petechiae, nose bleeding etc. Very rarely, it may present with ocular manifestations in the form of subretinal, preretinal, intra-retinal or vitreous haemorrhage.

Case: A 26-year-old female presented to the ophthalmology department with chief complaint of sudden painless diminution of vision in both eyes for the last one week. Her best corrected visual acuity for distance was 20/200 in right eye and 20/600 in left eye. Dilated fundus examination showed right eye sub-internal limiting membrane (ILM) haemorrhage, left eye large subretinal and intra-retinal haemorrhages over posterior pole, with Roth's spot in both eyes. Haematological investigations revealed moderate anaemia (Hemoglobin level of 7 gm %) and severe thrombocytopenia (25,000/ ul). Diagnosis of idiopathic thrombocytopenic purpura (ITP) was made after consultation with an internist. She was treated with systemic steroid and blood transfusion for systemic disease and kept under regular ophthalmic follow up. Over a period of six months, she showed gradual near total resolution of retinal hemorrhages in both eyes and improvement in visual acuity in both eyes.

Conclusion: This case gives a unique outlook to simultaneous varied ocular manifestations of ITP in a single patient and stresses upon thorough systemic evaluation in the presence of such manifestations. Optimization of systemic parameters without active ocular intervention in select cases may achieve favourable outcome.

Key words: Idiopathic thrombocytopenic purpura, Sub-internal limiting membrane haemorrhage, Retinal haemorrhages, Roth's spot.

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Introduction

Idiopathic thrombocytopenic purpura (ITP) is an autoimmune disease in which auto-antibodies are directed against platelets leading to thrombocytopenia which subsequently leads to their sequestration in spleen and resultant bleeding diathesis. ITP is mostly

a benign asymptomatic disorder. Rarely, ophthalmological involvement is seen (Holt JM et al., 1969). Here, we report a unique case of bilateral sudden onset diminution of vision in a young lady due to retinal hemorrhages at multiple levels (sub-internal limiting membrane, intra retinal, sub retinal and Roth's spot). Detailed systemic evaluation led to a diagnosis of ITP. Reversal of retinal hemorrhages and corresponding improvement in vision were noted gradually over six months after optimization of systemic parameters without any active ocular intervention.

Case history

A 26-year-old female presented with a complaint of sudden, painless diminution of vision in both eyes (BE) for the last one week. No history of trauma, headache, exposure to any toxic substance, or any significant systemic disease was elicited. Other than ocular complaints, she had a history of menorrhagia for the past three months but she did not seek any medical consultation for the same. Her best corrected visual acuity (BCVA) was 20/200 in right eye (RE) and 20/600 in left eye (LE). Extraocular movements were normal and painless in BE. Anterior segment examination including pupillary reactions were unremarkable in BE. Dilated fundus examination at presentation showed large premacular haemorrhage and few retinal haemorrhages with white centre (Roth's spots) in the RE (Figure 1a); large retinal hemorrhage over macula along with peripapillary Roth's spots in the LE (Figure 1b). The retinal vessels and the optic disc appeared normal in both eyes. Spectral domain optical coherence tomography (SD-OCT) of RE (Figure 2a) revealed elevated hyper reflective material beneath a hyper reflective membrane with posterior back shadow raising the probability of sub internal limiting membrane (ILM) or subhyaloid haemorrhage. SD-OCT of LE (Figure 2b) revealed the presence of subretinal and intraretinal haemorrhage with few intraretinal cystoid spaces. Physical examination by an internist showed multiple petechial rashes distributed all over the body. Laboratory workup demonstrated thrombocytopenia (platelets: 25000/µl) and moderate anaemia (Hb: 7 g/dl). Rest of the peripheral blood smear, coagulation profile, liver function tests, and renal function tests were unremarkable. Tests for HIV 1,2 and other common viral markers (HbsAg, HCV) were negative. Haematologist opinion was sought and a diagnosis of ITP was made. She was started on systemic steroids (intravenous methylprednisolone 1g/day for five days followed by oral steroids 1mg/kg body weight in tapering dose) and three units of whole blood transfusion were also done according to recommendation by haematologist. Follow up at 1 month revealed partial resolution of premacular bleed in RE (Figure 1c) with SD-OCT (Figure 2c) confirming presence of sub ILM bleed which has become organised. LE at 1 month similarly showed significant resolution of retinal haemorrhages with appearance of few hard exudates (Figure 1d) after decrease in retinal thickness (Figure 2d). She was managed with control of systemic parameters without any active ocular intervention. On subsequent follow up at six months, fundus examination revealed near total resolution of retinal haemorrhages and retinal edema (Figure 1e-f & 2 e-f) in BE with corresponding improvement in BCVA in BE. At final follow up at eight months, BCVA improved to 20/30 in RE and 20/25 in left eye with platelet count improved to 1, 30,000/µl and Hb level of 10 g/dl. She was advised monthly follow up at both ophthalmology and haematology clinic for further monitoring.

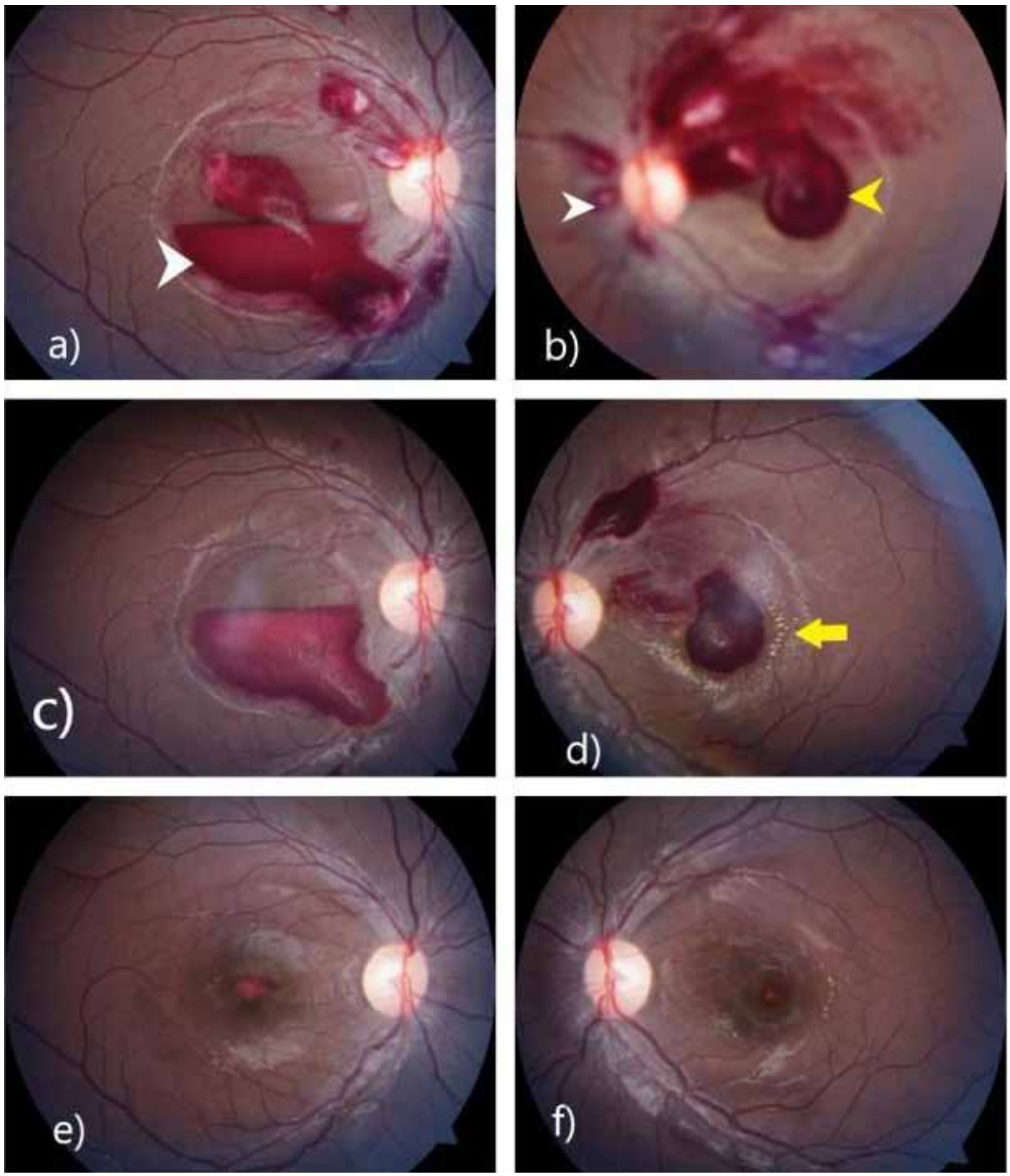

Figure 1: (a-b) Colour fundus photograph at presentation showing a large boat shaped premacular haemorrhage (white arrowhead) with few Roth's spots in right eye (RE) (1a); Large retinal haemorrhages at the posterior pole (yellow arrowhead) and peripapillary roth spots (white arrow head) in left eye (LE) (1b).

At one month follow-up (c-d), RE (c) showed resolution of Roth's spots and organised sub ILM haemorrhage. LE (d) showed partial resolution of retinal hemorrhages and appearance of a ring of hard exudates around fovea (yellow arrow).

At six months follow-up (e-f), both RE (e) and LE (f) showed near complete resolution of haemorrhages.

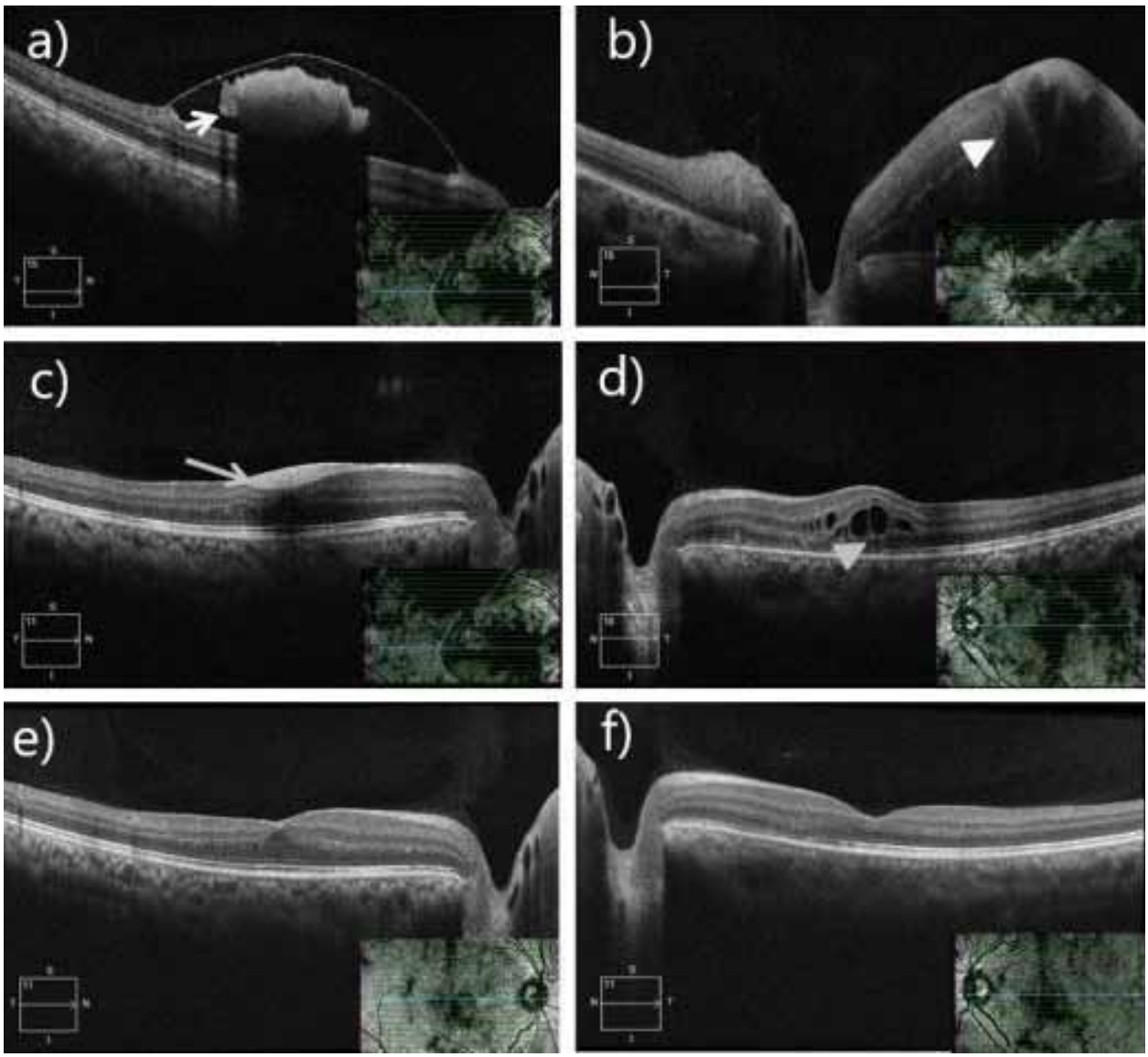

Figure 2: OCT macula of right eye (RE) (a) showing haemorrhage at subILM level (White arrow) and posterior back shadowing due to it. Left eye (LE) (b) shows an area of intraretinal and subretinal haemorrhage (White arrowhead) along with few intra retinal cystoid spaces.

At 1 month follow up (c-d), mild sub ILM haemorrhage (grey arrow) was noted at fovea of RE (c). LE (d) showed marked reduction in central macular thickness (CMT) with resolution of haemorrhages; few residual cystoid spaces at fovea (white arrowhead) were present.

Six months later (e-f), both eyes showed further reduction in CMT with restoration of foveal contour to near normality.

Discussion

ITP is a relatively uncommon autoimmune haematological condition wherein circulating auto-antibodies act against surface antigens of host platelets causing destruction. Destruction of platelets lead to low platelet count, occasionally leading to subsequent complications of bleeding. The usual systemic manifestations of ITP are petechial haemorrhages, bleeding from mucosa or gums, gastrointestinal tract, vagina, etc (DiFino SM et al., 1978). Ocular features though rare, may manifest as subconjunctival haemorrhage, (Sodhi PK et al., 2003) preretinal, intraretinal, subretinal and/or vitreous haemorrhage, (Shah PA et al., 2005; Majji A et al., 2010) which may be associated with terson type phenomenon, (Frankel CA et al., 1990). In the index case, we have demonstrated the presence of retinal haemorrhages at different levels (sub ILM without terson phenomenon, intra retinal, subretinal and Roth's spot) in the same patient and managed successfully with control of systemic parameters only without any active ocular intervention. Such presentation of simultaneous multi-layered retinal hemorrhages in a single patient as in our case, has rarely been reported in the literature earlier. Nd: YAG membranotomy (NYM) is an alternative option for management of dense premacular sub ILM haemorrhages, however success is governed by multiple factors like duration of bleed and degree of clot formation,(Oh BL et al.,2018). Although NYM management option was kept in consideration for RE at presentation, it could not be performed as the patient did not turn up for follow up at ophthalmology clinic for a period of 1 month after initial presentation because of intensive treatment under haematologist. Follow up at 1 month showed organised clot in RE precluding the role of NYM at that stage. Retinal haemorrhages in LE were not amenable to NYM at any stage of presentation. The other alternative mode of intervention in the form of pars plana vitrectomy is recommended for non-resolving dense premacular haemorrhage usually after a period of 3 months,(Adan A et al, 2008). However, our patient was not systematically fit in the initial period and subsequently showed spontaneous gradual resolution of haemorrhages with improvement in BCVA obviating need for any invasive surgical intervention. Thus, this case report highlights the protean retinal manifestations of ITP in a single patient and role of meticulous comprehensive systematic evaluation of a patient presenting with such retinal haemorrhages. Mere optimisation of systemic parameters in select cases may help in successful resolution of ocular manifestations of ITP without any invasive measures. However, long term follow up in collaboration with physicians is necessary for early detection and management of any recurrences.

Conclusion

This case gives a unique outlook to simultaneous varied ocular manifestations of ITP in a single patient and stresses upon thorough systemic evaluation in the presence of such manifestations. Optimization of systemic parameters without active ocular intervention in select cases may achieve favourable outcome.

References

Adan A, Pelegrin L, Alforja S, Giralt J (2008). Optical coherence tomography findings and management of sub-internal limiting membrane haemorrhage.Acta Ophthalmol;86:582-83.

DiFino SM, Lachant NA, Kirshner JJ, Gottlieb AJ (1978). Adult idiopathic thrombocytopenic purpura in adults. Long term results in a series of 41 patients. Ann Clin Res;10:83–86.

Frankel CA, Pastore DJ (1990). Idiopathic thrombocytopenic purpura with intracranial hemorrhage and vitreous. Clin Pediatr (Phila); 29: 725–728.

Holt JM, Gordon-Smith EC (1969). Retinal abnormalities in diseases of the blood. Br J Ophthalmol;53:145–160.

Oh BL, Yu HG(2018).Different responses of two consecutive Nd:YAG laser membranotomies at different locations in a 3 week-old sub-internal limiting membrane haemorrhage.Indian J Ophthalmol;66:1321-2.

Majji A, Bhatia K, Mathai A (2010). Spontaneous bilateral peripapillary, subhyaloid and vitreous hemorrhage with severe anemia secondary to idiopathic thrombocytopenic purpura. Indian J Ophthalmol; 58: 234–236.

Shah PA, Yang SS, Fung WE (2005). Idiopathic thrombocytopenic purpura with massive subretinal hemorrhage. Arch Ophthalmol; 123: 1612–1613.

Sodhi PK, Jose R (2003). Subconjunctival hemorrhage: the first presenting clinical feature of idiopathic thrombocytopenic purpura. Jpn J Ophthalmol; 47: 316–318.