Haematologica 1995; 80:194-195

OCULAR INVOLVEMENT IN ACUTE THROMBOTIC THROMBOCYTOPENIC PURPURA

Enrico Bobbio-Pallavicini*, Camillo Porta, Alessandra Brocchieri, Anna Saporiti, Fiorenzo Tacconi*

**Divisione di Medicina Generale, Ospedale Maggiore, Crema; Istituto di Terapia Medica, Università di Pavia, IRCCS Policlinico San Matteo, Pavia, Italy*

Thrombotic thrombocytopenic purpura (TTP) is an uncommon blood syndrome characterized by a pentad of symptoms including hemolytic anemia on a microangiopathic basis, thrombocytopenia with consequently severe hemorrhagic diathesis, neurologic changes of varying severity and/or impaired renal function.[1]

Bleeding usually involves mainly the skin and mucosae, but the gastroenteric, urogenital, central nervous and visual systems can also be struck. Ocular involvement during TTP is usually an underestimated, though far from uncommon, event occurring in 14-20% of patients reported in different literature series.[2-4] Ocular manifestations in the course of TTP are mainly detectable in the preterminal stage,[5] which is the likely reason they are usually reported in the literature as anecdotal.[6-8]

A 33-year-old woman referred to our Institute for the sudden onset of metrorrhagia accompanied by major neurologic symptoms was submitted to hematochemical tests, and the results allowed a diagnosis of acute TTP to be made (7 g/dL hemoglobin, 18×10^9/L platelets, 543 mU/L LDH, undosable traces of haptoglobin, negative Coombs' test and no antiplatelet antibodies, schistocytes and rare immature medullary forms in peripheral blood smears). During the tests the patient started complaining of visual disturbances including dimness of vision and transient left eye amaurosis. *Fundus oculi* examination demonstrated two peripapillary preretinal hemorrhages of different extent, above and below the optic papilla in the left eye (Figure 1), with no retinal vessel alterations at fluoroangiography (Figure 2); pupil motion and reflexes were normal. Complete remission was achieved after combined plasma exchange, platelet antiaggregating substances and cortisone administration. After a short while retinal bleeding resolved almost completely (Figure 3).

To date, the most accurate clinical classification of ophthalmologic manifestations in the course of thrombotic microangiopathies (both TTP and hemolytic-uremic syndromes, HUS) has been Percival's.[3,9] He subdivided ocular lesions according to their being caused by specific ocular lesions or their being the symptoms of the systemic disorders caused by thrombotic microangiopathy. Retinal detachment, choroidal bleeding, homonymous hemianopia belong to the first group, with the last of these usually being caused by hemorrhagic lesions at the occipital poles of the cerebral hemispheres.

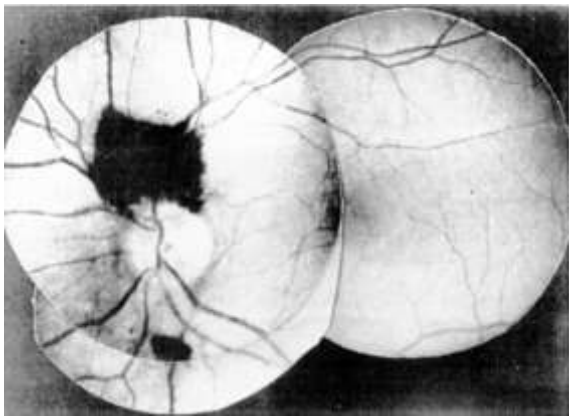

Figure 1 - Fundus examination of the left eye, performed right after acute TTP was diagnosed because of bilateral dimness of vision and transient left eye amaurosis, demonstrates two peripapillary preretinal hemorrhages of different extent, above and below the optic papilla.

Correspondence: Dr. Enrico Bobbio-Pallavicini, Divisione di Medicina Generale, Ospedale Maggiore di Crema, I-26013 Crema, Italy.
Received October 5, 1994; accepted January 11, 1995.

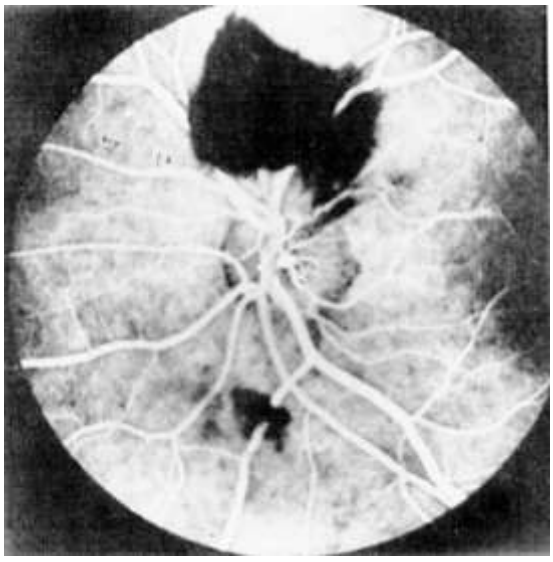

Figure 2. Subsequent fluoroangiography during the same visit showed no other retinal vessel alterations.

Diplopia, anisocoria, papilledema and retinal bleeding are also included in the first group.

In contrast, the ocular symptoms suggesting systemic damage include hypertensive retinopathy (especially in HUS), palpebral purpura or swelling, chemosis, subconjunctival bleeding, scleral jaundice and, finally, retinal bleeding.

In our patient, the ocular lesions had an atypical onset in the acute phase of the disease and were clearly demonstrated by fundus examination. They were likely caused by the severe thrombocytopenia our patient suffered from, rather than being secondary to systemic damage; this is supported not only by the disturbed vision that occurred in the acute phase, but also by fluoroangiographic findings which demonstrated no change whatsoever in the retinal vessels.

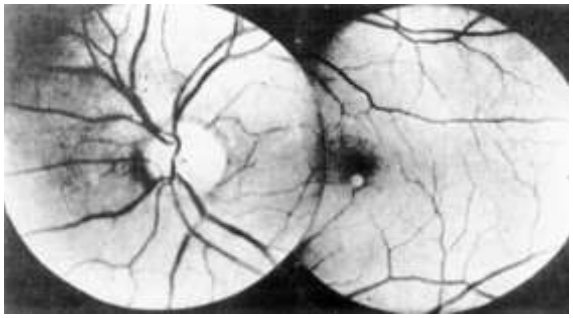

Figure 3. The ophthalmologic picture normalized after complete remission was achieved with plasma exchange, platelet anti-aggregating substances and cortisone administration.

To conclude, the case we have briefly reported must remind us that: a) the eye may be damaged in the course of TTP; b) such damage may occur even in the early phases of the disease, with possible subsequent diagnostic implications;[9] c) such symptoms can be cured, even in the short term, with adequate plasmapheresis (ref. #8 and this case) with or without the addition of plasma cryosupernatant[10] and, finally, d) this kind of ocular involvement may represent, as suggested by Douzinas et al.,[11] another prognostic factor in TTP patients, adding to and completing the classic score system by Eldor and Rose.[12]

References

1. Moschkovitz E. Hyaline thrombosis of the terminal arterioles and capillaries: a hitherto undescribed disease. Proc NY Pathol Soc 1924; 24:21-4.
2. Amorosi EL, Ultman JE. Thrombotic thrombocytopenia: report of 16 cases and review of the literature. Medicine 1966; 45:139-59.
3. Percival SPB. The eye and Moschkowitz's disease (thrombotic thrombocytopenic purpura). A review of 182 cases. Trans Ophtalmol Soc UK 1970; 30:375-82.
4. Ridolfi RL, Bell WR. Thrombotic thrombocytopenic purpura: report of 25 cases and review of the literature. Medicine 1981; 60:413-28.
5. Benson DO, Fitzgibbon JF, Goodnight SH. The visual system in thrombotic thrombocytopenic purpura. Arch Ophtalmol 1980; 12:413-7.
6. Lewellen DR Jr, Singerman LJ. Thrombotic thrombocytopenic purpura with optic disk neovascularization, vitreous hemorrhage, retinal detachment and optic atrophy. Am J Ophtalmol 1980; 89:840-4.
7. Slusher MM, Pugh H, Hackel R. Bilateral serous retinal detachment in thrombotic thrombocytopenic purpura. Arch Opthalmol 1990; 108: 744-5.
8. Wyszynski RE, Frank KE, Grossniklaus HE. Bilateral retinal detachments in thrombotic thrombocytopenic purpura. Graefe's Arch Clin Exp Ophtalmol 1988; 226:501-4.
9. Percival SPB. Ocular findings in thrombotic thrombocytopenic purpura (Moschkowitz's disease). Br J Ophtalmol 1970; 54:73-8.
10. Molinari E, Costamagna L, Perotti C, Isernia P, Pagani A, Salvaneschi L. Refractory thrombotic thrombocytopenic purpura: successful treatment by plasmapheresis with plasma cryosupernatant. Haematologica 1993; 78:389-92.
11. Douzinas EE, Markakis K, Karabinis A, et al. Early plasmapheresis in patients with thrombotic thrombocytopenic purpura. Critical Care Medicine 1992; 20:57-61.
12. Rose M, Eldor A. High incidence of relapses in thrombotic thrombocytopenic purpura: clinical study of 38 patients. Am J Med 1987; 83:437-44.