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 hematoochemical 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 fluorography (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.
Ocular involvement in TTP

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.

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; 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 cryosupernatant10 and, finally, d) this kind of ocular involvement may represent, as suggested by Douzinias et al.,11 another prognostic factor in TTP patients, adding to and completing the classic score system by Eldor and Rose.12

References