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SHORT COMMUNICATION

Case report

Bilateral non-arteritic anterior ischemic optic neuropathy in a patient with autoimmune thrombocytopenia

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PURPOSE. *To describe a patient with bilateral non-arteritic anterior ischemic optic neuropathy (NAION) and idiopathic autoimmune thrombocytopenia (ITP) with an extremely low platelet count.*
METHOD. *Case report.*
RESULTS. *Remarkably good recovery of visual acuity.*
CONCLUSIONS. *Bilateral non-arteritic anterior ischemic optic neuropathy can develop in the presence of a very low platelet count. (Eur J Ophthalmol 2000; 10: 180-2)*

KEY WORDS. *Non-arteritic anterior ischemic optic neuropathy, NAION, Thrombocytopenia, Aspirin*

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INTRODUCTION

Systemic hypertension, diabetes mellitus, hyperlipidemia, cardiovascular disease and a small and crowded disc are all risk factors for non-arteritic anterior ischemic optic neuropathy (NAION). As thrombocytes are believed to be involved in the pathogenesis, aspirin is currently recommended for the protection of the fellow eye.

We describe a patient who developed bilateral NAION in the presence of an extremely low platelet count.

Case report

A 68-year-old man presented with sudden painless loss of vision in his right eye. The history was remarkable for idiopathic autoimmunethrombocytopenia (ITP) diagnosed 18 years earlier. Splenectomy had been performed to improve the low platelet count, but had limited effect. Therefore the patient was placed on oral prednisolone, 5 mg/day.

On examination visual acuity was 20/400 OD and 20/25 OS. Both optic discs were prominent, with splinter hemorrhages (Fig. 1). The pupils reacted sluggishly to direct stimulation. In addition the OD had a relative afferent pupillary defect. No Ishihara plates were identified with the right eye but 16 out of 17 were identified correctly with the left eye.

Fluorescein angiography showed late staining of both optic discs (Fig. 2). The right visual field was concentrically constricted, with a superiotemporal field cut and an enlarged blind spot. The left visual field had an enlarged blind spot and moderate concentric constriction.

Laboratory tests showed a platelet count of 15,000. ESR was 56 mm/hour, C-reactive protein (CRP) was below 2. RBC and WBC were within the normal range as were all parameters for coagulopathies and collagen vascular disorders, such as aPTT, thrombin time, lupus anticoagulant, complement C4 and CH 100 alternate levels; the patient had negative rheumatoid factor, negative Anti-ds-DNA antibodies, normal titres for ANCA, anti-cardiolipin and anti-β-2-glyco-

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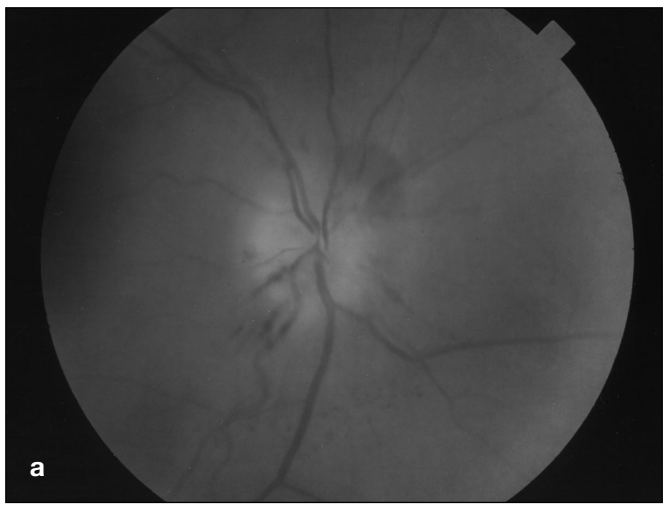


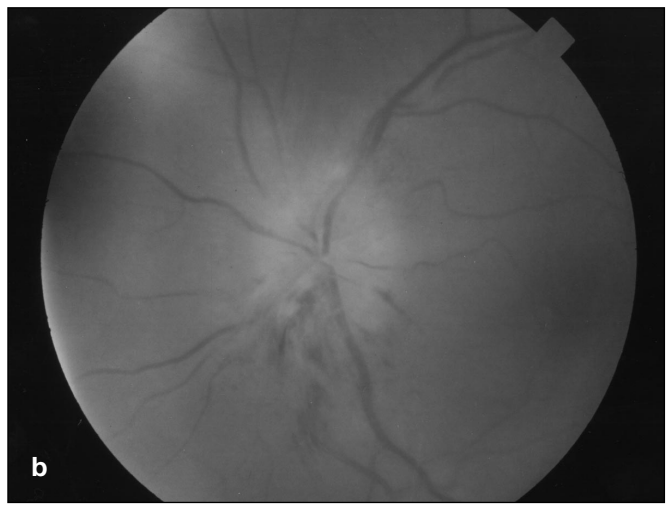
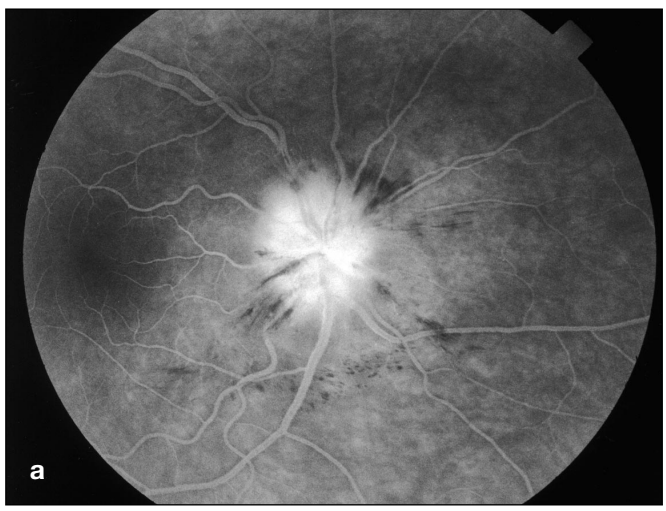


Fig. 1 - *Swelling of the optic disc with splinter hemorrhages in the inferotemporal and superonasal quadrant, right eye.*
1b *- Swelling of the optic disc with scattered hemorrhages predominantly in the inferotemporal and inferonasal quadrants, left eye.*



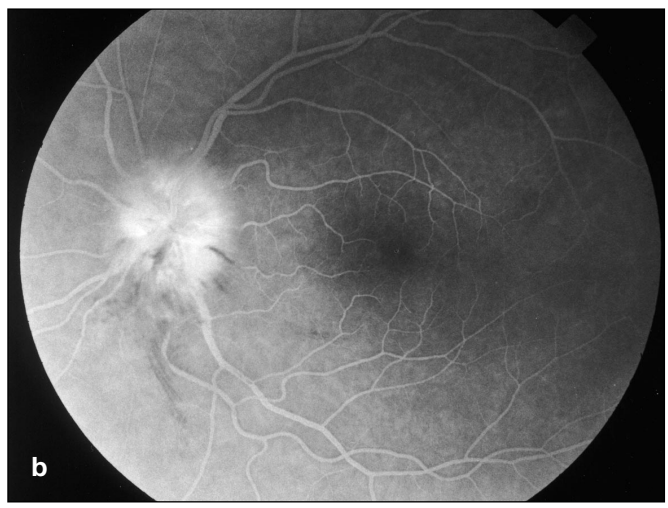


Fig. 2 - *Fluorescein angiography of* **a)** *the right and* **b)** *the left eye, with diffuse staining of the optic discs in the late phase due to ischemic edema. Besides splinter hemorrhages there were no signs of choroidal or retinal perfusion deficits, especially no tortuosities or other signs of venous thrombosis. The retinal arteries show some irregularities suggesting a low degree of arteriosclerosis. The perifoveal capillary net is intact, and there is no macular edema in the late phase of the angiogram.*

protein-1 antibodies, normal serum protein electrophoresis. Although there was no history or clinical indication for giant cell arteritis, temporal artery biopsy was performed and found to be normal. On the basis of the history, the clinical presentation and the laboratory results NAION was diagnosed by exclusion.

In order to raise the low platelet count so as to prevent spontaneous bleeding, methylprednisolone (4 x 250 mg/day IV for three days) was started. One week later visual acuity was 20/200 OD. The platelet count improved to 62,000. Corticosteroid treatment was reduced to 50 mg/ day p.o.

Two weeks after the visual loss in OD the patient started to lose vision in his left eye. Visual acuity was 20/150 OD and 20/200 OS. Laboratory tests indicated a platelet count of 51,000 and ESR 4-mm.

Four months after the first examination visual acuity had improved to 20/60 OD and 20/40 OS. The patient was able to read 4/15 Ishihara plates OD and 7/15 OS.

DISCUSSION

The etiology of NAION is not yet fully understood. A microvascular event in the optic nerve portion behind the lamina cribrosa is the postulated pathophysiological mechanism (1, 2). NAION usually presents in one eye, but less frequently the disease affects both eyes simultaneously (2). As no treatment is known to be effective, treatment strategies tend to focus on protection of the fellow eye. Although aspirin has no beneficial effect on the visual outcome in patients with NAION (4), it is currently recommended for the protection of the fellow eye (3, 5), since it inhibits platelet adhesion and aggregation.

Our patient was found to have ITP by exclusion of any other diseases including proliferative disorders or systemic vasculitis. The hematologists assumed the initally elevated sedimentation rate was due to the ITP. This was supported by the normal titers for CRP and the normal values for the markers of autoimmune disease. Despite his extremely low platelet count this patient developed bilateral NAION with severe visual loss.

Platelet function depends either on their number or their function. Recovery of vision is known to occur in patients with NAION. The remarkably good visual recovery in our patient might have been facilitated by the low platelet count and probably to some extent by the reduction of nerve fiber edema by the steroid medication.

Further studies are necessary to determine whether more potent platelet inhibitors, such as ticlopidine or dipyridamole as used in the prevention of strokes, are more effective in preventing and treating NAION in patients with normal platelet counts.

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