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Sudden onset blindness treated with pulse steroid therapy in a patient with microscopic polyangiitis

Case Report

Yoshiro Horai*, Tomoya Miyamura, Karin Shimada, Soichiro Takahama, Rumi Minami, Masahiro Yamamoto, Eiichi Suematsu

Department of Internal Medicine and Rheumatology, Clinical Research Center, National Hospital Organization, Kyushu Medical Center, 1-8-1 Jigyohama Chuo-ku, Fukuoka, 810-8563, Japan.

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Abstract: We report a 71-year-old male with microscopic polyangiitis (MPA) who developed sudden-onset, progressive, bilateral visual loss associated with a relapse of MPA symptoms. The patient was referred to our hospital, and treated with intravenous pulse steroid therapy and high-dose oral prednisolone. Although the right eye remained vision deficient, visual acuity in the left eye recovered. Ocular manifestations of MPA are quite uncommon. This case emphasizes the necessity of early detection and initiation of prompt therapy where ocular manifestations of MPA occur.

Keywords: *Acute blindness • Pulse steroid therapy • Microscopic polyangiitis*

1. Introduction

Microscopic polyangiitis (MPA) is an autoinflammatory disease characterized by systemic small vessel vasculitis. In MPA, myeloperoxidase anti-neutrophil cytoplasmic antibody (MPO-ANCA) is considered to play an important role in the pathogenesis of MPA [1]. However, the association between positivity of serum ANCA testing and the clinical features of MPA remains unclear.

MPA is a systemic disease, which affects many organ systems, including the lungs, kidneys gastrointestinal tracts, and peripheral nerves. However, cases of eye involvement have rarely been reported. In this study, we describe a case of MPA associated with sudden onset blindness, where MPO-ANCA levels were not elevated.

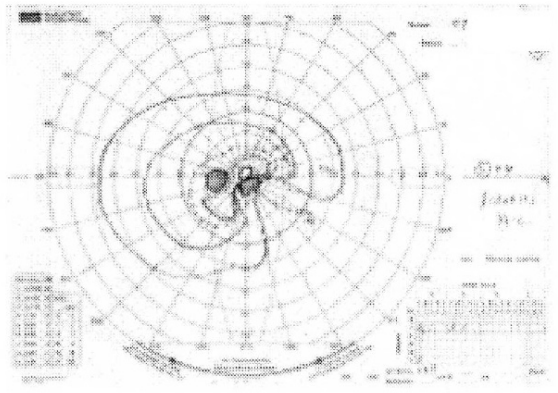
2. Case report

A 71-year-old male was admitted to a local hospital due to pyrexia, general malaise, and stiffness in his arms and legs in May 2005. He was diagnosed to have microscopic polyangiitis (MPA) because of renal dysfunction, mononeuritis multiplex, elevated serum CRP levels, and a positive finding for MPO-ANCA (441.0 EU/ml). Renal biopsy specimens revealed vasculitis and necrotizing glomerulonephritis, which were consistent with the renal pathology of MPA. Pulsed steroid therapy (500 mg/day of intravenous methylprednisolone for 3 consecutive days) was followed by 1 mg/kg/day of oral prednisolone (PSL). He suffered repeated remission and deterioration of his condition; therefore, the PSL dosage was titered according to disease activity. He suffered from an episode of herpes zoster in January 2010, which was treated with anti-herpetic agents.

Later in 2010, he developed a persistent headache and consulted a neurovascular surgery department at our hospital. Computed tomography (CT), magnetic resonance angiography, and single photon emission CT were performed. However, no abnormal findings were found. Tension-type headache was suspected and a trial of non-steroidal anti-inflammatory drugs was initiated. About 1 month after the development of headache, he suddenly lost right visual acuity and left visual acuity was severely deteriorated after a few days and he was admitted to our hospital immediately in mid-August 2010.

* *E-mail: yoshirohorai0518@qmed.hosp.go.jp*

Figure 1 Visual field test on admission.



A visual field defect in the lower temporal quadrant of the left eye was detected.

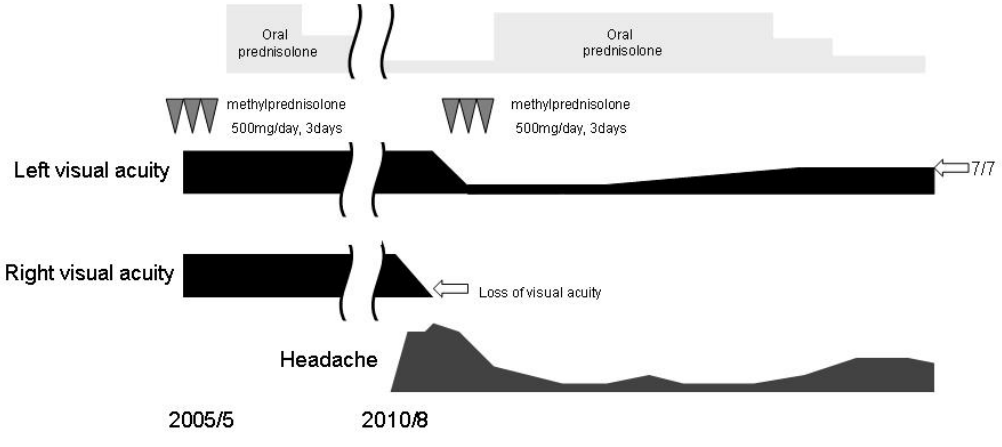
A physical examination on admission showed bilateral worsening of pretibial edema and sensory disturbance in the bilateral lower extremities. A headache located at the back of the head persisted, but neither swelling nor tenderness of temporal arteries was found. Post herpes zoster hyperpigmentation and neuralgia on the left side of the back were found. The results of laboratory tests on admission were: white blood cell count, 4,200/μl (neutrophils 81.3%, lymphocytes 14.2%, monocytes 4.5%); hemoglobin, 10.2 g/dl; platelet count, 170,000/μl; C-reactive protein, 2.27 mg/dl; erythrocyte sedimentation rate, 22 mm/hour; blood urea nitrogen, 22 mg/dl; serum creatinine, 1.3 mg/dl; Na^+, 139 mEq/L; K^+, 4.7 mEq/l; and Cl-, 107 mEq/l, prothrombin time, 12.0 seconds; international normalized ratio, 0.90; activated partial thromboplastin time, 23.0 seconds. Tests for protein and occult blood in urine were both negative. The microscopic examination of the urine sediment showed no blood cells or cellular casts. The results of tests to detect autoantibodies on admission were as follows: antinuclear antibody (ANA) was negative, anti-cardiolipin antibody IgG was negative (<8 U/ml), anti-β2-glycoprotein-I antibody was negative (<1.2 U/ml), lupus anticoagulant (dilute Russell's viper venom time) was negative (1.01), anti-double-stranded DNA antibody was negative. There was no positivity for MPO-ANCA (MPO-ANCA) or proteinase 3-ANCA. Herpes simplex virus and varicella zoster virus tests were done; however, no significant increases were observed. Visual field test revealed vision loss in right eye and the visual field defect in the lower temporal quadrant of the left eye (Figure 1). Fluorescein angiography showed no disturbance of the microcirculation or leakage from the optic papilla. No significant carotid artery stenosis was found on carotid ultrasound.

Based on the fact that sudden blindness occurred with increased general disease activity of MPA, and other causes of vision loss were not found, a diagnosis was made of MPA-associated sudden blindness. High-dose intravenous pulse steroid therapy was initiated (500 mg/day for 3 days) followed by 40 mg/day of PSL on a reducing dose regimen. Although his right visual acuity did not improve, vision in the left eye improved to 7/7. The dose of PSL was decreased to 25 mg/day without there being any relapse and he was eventually discharged from hospital (Figure 2).

3. Discussion

MPA is a systemic inflammatory disease characterized by small vessel vasculitis. Although the etiology and pathogenesis of MPA is still unknown, MPA, along with Wegener's granulomatosis (WG) and Churg-Strauss syndrome (CSS), is considered part of the family of ANCA-associated systemic vasculitides, based on the fact that the prevalence of MPO-ANCA in MPA is 70-

Figure 2 Clinical course.



80% [2]. Although it is reported that titers of MPO-ANCA can be useful for monitoring disease activity in patients with MPA [3], there are very few cases of MPA where titers of MPO-ANCA are not associated with systemic disease activity and emergence of new symptoms. In this case, MPO-ANCA remained negative despite visual loss occurring. However, exacerbations of mononeuritis multiplex, renal dysfunction and slightly elevated CRP levels, which suggested increased MPA disease activity, were observed. One possibility is that MPO-ANCA tests were pseudo-negative due to a limitation of the direct enzyme-linked immunosorbent assay (ELISA). Tamura et al. reported cases of refractory WG treated with rituximab. In these cases, capture MPO-ANCA ELISA tests were positive, despite direct MPO-ANCA ELISA tests being negative when relapses of WG were observed [4]. Their report suggested that relapse of ANCA-associated systemic vasculitis is possible even where direct MPO-ANCA ELISA tests are negative, and capture ELISA methods might be useful.

While ocular manifestations of WG, such as scleritis, episcleritis, conjunctivitis and orbital mass, are well recognized, ocular involvement in other ANCA-associated systemic vasculitides, such as MPA and CSS, are rarely reported. At the referral to our department, we suspected temporal arteritis (TA) because headache and visual disturbance were found. However the possibility of TA was denied because swelling or tenderness of temporal arteries was not found and abnormality of the temporal arteries by magnetic resonance angiography was not apparent. We first Guillevin et al. analyzed the clinical symptoms, laboratory findings, and outcomes in 85 patients with MPA; only in one patient was eye involvement found (2). Most reports of acute loss of vision in ANCA-associated vasculitis were reported as being complications of ANCA-associated systemic vasculitis, without there being a specific diagnosis of either WG, CSS, or MPA. Therefore the actual frequency of eye complications in MPA is presently unknown. We found two reports describing cases of acute vision loss in patients diagnosed with MPA. Duran et al. reported a case of a 67-year old male with ANCA-associated vasculitis presenting with acute vision loss due to ischemic optic neuropathy [5]. In the report, the authors diagnosed him as having likely WG or MPA. However, the diagnosis of MPA was unlikely because the patient lacked symptoms of the ears, orbits, nose, sinuses, and lungs. The other case, an 80-year-old female with MPA who presented with subacute optic neuropathy was reported by Altaie R [6]. Both of these cases were treated with high-dose PSL and visual symptoms improved. This case and the two cases mentioned above have several common features: older age, renal manifestations ascertained by renal biopsy, and improvements of ocular manifestations following PSL therapy. In the present case, we found the clinical features of ischaemic optic neuropathy. However, clinical course representing acute progressive vision loss was not incompatible with optic neuropathy associated with systemic small-vessel vasculitis. Therefore we suspected the possibility of arteritic optic neuropathy and started high-dose PSL, which resulted in a satisfactory response.

In this study, we report a patient with sudden blindness associated with MPA. Although eye manifestations in patients with MPA are rare, symptoms can become severe. Prompt initiation of immunosuppressive therapy should be considered for patients with MPA complicating vision impairment.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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