

A case of antineutrophil cytoplasmic antibody associated vasculitis with presenting optic neuritis

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Introduction: Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) affects systemic small vessels and is characterized by the presence of ANCA in the serum. The clinical manifestations of AAV depend on the affected vascular bed. Optic neuritis is not a common feature of AAV, but several case reports have documented its occurrence. Here, we present a case of a 53-year-old man with AAV who presented with optic neuritis as the initial clinical symptom.

Case: A 53-year-old man presented with a temporary visual impairment in his right eye that had occurred three weeks ago. Examination revealed relative afferent pupillary defect and papilledema in the right eye. One week later, he was also diagnosed with rheumatoid arthritis due to hand stiffness, arthralgia, and claudication. He was prescribed methylprednisolone (8mg/day) and hydroxychloroquine (200mg/day), but he was referred to our hospital due to the persistent symptoms. Laboratory findings revealed an elevated erythrocyte sedimentation rate (118mm/hour) and high-sensitivity C-reactive protein (17.26mg/dL). Further tests showed positive results for c-ANCA (84.3AU/ml) and p-ANCA (>100AU/ml). Urinalysis showed microscopic hematuria and mild proteinuria, so a renal biopsy was not performed. Orbital MRI showed diffuse enhancement of the intraorbital segment of both optic nerves, suggesting optic neuritis (Figure 1). Femoral MRI revealed diffuse atrophic changes in the right thigh, consistent with myositis. Nerve conduction studies showed left median motor neuropathy. He underwent early steroid pulse therapy for three days, followed by rituximab induction therapy for up to the third cycle. However, during the fourth cycle, he developed a skin rash with pruritus all over the body, which was thought to be caused by the drug. So rituximab was discontinued, and he is currently being treated with mycophenolate mofetil. The visual disturbance in the right eye is gradually improving.

Discussion: Ocular involvement is extremely rare and can easily be overlooked during the evaluation of AAV. However, it is important to actively evaluate and manage it due to its potential for rapid progression and vision loss.

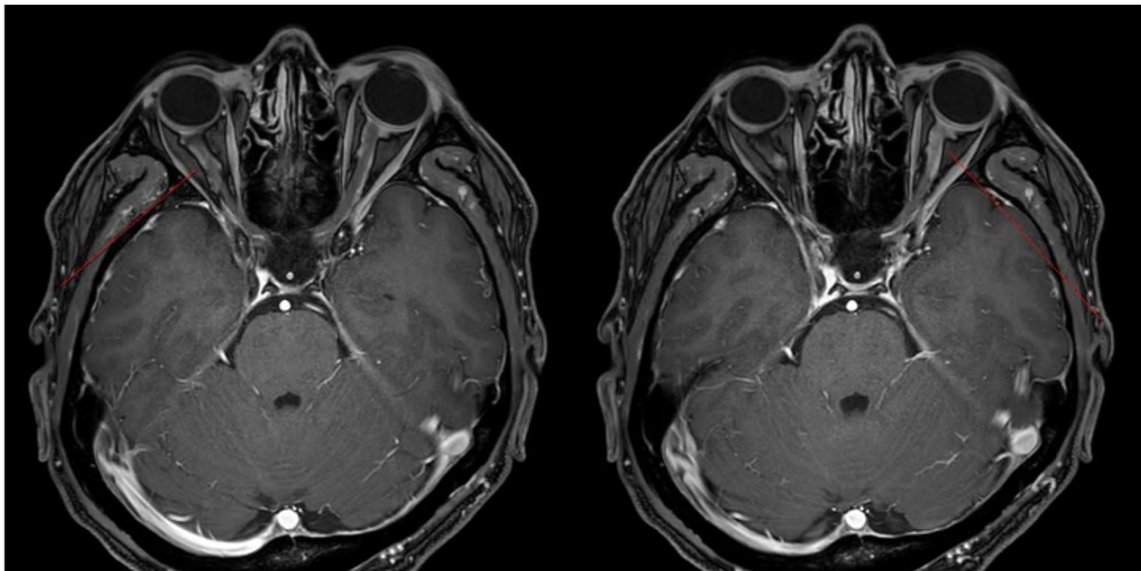


Figure 1. The orbit MRI showed diffuse enhancement of the intra-orbital segment of both optic nerves.