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Optic Neuritis as an Atypical Initial Symptom of Microscopic Polyangiitis: A Case Report

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Case Study : Microscopic polyangiitis (MPA) is a systemic vasculitis that mainly affects the kidneys, lungs, skin and joints, causing inflammation and damage. However, optic neuritis as an initial MPA symptom is extremely rare. We describe the case of a patient whose initial manifestation of MPA was visual impairment, highlighting the intricate and systemic nature of this condition. A 53-year-old male developed right eye visual impairment three weeks before visiting our clinic. Two weeks ago, he was diagnosed with rheumatoid arthritis at a private clinic due to joint pain and was prescribed methylprednisolone. Despite treatment, persistent symptoms such as fever, muscle aches and walking difficulties due to weakness required a referral to our hospital. Laboratory tests showed high C-reactive protein (17.26 mg/dL), elevated c-ANCA (84.3 AU/mL), p-ANCA (>100 AU/mL), and increased muscle enzymes. Urinalysis revealed microscopic hematuria and mild proteinuria, with a spot urine protein/creatinine ratio of 0.7 mg/mg. MRI scans showed diffuse enhancement of the intra-orbital segments of both optic nerves, indicative of optic neuritis, and atrophy in the right thigh muscles, consistent with myositis. The patient underwent steroid pulse therapy (methylprednisolone 500mg/day) and three cycles of rituximab (once-weekly doses of 375 mg/m2). Due to a rash attributed to rituximab, the treatment was switched to mycophenolate mofetil. About a month later, serum creatinine rose from 1.3 to 4.3 mg/dL, with a urine protein/creatinine ratio of 5.4 mg/mg. A renal biopsy showed cellular crescents without immune complex deposition, aligning with ANCA-associated pauci-immune glomerulonephritis. Treatment began with steroid pulse therapy and cyclophosphamide (750 mg biweekly for three cycles), followed by consolidation therapy (500 mg every three weeks for three cycles). The patient's renal function and right eye vision both improved over time. This case highlights the need for vigilant monitoring in patients with AAV presenting unusual symptoms such as optic neuritis.