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RESEARCH ARTICLE

# MPO-ANCA-ASSOCIATED VASCULITIS PRESENTING WITH PITUITARY MASS: EXPANDING THE CLINICAL SPECTRUM

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Abstract: *Background:* Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a small-vessel necrotizing vasculitis that predominantly affects upper respiratory tract ,kidneys, lungs, and peripheral nerves. Pituitary involvement is exceedingly rare, more commonly associated with granulomatosis with polyangiitis (GPA) and proteinase 3 (PR3)-ANCA positivity. MPO-ANCA-positive vasculitis with pituitary mass formation is exceptionally uncommon. **Case Presentation**:We report a 63-year-old female with type 2 diabetes mellitus who presented with dyspnea, bilateral pedal edema, and neuropathic symptoms. Evaluation revealed heart failure with reduced ejection fraction (EF 30%), mononeuritis multiplex, and renal involvement. Serology was positive for MPO-ANCA, confirming AAV. Subsequently, she developed headache and bitemporal visual field defects. Brain MRI revealed a pituitary mass with suprasellar extension compressing the optic chiasma. Hormonal assays were normal. Immunosuppressive therapy led to significant regression of the lesion and systemic improvement. ***Conclusion***:This case highlights the rare occurrence of pituitary involvement in MPO-ANCA vasculitis. Recognition of this manifestation is essential as radiological regression following immunosuppression distinguishes inflammatory infiltration from neoplasia.

Keywords:

## INTRODUCTION

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) represents a group of autoimmune small-vessel vasculitides, including microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA), and eosinophilic granulomatosis with polyangiitis (EGPA). Among these, MPA is primarily characterized by myeloperoxidase (MPO)-ANCA positivity and affects kidneys, lungs, and peripheral nerves. Pituitary gland involvement is exceptionally rare, with a reported incidence of 1–3% among AAV cases, typically seen in GPA rather than MPA (1,5).

Pituitary involvement may manifest as diabetes insipidus, hypopituitarism, or mass effects due to inflammatory infiltration. In most cases, radiological findings mimic pituitary adenomas, posing a diagnostic challenge (3). Here, we report a rare case of MPO-ANCA-associated vasculitis presenting as a pituitary mass with suprasellar extension, which regressed following immunosuppressive therapy, thus expanding the clinical spectrum of this disease entity.
Myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA)-associated vasculitis (AAV) is a small-vessel necrotizing vasculitis that most commonly affects the kidneys, lungs, peripheral nerves, and skin. However, central nervous system (CNS) involvement is rare, and pituitary involvement is even more uncommon, often posing a diagnostic challenge. We report a 63-year-old female with type 2 diabetes mellitus who presented with dyspnea, bilateral pedal edema, and neuropathic symptoms. Evaluation revealed heart failure with reduced ejection fraction (30%), mononeuritis multiplex, and renal dysfunction. Serological testing confirmed the presence of MPO-ANCA, consistent with microscopic polyangiitis (MPA). While on immunosuppressive therapy, the patient developed new-onset headache, blurring of vision, and bitemporal visual field defects. Magnetic resonance imaging (MRI) of the brain revealed a pituitary mass with suprasellar extension compressing the optic chiasma. Pituitary hormonal profile was normal, excluding a functioning adenoma. In view of concurrent systemic vasculitis and absence of hormonal abnormality, a diagnosis of pituitary involvement secondary to AAV was made. Immunosuppressive therapy with corticosteroids and cyclophosphamide resulted in remarkable regression of the lesion and resolution of visual symptoms.

Pituitary involvement in AAV is exceedingly rare, with limited cases documented in the literature, mostly

associated with granulomatosis with polyangiitis (GPA) and rarely with MPO-ANCA-positive MPA [1,2]. The pathogenesis is attributed to granulomatous inflammation or vasculitic necrosis within the pituitary vasculature, leading to mass effect or hormonal dysfunction [3]. Clinical manifestations may include headache, visual disturbances, diabetes insipidus, or anterior pituitary insufficiency. Radiologically, pituitary lesions may mimic adenomas or hypophysitis, often leading to misdiagnosis. The absence of endocrine dysfunction in this case further complicated the diagnostic process. Early diagnosis through MRI and ANCA testing is crucial, as timely initiation of immunosuppressive therapy can lead to complete or near-complete resolution of pituitary lesions without surgical intervention [4,5].

This case underscores the need for clinicians to consider pituitary involvement as a rare but reversible manifestation of MPO-ANCA-associated vasculitis, especially in patients presenting with new neurological or visual symptoms during systemic disease activity.

**Case Presentation**

A 63-year-old woman, a known case of type 2 diabetes mellitus for 10 years, presented with progressive exertional dyspnea, bilateral pedal edema, and tingling sensations in both lower limbs over a period of two months. The symptoms gradually worsened, limiting her daily activities. There was no history of fever, chest pain, or orthopnea. On clinical examination, she was afebrile with mild pedal edema, bibasal crepitations on lung auscultation, and reduced ankle reflexes suggestive of peripheral neuropathy. Her blood pressure was mildly elevated, and cardiovascular examination revealed a displaced apex beat. Laboratory investigations showed a hemoglobin level of 10.8 g/dL, serum creatinine of 1.4 mg/dL, and 24-hour urinary protein excretion of 810 mg with microscopic hematuria. Echocardiography demonstrated global left ventricular hypokinesia with an ejection fraction of 30%, consistent with heart failure with reduced ejection fraction (HFrEF).

Given the combination of cardiac, renal, and neurological manifestations, a systemic vasculitic process was suspected. Nerve conduction studies revealed severe axonal sensory-motor polyneuropathy, consistent with mononeuritis multiplex. Further immunological workup demonstrated positive myeloperoxidase-antineutrophil cytoplasmic antibodies (MPO-ANCA 234 U/ml) with negative proteinase-3 ANCA, establishing a diagnosis of microscopic polyangiitis (MPA). Chest computed tomography (CT) revealed bilateral patchy ground-glass opacities and mediastinal lymphadenopathy, supporting pulmonary vasculitic involvement (Fig.1). Collectively, these findings confirmed multisystem involvement due to MPO-ANCA-associated vasculitis.

During hospitalization, the patient developed new-onset, progressively worsening headaches associated with blurred vision and bitemporal visual field defects. Fundoscopic examination was normal, and no cranial nerve palsies were observed. Magnetic resonance imaging (MRI) of the brain showed a 20 × 20 × 21 mm sellar mass with suprasellar extension compressing the optic chiasma and medial frontal lobes. The lesion appeared heterogeneously hyperintense on T2-weighted images with foci of hemorrhage and lacked the posterior pituitary bright spot (Fig 2.). Pituitary hormonal assays, including TSH, ACTH, prolactin, LH, FSH, and cortisol, were within normal limits, excluding hypopituitarism or functional adenoma. In the context of active systemic vasculitis, pituitary involvement secondary to vasculitic infiltration was considered the most plausible diagnosis.

The patient was started on intravenous methylprednisolone pulses (1 g/day for three consecutive days), followed by oral prednisolone at 1 mg/kg/day. In addition, she received intravenous cyclophosphamide (15 mg/kg every two weeks) as induction therapy. Despite partial improvement, persistent neurological deficits prompted administration of rituximab (1 g intravenously, two doses given two weeks apart). She tolerated immunosuppressive therapy well, with close glycemic monitoring and prophylaxis against opportunistic infections.

Over the ensuing three months, the patient's cardiac function improved significantly, with left ventricular ejection fraction increasing to 53%. Neuropathic symptoms stabilized, renal parameters normalized, and there was no recurrence of pulmonary manifestations. Follow-up MRI at three months demonstrated substantial mild reduction in the size of the pituitary lesion with resolution of hemorrhagic components (Fig 3.), confirming its inflammatory and reversible nature. Her visual symptoms showed partial recovery, with improvement in visual acuity and field defects. The patient remained in clinical remission with improvement in BVAS (from 34 to 7), underscoring that prompt recognition and aggressive immunosuppressive therapy in MPO-ANCA-associated vasculitis can result in both systemic and neuroendocrine recovery, even in rare presentations such as pituitary involvement.

# RESULTS AND OBSERVATIONS:

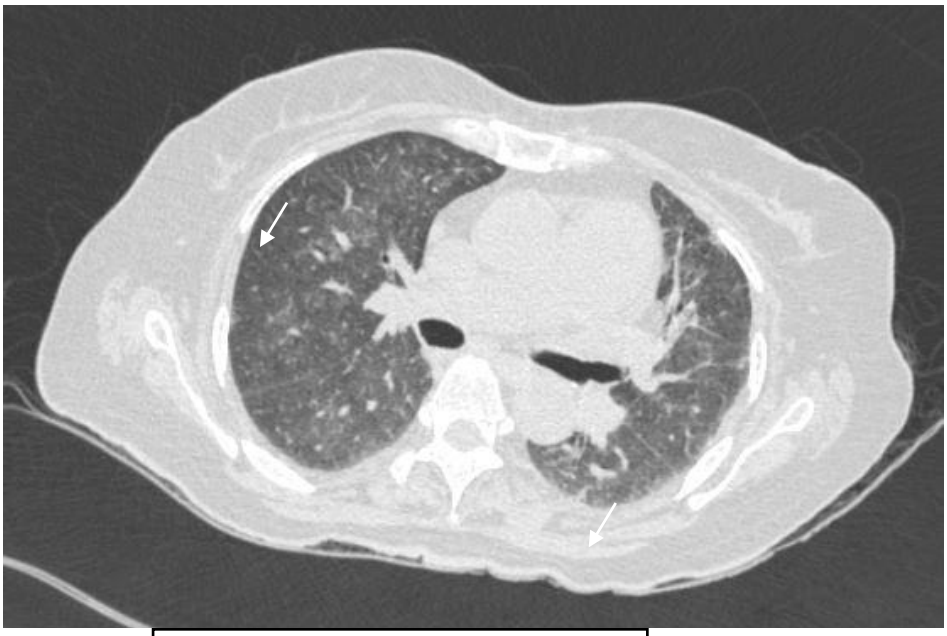

Fig 1. CT thorax

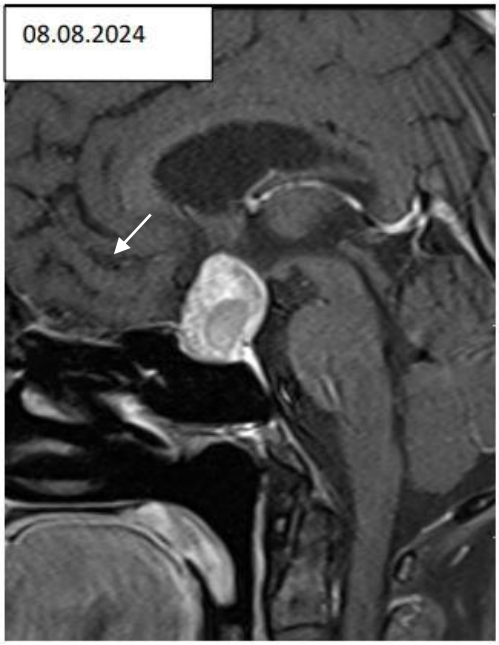


Fig 2. Pituitary mass

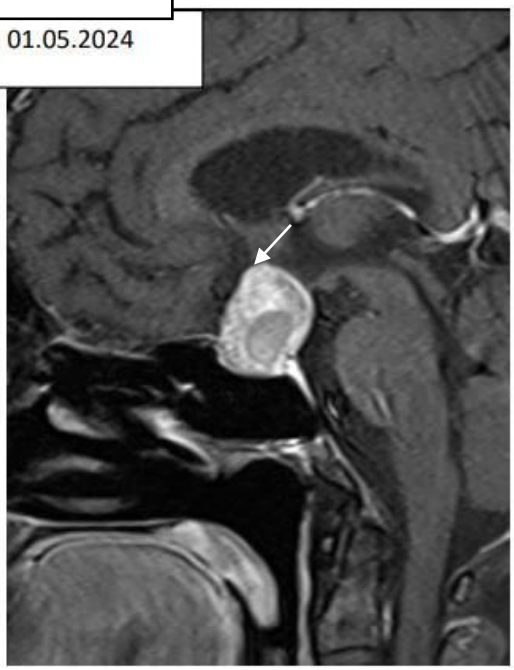


Fig 3. Pituitary mass

# DISCUSSION

Pituitary involvement in antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a rare but increasingly recognized manifestation, largely owing to the advancement of neuroimaging and heightened clinical awareness. The reported prevalence of pituitary lesions in AAV ranges from 1–3%, with the majority of cases occurring in granulomatosis with polyangiitis (GPA) and proteinase 3 (PR3)-ANCA-positive patients [6,8,10]. In contrast, microscopic polyangiitis (MPA) associated with myeloperoxidase (MPO)-ANCA positivity and pituitary involvement is exceedingly uncommon, with only isolated case reports and small series described in the literature.

The underlying pathophysiological mechanisms involve either granulomatous inflammation or necrotizing vasculitis affecting the hypophyseal vasculature, leading to ischemia, necrosis, or fibrosis of the gland

and adjacent structures [7]. The inflammatory process can extend into the infundibulum, optic chiasma, or hypothalamus, resulting in a spectrum of clinical manifestations. Patients may present with headache, visual field defects due to chiasmal compression, cranial neuropathies, or features of anterior and/or posterior pituitary dysfunction. Diabetes insipidus is reported as the most frequent endocrine abnormality, often due to involvement of the posterior pituitary or infundibulum. However, as demonstrated in our case, the absence of hormonal derangements does not preclude pituitary vasculitis, emphasizing that radiological and systemic correlation is essential for diagnosis [6,9].

Magnetic resonance imaging (MRI) remains the diagnostic modality of choice in evaluating pituitary involvement. Vasculitic or inflammatory pituitary lesions can closely mimic pituitary macroadenomas in imaging appearance. Typical features suggesting an inflammatory rather than neoplastic etiology include heterogeneous enhancement, thickening of the pituitary stalk, absence of the posterior pituitary bright spot, and suprasellar extension compressing the optic chiasma [7,8]. A hallmark feature supporting an inflammatory origin is the regression of the lesion following immunosuppressive therapy, as observed in this patient. Such radiological improvement, coupled with systemic remission of vasculitic features, is a strong indicator of vasculitic infiltration rather than primary pituitary neoplasia.

The treatment approach for pituitary involvement in AAV parallels that of systemic disease. High-dose corticosteroids remain the cornerstone of therapy, often combined with cytotoxic agents such as cyclophosphamide or biological agents like rituximab for induction of remission [9]. Rituximab, a monoclonal antibody targeting CD20-positive B cells, has shown favorable outcomes in refractory or relapsing cases and may be particularly useful when cyclophosphamide toxicity is a concern. In most reported cases, timely initiation of immunosuppressive therapy has resulted in substantial or complete resolution of pituitary lesions and recovery of endocrine and visual functions. Delay in treatment, however, may lead to irreversible pituitary insufficiency and persistent visual deficits [10].

This case adds to the limited literature describing MPO-ANCA-associated vasculitis presenting with a pituitary mass, underscoring the importance of recognizing such atypical central nervous system (CNS) manifestations. Awareness of this rare presentation can help prevent misdiagnosis as pituitary adenoma and avoid unnecessary surgical intervention. Early recognition and aggressive immunosuppressive therapy can lead to dramatic systemic and radiological improvement, highlighting the reversibility of inflammatory pituitary lesions in AAV when treated appropriately.

# CONCLUSION

Pituitary involvement in MPO-ANCA-associated vasculitis is an uncommon yet significant manifestation that can clinically and radiologically mimic pituitary adenoma. The absence of hormonal dysfunction should not exclude the diagnosis, particularly when accompanied by systemic vasculitic features such as renal, neurological, or pulmonary involvement. Magnetic resonance imaging (MRI) regression following corticosteroid or immunosuppressive therapy strongly supports an inflammatory origin rather than neoplastic pathology. Early recognition and initiation of treatment are essential to prevent permanent endocrine or visual deficits. Multidisciplinary collaboration among rheumatology, endocrinology, and neurology teams ensures comprehensive care, facilitates accurate diagnosis, and optimizes patient outcomes through timely immunomodulatory intervention.

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