

Clinical Image

Digital infarct and mononeuritis in a middle aged female: always suspect Antineutrophil cytoplasmic antibodies associated vasculitis

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ABSTRACT

Vasculitis is a process caused by inflammation of blood vessel walls and results in a variety of disorders. Small-vessel vasculitis (vasculitis involving arteries, venules and capillaries) should be suspected in any patient who presents with a multisystem disease that is not caused by an infectious or malignant process. Testing for Antineutrophil cytoplasmic antibody (ANCA) is the basis of classification of small vessel vasculitis into ANCA associated and non - ANCA associated vasculitis. Apart from cutaneous manifestations like palpable purpura and vasculitic urticaria, digital gangrene in a patient with evidence of mononeuritis multiplex is highly suggestive of ANCA associated vasculitis (AAV). Clinically most of these vasculitides have overlapping clinical presentations and similar treatment. Early diagnosis and rapid initiation of treatment of AAV is recommended rather than ordering for definitive tests (e.g. histopathology or angiograms) since delay in treatment can result in serious end organ damage (pulmonary or renal).

Keywords: Antineutrophil cytoplasmic antibody vasculitis, Digital gangrene, Digital infarct, Mononeuritis multiplex

INTRODUCTION

A 38 year old house maker, mother of 2 children from Indian state of Uttar Pradesh presented with a 15 day history of progressive weakness of all four limbs, skin rashes and polyarthralgia. She was conscious, anxious, afebrile with normal hemodynamic parameters. Skin examination showed multiple, small, non-tender, purpuric lesions (largest measuring 1cm X 0.7cm) over both hands and pretibial regions.

Neurological examination revealed flaccid weakness and absent reflexes (LMN type) in both upper limbs (left > right) and left lower limb with sensory loss limited to ulnar nerve distribution of left hand and common peroneal nerve distribution of left leg. Rest of the

systemic examinations were insignificant. Routine blood investigations revealed anemia and thrombocytosis. An urgent Nerve conduction study revealed Mononeuritis multiplex. Anti-nuclear antibody testing (2+, homogenous), Anti-MPO (>100IU/ml) and proteinuria (spot urine- 200mg) confirmed the diagnosis as ANCA associated vasculitis (AAV). The patient eventually developed excruciating finger pain, ecchymotic patches, necrotic ulceration, digital gangrene (left little finger), blackening of other fingers and ulnar clawing. She was put on oral corticosteroid (Prednisolone- 1mg/kg/day) which temporarily resolved the pain, but gangrene progressed to involve all fingers of left hand. Authors have planned for pulse cyclophosphamide and skin biopsy, but attendants insisted on transferring the patient to higher centre.



Figure 1: Digital gangrene and mononeuritis.

DISCUSSION

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a group of disorders characterized by inflammation and destruction of small- and medium-sized blood vessels and the presence of circulating ANCA.¹ ANCA-associated small-vessel vasculitis includes microscopic polyangiitis, Wegener's granulomatosis,

Churg-Strauss syndrome, and drug-induced vasculitis.² The rapid diagnosis of ANCA associated vasculitis is crucial in preventing end organ damage and fatal outcome.³ Treatment consisting of corticosteroids and immunosuppressive should be started early when indicated without making unnecessary delay for a definitive diagnosis.

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REFERENCES

1. Geetha D, Jefferson JA. ANCA-Associated Vasculitis: Core Curriculum 2020. Am J Kidney Dis. 2019 Jul 26.
2. Jennette JC, Falk RJ. Small-vessel vasculitis. N Engl J Med. 1997;337:1512-23.
3. Mansi IA, Opran A, Rosner F. ANCA-associated small-vessel vasculitis. Am Family Physician. 2002 Apr 15;65(8):1615.

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