

A RARE CASE OF MICROSCOPIC POLYANGIITIS**Harshita Ganji*¹, Raghavendra Prasad² and Prabhakar Kamarthy³**^{1,2,3} MBBS, MD GENERAL MEDICINE,

Sri Devaraj URS Academy of Higher Education And Research, Kolar, Karnataka, India.

Corresponding Author: Dr. Harshita Ganji

MBBS, MD GENERAL MEDICINE, Sri Devaraj URS Academy Of Higher Education And Research, Kolar, Karnataka, India

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ABSTRACT

INTRODUCTION: Microscopic polyangiitis (MPA) is vasculitis of small vessels. The incidence of MPA is approximately 1:100.000 per year. A dermato-pulmonary-renal syndrome is the feature of the disease. Lung hemorrhage is observed in 12 to 29% of the patients with MPA and is an important contributory factor to morbidity and mortality. **CASE REPORT:** A 32 year old male patient presented with history of decreased urine output since 4 days, generalised weakness since 1 week, blackish discoloration of tips of the fingers and toes. On examination, Gangrene is present over distal aspect of fingers and toes. Patient was started on hemodialysis in view of deranged blood urea and serum creatinine. Patient developed dyspnoea on 4th day. Chest ray was done and it showed bilateral pulmonary infiltrations. Perinuclear Anti-Neutrophil Cytoplasmic Antibodies (p-ANCA) specific to myeloperoxidase shows positive. Patient was started on methylprednisolone 125mg/kg/day. Patient had sudden onset of chest pain followed by cardiac arrest and death. **CONCLUSION:** A Young male presenting with above clinical scenario, vasculitis should be suspected. Usually it occurs in 50-70 years males, but occurring in a 32 yr old male is a rare presentation.

KEYWORDS: Microscopic polyangiitis, Perinuclear Anti-Neutrophil Cytoplasmic Antibodies, methylprednisolone, vasculitis

INTRODUCTION

Microscopic polyangiitis (MPA) is vasculitis of small vessels. It was initially considered as a microscopic form of polyarteritis nodosa (PAN). The incidence of MPA is approximately 1:100.000 per year. Renal failure and pulmonary involvement are the major causes of morbidity and mortality in MPA¹. A dermato-pulmonary-renal syndrome is the feature of the disease. Lung hemorrhage is observed in 12 to 29% of the patients with MPA and is an important contributory factor to morbidity and mortality. Some patients with small-vessel lung vasculitis may present clinical, radiologic and functional findings consistent with an interstitial process mimicking idiopathic pulmonary fibrosis. Musculoskeletal involvement (myalgias, arthralgias and arthritis) are present in 65 to 72% of the patients. Cutaneous lesions (purpura, splinter hemorrhages) are found in 44 to 58% of the patients. Gastrointestinal symptoms are characterized by abdominal pain (32 to 58%) and digestive tract bleeding (29%). Peripheral neuropathy is found in only 14 to 36% of the cases, thus occurring less frequently than in PAN.¹ We report a 32 year old male with renal failure, pulmonary involvement and distal gangrene.

CASE REPORT

A 32 year old male patient presented with history of

- Decreased urine output since 4 days.
- Generalised weakness since 1 week.
- Blackish discoloration of tips of the fingers and toes.

Patient is not a known case of hypertension and diabetes. On examination, Gangrene is present over distal aspect of fingers and toes (FIGURES 1 AND 2). Patient was started on hemodialysis in view of deranged blood urea and serum creatinine. 4 cycles of hemodialysis was done. Patient developed dyspnoea on 4th day. Chest ray was done and it showed bilateral pulmonary infiltrations (FIGURE 3). CT thorax showed bilateral peribronchovascular nodular infiltration (FIGURE 4). p-ANCA specific to myeloperoxidase shows positive. Renal biopsy was done and it showed focal segmental crescentic glomerulonephritis without immune-complex deposition. Patient was started on methylprednisolone 125mg/kg/day. Patient had sudden onset of chest pain followed by cardiac arrest and death.

INVESTIGATIONS:

Blood urea-127 mg/dl.

Serum creatinine- 6.0 mg/dl.

Serum sodium-132 meq/l.

Serum potassium- 4.4 meq/l.

Random blood sugar-
90mg/dl.

Hemoglobin-8.0 gm%.

Platelet count- 4.52 lakh/
cu.mm

Total leucocyte count- 11,000
cells/cu.mm.

Urine albumin- present

P-ANCA – POSITIVE

Renal biopsy : Focal
segmental crescentic
glomerulonephritis

Chest xray- Bilateral
pulmonary infiltrations.

CT thorax- Bilateral
peribronchovascular Nodular
infiltration



FIGURE 1



FIGURE 2

**FIGURE 1 AND 2 : SHOWS DISTAL
GANGRENE OF FINGERS AND TOES**

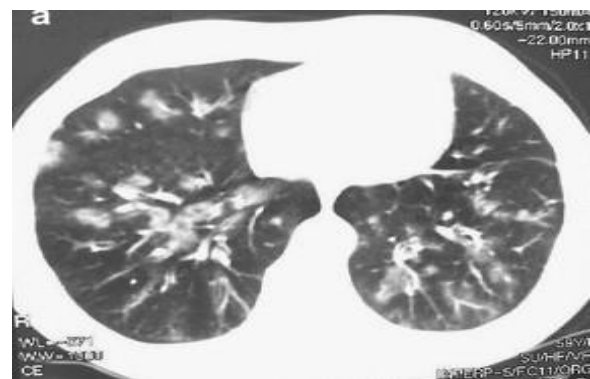


FIGURE 3



FIGURE 4

**FIGURE 3: CT THORAX SHOWING
BILATERAL PERIBRONCHOVASCULAR
NODULAR INFILTRATION**

**FIGURE 4 : CHEST XRAY SHOWING
BILATERAL PULMONARY INFILTRATIONS**

DISCUSSION

MPA is one of the vasculitides that is included in the pulmonary renal syndromes. The incidence of MPA is

approximately 1:100,000 per year. Males are more commonly affected than females. RPGN is one of its major characteristics. Other clinical symptoms such as alveolar haemorrhage, cutaneous, musculoskeletal, gastrointestinal system, neurological and ear-nose-throat involvement may also be found but occur less frequently.^[1]

- Lauque et al. showed that the median time between onset of symptom and diagnosis of MPA was highly variable between 2 weeks and 10 years.^[1]
- ANCA antibodies are useful diagnostic markers for MPA. Circulating ANCA antibodies are present in 74.5% of patients with MPA. The p-ANCA pattern with antibodies to MPO is most closely associated with MPA. Lauque et al. revealed that p-ANCA antibodies were present in 93% of MPA patient. In this case, p-ANCA with anti-MPO activity was positive.^[1]
- Ando et al. reviewed the CT thorax findings in 51 MPA patients with pulmonary involvement. The CT findings consisted of ground-glass attenuation in 94% of patients, consolidation in 78%, thickening of bronchovascular bundles in 51%, honeycombing in 37%, nodules larger than 1 cm in 29%, bronchiectasis in 27%, pleural effusion in 27% and enlarged mediastinal lymph nodes in 18%.^[2]
- Agard et al found that the most common presenting clinical symptoms attributable to vasculitis in 36 MPA patients were general symptoms (fever, weight loss, asthenia) (61%), followed by myalgias (43%) and arthralgias (29%). Alveolar hemorrhage occurred in 11% of the patients.^[3]
- The severity of pulmonary and renal involvement often requires intensive treatment with high doses of corticosteroids and pulse cyclophosphamide and if needed hemodialysis. Renal function may improve markedly with early treatment in most patient even in those with initial severe renal failure. Relapses are frequent in MPA, occurring in 25–54% of patient who survive the first months.^[4]

CONCLUSION

A Young male presenting with above clinical scenario, vasculitis should be suspected. Usually it occurs in 50-70 years males, but occurring in a 32 yr old male is a rare presentation.

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