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Case Report

Digital gangrene a rare early manifestation of Wegener's granulomatosis: a case report

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ABSTRACT

Wegener's granulomatosis (WG) or Granulomatosis with polyangiitis (GPA) rarely presents with digital gangrene being an early manifestation. We report a case of 60-year-old woman who presented with fever and cough with occasional hemoptysis along with painful bilateral feet digital gangrene. X ray chest and HRCT thorax of the patient was suggestive of multiple nodulo-cavitatory lesions. HPE of the lung lesion showed granulomatous pathology. CT Angiography was suggestive of multiple calcified and non-calcified lesions in infra renal aorta with stenosis of right dorsalis pedis artery. Biopsy of the skin lesion revealed a leucocytoclastic vasculitis. We eventually diagnosed GPA in light of clinical symptomatology with HPE of lung mass showing granulomatous lesion along with a positive cANCA. A few months after starting combined cyclophosphamide and steroids patient had significant symptomatic improvement. Hence Wegener's granulomatosis should always be considered as a possibility while evaluating a patient of digital gangrene specially with pulmonary and renal involvement.

Keywords: Wegener's granulomatosis, Granulomatosis with polyangiitis, Digital gangrene

INTRODUCTION

Granulomatosis with polyangiitis (GPA) or Wegener's granulomatosis (WG) is a systemic disorder that involves both granulomatosis and polyangiitis. The condition was originally named by Friedrich Wegener, who described the disease in 1936.[1] It is a form of multisystem vasculitis (inflammation of blood vessels) that affects small- and medium-size vessels in many organs. The inflammatory process associated may involve various territories conditioning multiple clinical presentations. Digital ischemia due to Wegener's granulomatosis is extremely rare.[9] In known literature, there are less than 10 reported cases of Wegener's granulomatosis presenting as digital gangrene. Hence this is a unique case report of Wegener's granulomatosis presenting as digital gangrene which is a rare early manifestation.

CASE REPORT

A 60 year female presented with fever and cough with occasional hemoptysis since 1 month and pain bilateral feet with bluish black discoloration since 8 days with similar past history of fever with cough 1 year back diagnosed as sputum smear negative pulmonary TB on the basis of radiographic evidence (X ray changes), she took ATT for 6 months but did not respond satisfactorily. No significant past, family or personal history was elicited. Examination revealed gangrene of bilateral toe, on palpation feeble pulsation of bilateral posterior tibial artery and left dorsalis pedis artery and absent pulsation in right dorsalis pedis artery, auscultation of the chest revealed bilateral crepitations and rhonchi with no other significant finding on systemic examination. On investigations patient had mild anaemia with leucocytosis and mild thrombocytosis along with positive CRP and

RF; other investigations viz. RFT, LFT, lipid profile, USG abdomen, ECG, fundoscopy were WNL. Sputum for AFB was negative. X ray chest and HRCT thorax was suggestive of multiple nodulo-cavitatory lesions. HPE of lung lesion showed granulomatous pathology. CT Angiography showed multiple calcified and non-calcified lesion of infra renal aorta with stenosis of Right dorsalis pedis artery. Biopsy of the skin lesion showed leucocytoclastic vasculitic lesion. We diagnosed GPA in light of clinical symptomatology with HPE of lung mass showing granulomatous lesion with positive cANCA. A few months after starting combined cyclophosphamide and steroids patient had significant symptomatic improvement.

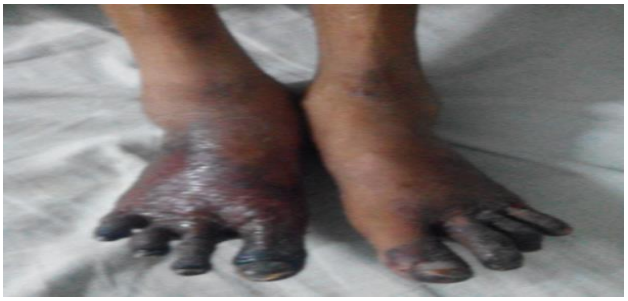

Figure 1: Clinical photograph of the patient showing digital gangrene of B/L lower limbs.

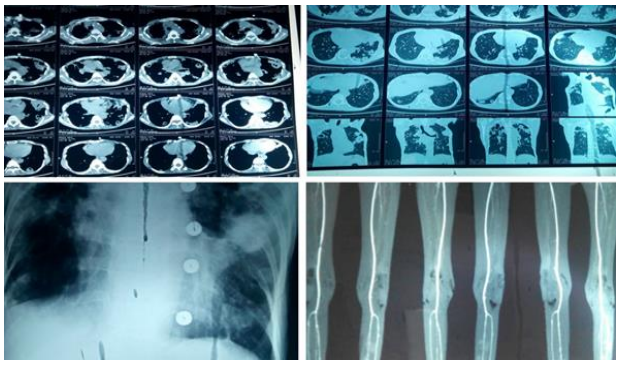

Figure 2: HRCT thorax images and CXR image showing multiple nodulo cavitatory lesions in B/L lung fields. Ct angiography of lower limbs showing multiple calcified and non-calcified lesion of infra renal aorta with stenosis of right dorsalis pedis artery (Clockwise from top left).

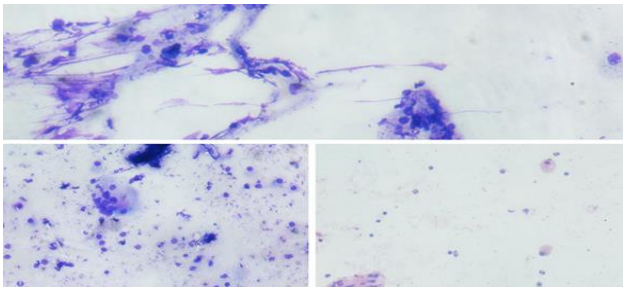

Figure 3: HPE slides showing granulomatous pathology.

DISCUSSION

The classical triad of the disease includes upper airway, lung, and kidney in most of the patients. Its causes are unknown, although microbes, such as bacteria and viruses, as well as genetics have been implicated in its pathogenesis.[2,3] Inflammation with granuloma formation against a nonspecific inflammatory background is the classical tissue abnormality in all organs affected by GPA.[4] Initial signs are extremely variable, and diagnosis can be severely delayed due to the nonspecific nature of the symptoms. In general, rhinitis is the first sign in most people.[2,4] Our patient also had sinusitis , B/L lung nodulo cavitatory opacities along with proteinuria and digital gangrene of B/L lower limbs. The disease is highly associated with the presence of antineutrophil cytoplasmic autoantibodies (ANCA) directed against proteinase 3 (PR3) or myeloperoxidase (MPO). 40 - 50% of WG patients have cutaneous manifestations still it is very rare as a presenting feature with a frequency of only 1% to 13%.[5,6] Skin findings include palpable and nonpalpable purpura, papules, subcutaneous nodules, ulcers, digital necrosis, splinter haemorrhages, and vesiculobullous lesions.[7] Diagnosis can reliably be made by histopathological examination along with clinical and immunological correlation. On histopathological examination, a biopsy will show leucocytoclastic vasculitis with necrotic changes and granulomatous inflammation (clumps of typically arranged white blood cells) on microscopy. In our patient too HPE was suggestive of leucocytoclastic vasculitis along with a positive cANCA. The standard treatment for GPA is cyclophosphamide and high dose corticosteroids for remission induction and less toxic immunosuppressant like azathioprine, leflunomide, methotrexate or mycophenolate mofetil.[8]

Our patient was also started on cyclophosphamide along with corticosteroids. With improved treatment regimens now the 5 year survival rate is around 80% however long term consequences like chronic kidney disease are common. Our patient has shown significant symptomatic improvement and is on regular follow up now.

CONCLUSION

Although digital gangrene is a very rare manifestation of Wegener's granulomatosis however its possibility should still be considered while evaluating a patient of digital gangrene with pulmonary and renal involvement to avoid diagnostic pitfalls and mismanagement

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