



A Case of ANCA Negative Granulomatosis with Polyangiitis Presenting with Peripheral Vascular Disease

Authors

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Abstract

Granulomatous with Polyangiitis (GPA) rarely presents as the peripheral vascular disease is an early manifestation. Most of the cases of GPA have ANCA positive (90%), and 10% are ANCA negative. A case of 32 yr old female who presented with painful gangrene of digits of both upper limb and lower limb and purpura on the lower leg and nasal crusting. CT angiogram of upper limbs shown radial and ulnar cut off and lower limbs narrowed lumen and non-visualisation of the posterior tibial artery. HRCT chest shows focal consolidation and cavitory lesions and soft tissue nodules in the right middle and lower lobes suggestive of granulomatous etiology. biopsy of skin shown vessels occluded with thrombi and transmural inflammation of vessel wall and renal involvement in the form of albuminuria, ANCA were negative. In view of clinical features and HRCT findings and biopsy reports diagnosed as ANCA negative GPA. According to ACR criteria, nasal involvement, lower respiratory tract involvement, renal involvement, and ANCA positivity, 2/4 is enough to diagnose GPA. In our case, three of them are seen. Patient was started on steroids and cyclophosphamide therapy; there was no progression of gangrene, and patient shown symptomatic improvement after treatment. There were less than 20 reported cases of GPA presenting as peripheral vascular disease. Hence GPA should be considered as a possibility in a patient with peripheral vascular disease.

Introduction

Granulomatosis with polyangiitis (GPA) or previously called Wegener's granulomatosis is a systemic autoimmune disorder that involves vessels causing both granulomatosis and polyangiitis. Friedrich Wegener first explained this in 1936¹. It is a multisystem disorder involving small and medium-sized vessels causing vasculitis in every organ possible and give rise manifestations of corresponding involvement². Most cases of GPA are ANCA positive, but 10 % of cases are ANCA negative. There are very fewer cases of GPA presenting as digital gangrene. Hence this is a unique case of ANCA negative GPA presenting as peripheral vascular disease.

Case Report

A 32 yr old female presented to the hospital with complaints of fever, arthralgia, nasal crusting since six months and 15 dayshistory of blackish discoloration of fingers and toes which are painful. No significant past, family, or personal history present. On examination, she was found to have pallor, reddish discoloration of the conjunctiva, Episcleritis, nasal crusting with ulceration and on filthrum and blackish discoloration of toes and fingers and palpable purpura on the lower limbs, with feeble dorsal is pedis artery pulsations on both sides and absent radial artery pulsations on both sides. On chest examination, mild crepitation was heard on right side and no other significant finding.

Investigations

HB	7.2gm/dl	S.creatinine	0.6mg/dl
TC	13600	B.urea	24 mg/dl
DC	P-70,L-23,E-6,M-1	T.bilirubin	0.7 mg/dl
PLATELET	2.14lakh/dl	Uric acid	1.3mg/dl
PS	Microcytichypochromic anemia	CUE	Alb-1+,sugar-nil,4-6 pus cells,2-3epithelial cells,no RBC
ESR	60mm/hr	24 hr urinary protein	960 mg /24 Hrs
CRP	Positive(70.2mg/l)	Blood C/S	sterile
ANA	Negative	HIV	negative
p-ANCA	negative	HBsAg	negative
c-ANCA	negative	HCV	negative

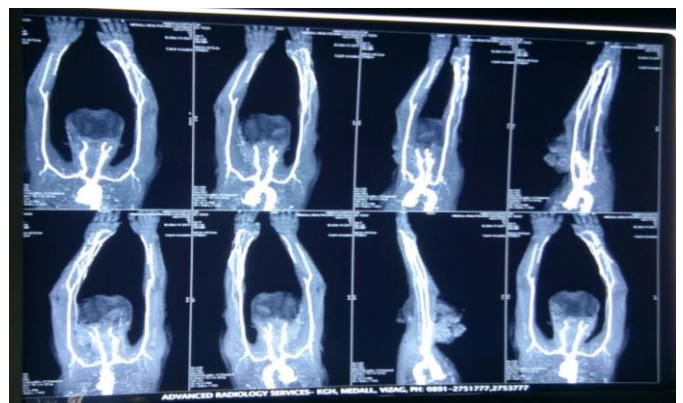


HRCT Chest -patchy air space opacities, focal consolidation, cavitory lesions and nodules noted in bilateral upper lobes and the right middle and lower lobes-diagnostic possibility of granulomatous etiology.



CT Angio of Upper Limbs

- 1) Total luminal occlusion with thrombosis of proximal right radial artery involving a segment of 5cm
- 2) Total luminal occlusion of right distal ulnar artery and its branches in hand
- 3) Sparse contrast opacification with near overall luminal narrowing of left distal ulnar artery and its branches



CT Angio of Lower Limbs

- 1) the narrowed lumen of upper 1/3rd of posterior tibial artery
- 2) Non visualized the lower 2/3rd posterior tibial artery



HPE of Skin Biopsy- Dermis showing occluded vessels with thrombi, fibrin material, and transmural inflammatory cells. There is an exhaustion of RBC and perivascular mixed inflammatory cells comprising of neutrophils, lymphocytes and nuclear debris suggestive of vasculitis.

Discussion

Wegener's granulomatosis was recently changed as GPA in 2011. ACR CRITERIA for GPA includes nasal or oral inflammation, abnormal chest radiograph, abnormal urinary sediment, granulomatous inflammation on biopsy of artery⁴ or perivascular area. Two or more of these criteria yielded high sensitivity. Our patient has nasal inflammation and pulmonary nodules, and biopsy showing vasculitis and eye involvement in the form of episcleritis and digital gangrene and palpable purpura. Bilateral or unilateral pulmonary infiltrates are present in nearly 50% of patients initially, with lung disease eventually developing in 85–90% of patients. Pleural effusion has also been reported in 12% of cases⁵. ANCA is mostly positive in 90% of patients, but in our patient, it is negative, which is seen in 10 percent of cases.

The standard treatment for GPA is cyclophosphamide and high dose corticosteroids for remission induction and less toxic immunosuppressant like azathioprine, leflunomide, methotrexate, or mycophenolatemofetil.

Conclusion

GPA presenting as digital gangrene is very rare, and ANCA negative in GPA is also unique. There were less than 20 reported cases of GPA presenting as peripheral vascular disease, so patient presenting with peripheral vascular disease, GPA should be considered in evaluating the cases.

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