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Case Report

Perinuclear anti neutrophil cytoplasmic antibody vasculitis with nonspecific muscle pain

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ABSTRACT

This is a case report describing p-ANCA vasculitis presenting with nonspecific muscle pain. It is a very uncommon finding seen in p-ANCA vasculitis patients which they usually present with palpable purpura alone. In this case, along with nonspecific muscle pain, renal involvement of the disease has been explained and there are no upper and lower respiratory tract complaints which favours more towards microscopic polyangitis.

Keywords: Non-specific muscle pain, p-ANCA vasculitis, Palpable purpura

INTRODUCTION

Anti-Neutrophil Cytoplasmic Antibody (ANCA) associated vasculitis (AAV) is a group of small vessels vasculitides characterized by destruction and inflammation of small blood vessels by neutrophils. There are two major ANCA autoantibodies- the cytoplasmic (c-ANCA), which confers antigen specificity for proteinase-3 and perinuclear (p-ANCA), with specificity for Myeloperoxidase enzyme. The cytoplasmic and perinuclear forms refer to the pattern of reactivity seen by indirect immunofluorescence test on alcohol-fixed test cells exposed to patients's serum carrying ANCA antibodies.[1]

CASE REPORT

A 45 year old postmenopausal married female hailing from Sivsagar district, Assam state, presented to medicine department with complaints of chest pain and back pain for 2 months and skin rash which are non-itchy started on both lower limb followed by both upper limb for 2 weeks. There was no history of fever, cough, difficulty in breathing, joint pain, loss of appetite, loss of weight. There was no history of easy fatiguability, nasal congestion, numbness and tingling sensation over limbs. There was no past history of Bronchial asthma.

Patient was newly diagnosed as hypertension of systolic blood pressure 140 mmHg and diastolic blood pressure of 90 mmHg. On general examination, mildly pale, no conjunctival congestion, no nasal polyp, no oral ulcers, no lymphadenopathy, no pedal edema were found. On systemic examination, no wheeze heard over chest, sensations were absolutely normal, no spinal tenderness. On Local examination, palpable erythematous non blanchable papular rash present over bilateral lower limb and bilateral upper limb (over both flexor and extensor surfaces) (Figure 1).

On investigation, Hb% was 9.1 gm/dl, total WBC count was 6200 cells/mm^3, peripheral blood smear showed mild anisocytosis with mild hypochromia, ESR was 140 mm/hour, CRP was 4.73 mg/dl, urine routine investigation showed 4-6 WBC/Hpf, 5-10 RBCs/ Hpf, Albumin- 3+, Urine culture and sensitivity shows no growth of any microorganism, Liver function tests were normal, Kidney

function tests were normal. ECG was within normal limits, X-ray chest (Postero-anterior view) was normal, X-ray dorso-lumbar spine was normal, X-ray lumbo-sacral spine showed degenerative changes in multiple vertebral levels. HRCT thorax revealed fibrotic opacity is noted in apico-posterior segment of left upper lobe suggestive of sequale of past pulmonary infection without any evidence of present activity. Despite of all biochemical and radiological investigation, the cause for chest pain and back pain was not clearly found so that authors provisionally diagnosed it to be a nonspecific muscle pain.

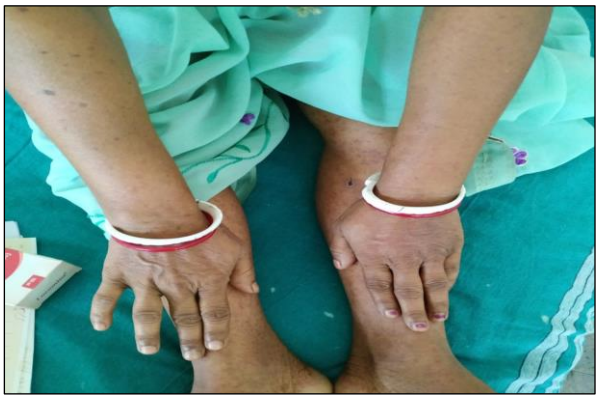

Figure 1: Palpable purpura over bilateral upper limbs and lower limbs.

Based on clinical knowledge and experience authors have proceeded for further investigation, to rule out Systemic lupus erythematosus authors did ANA IFA which was negative (1:80 dilution). Then authors have proceeded to do ANCA profile, in that c-ANCA found to be negative but p-ANCA was in borderline [Myeloperoxidase- 1+].

Based on clinical findings, biochemical and immunological tests authors have provisionally diagnosed it would be a case of p-ANCA vasculitis and then authors have proceeded for histopathological examination of skin biopsy from leg which showed a mass of edematous and haemorrhagic fibro-collagenous tissue with areas of perivascular chronic inflammatory changes picture suggestive of vasculitis.

Then authors have started on oral PREDNISOLONE 1 mg/kg daily and anti-hypertensive drugs following which back pain and chest pain got subsided and skin rashes got subsided and there are no further episode of skin rashes during follow-up but the patient developed bilateral pitting pedal edema and authors have repeated 24 hour urinary protein which was found out to be 3.20 gram with a total volume of 2 litre in 24 hours and now the renal involvement of the disease has been confirmed and authors have started on tablet Mycophenolate mofetil 500 mg twice daily and there was no complaint during follow-up.

DISCUSSION

As per recent study, ANCA associated vasculitis have annual incidence ranging from 1.2 to 2.0 cases per 1,00,000 individuals and a prevalence of 4.6-18.4 cases per 1,00,000 individuals.[2] The term "polyangiitis" is preferred to "polyarteritis" for Microscopic Polyangitis because of the tendency of the disease to involve veins as well as arteries. The Chapel Hill Consensus Conference (2012) defined Microscopic Polyangitis as a process that i) involves necrotizing vasculitis with few or no immune deposits; ii) affects small blood vessels (capillaries, arterioles, or venules) and possibly medium-sized vessels, as well; and iii) demonstrates a tropism for the kidneys and lungs.[3] According to study done by Shunichiro Hanai et al, the diagnosis of Microscopic polyangitis is very difficult only by constitutional symptoms like such as nonspecific muscle pain, but in this case along with nonspecific muscle pain, patient was having skin manifestations like palpable purpura which help us to diagnose the condition.[4]

CONCLUSION

Thus, from this case report authors have found that rare disease like Microscopic Polyangitis can present with trivial complaint like muscle pain and therefore in this type of presentation, a high degree of clinical suspiction, laboratory findings and skin biopsy are required to establish the disease.

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