CASE REPORT

ACRO-OSTEOLYSIS AND MONONEURITIS MULTIPLEX AS A PRESENTING SYMPTOM OF SYSTEMIC ANGIITIS OF WEGENER’S TYPE

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ABSTRACT

Wegener’s granulomatosis is a multisystem disorder involving small- and medium-sized vessels, leading to granuloma formation and involvement of upper and lower respiratory tract with or without glomerulonephritis. However, limited forms of angiitis and granulomatosis of the Wegener’s type with oligosymptomatic and atypical site involvement are known to occur. We present here a rare case of limited form of angiitis and granulomatosis of Wegener’s type who presented sequentially with spontaneous resolution of digits with acro-osteolysis and mononeuritis multiplex over a period of 10 months. His vasculitic workup revealed high proteinase 3 antibodies (c-ANCA) titters and an almost asymptomatic lung involvement, detected on high-resolution computed tomography of chest. The patient was aggressively treated with immunosuppressive therapy, following which he showed good improvement.

Key words: Acro-osteolysis, mononeuritis multiplex, Wegener’s granulomatosis

Wegener’s granulomatosis, a disorder characterized by necrotizing vasculitis of small arteries and veins, typically involves multiple systems, but the upper and lower respiratory tract, with or without kidneys, suffer the brunt of the disease burden. The diagnosis is simple, provided typical sites are involved and tissue diagnosis is available. However, in the limited forms, when rare presenting symptoms are present or where unusual sites are involved, the characteristic radiological features and surrogate parameters for granulomatous inflammation assume increasing importance.

CASE REPORT

A 49-year-old male presented with history of bluish discoloration of the fingertips of both hands culminating into spontaneous resorption of fingertips of 10 months duration. He was empirically treated outside with low-dose oral steroids (30 mg/day) and analgesics without much relief. Later he
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presented with numbness and burning paresthesias on the anterior and lateral aspects of left leg and dorsum of left foot of 4 weeks’ duration, followed by difficulty in lifting left foot against gravity. Patient also had intermittent fever (up to 101°F) with generalized myalgia and weight loss of about 8 kg. There was no history of cough, hemoptysis, pleuritic chest pain, epistaxis, ulceration or deformity of nose. There was no history of joint pains and/or swelling, oral or genital ulcers, photosensitivity, skin rash or dryness of eyes.

Examination revealed resorption of fingertips of both hands (left > right) [Figure 1]. The motor system examination revealed decreased tone across left ankle joint, motor power of grade 2/5 (on MRC Scale) in the left middle and ring fingers [Figure 2]. A nerve biopsy taken from left sural nerve revealed evidence of axonal loss. Nerve conduction study revealed bilaterally nonstimulable lower limb motor peroneal and ulnar nerves was normal, supporting the clinical diagnosis of mononeuritis multiplex with confluence. Investigations revealed normal hemoglobin and a normal total and differential leukocyte count. The absolute eosinophil count was 100/mm³. The ESR was 45 mm in the first hour. The vasculitic and immune workup revealed c-reactive protein (CRP) positivity, proteinase 3 antibodies (c-ANCA) titers of 48.40 units/ml (reference range <7.00 units/ml), antmyeloperoxidase antibodies (p-ANCA) titers of 4.50 units/ml (reference range <7.00 units/ml). Other immunological investigations like antinuclear antibody, rheumatoid factor, cryoglobulins, hepatitis B virus surface antigen, hepatitis C IgG antibody and HIV (1 and 2) serology were negative. The 24-hour urinary proteins were nil on two occasions, and no red blood cells or casts were detected in the urine. His X-ray of both hands showed acro-osteolysis of distal phalanges of bilateral index fingers and ray of both hands showed acro-osteolysis of both hands (Rt. > Lt.) X-rays showing spontaneous resorption of distal phalanges of both hands (Rt. > Lt.)

In the view of c-ANCA positivity, patient was actively worked up for Wegener’s granulomatosis. The high-resolution computed tomography (HRCT) of chest showed a solitary 5 mm × 10 mm cavitating nodule in the lingula abutting the pericardium [Figure 3]. The microscopic examination of sputum for acid fast bacilli and malignant cells was negative on three consecutive occasions. A fine needle aspiration cytology/transthoracic biopsy from the cavitating nodule was planned for tissue diagnosis but was refused by the patient. Computed tomography of paranasal sinuses was essentially normal. Considering the radiological finding of cavitating nodule as a surrogate parameter for active granulomatous inflammation of lower respiratory tract, c-ANCA titer positivity and lack of eosinophilia in blood, this patient satisfied the Sorenson’s criteria for Wegener’s Granulomatosis. Because of clinical worsening, monthly pulse Cyclophosphamide was started and the dose of oral steroids was optimized. On 6-month follow-up, the patient showed gradual improvement in neurological symptoms, repeat CT chest revealed resolution of lesions and c-ANCA titers reduced to 7.50 units/ml.

DISCUSSION

The presenting signs and symptoms of Wegener’s granulomatosis are attributable to involvement of ear, nose and throat (ENT) in 73%, lungs 45%, kidney 18% and eye in 15% of cases.

The involvement of nervous system as a presenting manifestation is distinctly rare and includes cerebrovascular events in 4%, peripheral neuropathy in 2.4% and cranial neuropathy in 2.2% of cases and occasionally seizures. To our knowledge, acro-osteolysis of digits as a presenting symptom of Wegener’s granulomatosis has not been previously reported in the literature. The recognition of rare manifestations of this relatively uncommon disease is extremely important, as the spontaneous 2-year mortality rate of Wegener’s granulomatosis of 93% has declined dramatically to a 5-year survival exceeding 60% in the 1990s since the introduction of immunosuppressive therapy.

The American College of Rheumatology (ACR) criteria for Wegener’s granulomatosis mandates demonstration of granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area (artery or arteriole) on biopsy. This is not always possible because of the segmental and dynamic nature of the vasculitis process and difficulty in obtaining representative biopsies from inaccessible areas, as in our case owing to close proximity of lesion to the heart. In such a situation, noninvasive investigation like CT chest showing a cavitating pulmonary nodule reflects an active pulmonary disease in patients with Wegener’s granulomatosis, which usually...

Figure 1: Figure showing resorption of fingertips of both hands (Rt. > Lt.)

Figure 2: X-rays showing spontaneous resorption of distal phalanges of both hands (Rt. > Lt.)

Figure 3: CECT of chest showing cavitating nodular lesion in the lingula abutting pericardium

Figure showing resorption of fingertips of both hands (Rt. > Lt.)
shows resolution with therapy.[5] Likewise, Sorensen et al. considered radiologically demonstrated pulmonary infiltrates or cavitations of more than 1 month's duration in patients suspected of having Wegener's granulomatosis as surrogate marker of granulomatous inflammation of lower airways, provided infection and malignancy are ruled out.[5] In view of negative sputum samples for acid fast bacilli and malignant cells, presence of surrogate markers like c-ANCA and cavitating nodule seen on HRCT chest in a patient with evidence of systemic vasculitis (spontaneous acro-osteolysis and digital resorption, foot drop and history of fever with weight loss), we had no better diagnosis than systemic vasculitis and granulomatosis of the Wegener's type.

REFERENCES


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