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Case Report

Extensive gastrointestinal tract and thyroid involvement with Wegener’s granulomatosis

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Wegener’s granulomatosis (WG) is a pauci-immune systemic vasculitis involving small to medium sized blood vessels of the respiratory tract and renal vasculature. We report a 34-year-old lady with extensive gastrointestinal tract, pancreas and thyroid involvement. Literature review revealed only two prior reports of esophageal involvement, two reports of pancreatic involvement and few cases of thyroid involvement. [Indian J Gastroenterol 2007;26:290-291]

Case Report

A 34-year-old white woman was being treated with various antibiotics since 9-10 weeks for symptoms of refractory sinusitis and epistaxis. During the same period, she developed small erythematous papules on her lower extremities, which evolved into target-like lesions with central necrosis (Fig 1), and necrosis of fingers and toes due to digital ischemia. Laboratory studies revealed acute renal failure.

Her temperature was 95.8 °F, and blood pressure 126/78 mmHg. She had painful sores in her mouth, erosions in the nasal cartilage and palate, left-sided otitis media, target-like lesions with central necrosis on her lower extremities and digital ischemia (Fig 1), and necrosis of fingers and toes due to digital ischemia. Radiography showed diffuse bilateral pulmonary infiltrates and CT scan of the sinuses revealed pan-sinusitis. Negative blood cultures and a negative transesophageal echocardiogram ruled out infective endocarditis.

Investigations: WBC count 22,010/mm³, hemoglobin 8.9 gm/dL, hematocrit 27.9%, platelet count 227,000/mm³, ESR 113 mm/h and creatinine 6.1 mg/dL. CRP 44.5 mg/dL, antineutrophil cytoplasmic antibody (C-ANCA) titer 1:320, antiproteinase-3 antibody (anti PR-3) >100 U/mL (normal <3.5), TSH 18.8 U IU/mL (0.3-4.8), thyroid peroxidase antibodies 380 IU/mL, T3 uptake 39%, T4 2.8 mg/dL (4.5-12.5), C3 147 mg/dL, C4 38 mg/dL and elevated rheumatoid factor 115.9 IU/mL (0-13.9). Tests for myeloperoxidase antibody (MPO), anti-cardiolipin antibody, anti-glomerular basement membrane antibody, antinuclear antibody (ANA), double-stranded (dS) DNA antibody and hepatitis were negative. Urine analysis showed hematuria.

Punch biopsy of the skin lesions showed dense superficial and deep perivascular and intervascular infiltrates of lymphocytes, histiocytes and neutrophils with prominent fibrinoid change in perivascular connective tissue in papillary dermis, mid and deep reticular dermis and subcutaneous adipose tissue. Histology was consistent with necrotizing vasculitis involving small vessels. Mucosal biopsy of the oropharyngeal lesion showed acute surface ulceration.

She fulfilled the American College of Rheumatology 1990 classification criteria and Sorensen’s diagnostic criteria for WG. Methylprednisolone (60 mg intravenously 6 hourly) and cyclophosphamide (100 mg intravenously once a day), hemodialysis and thyroid replacement were initiated.

A week after presentation, the patient complained of epigastric discomfort and dysphagia. MRI of the abdomen showed an inhomogeneous mass in the body of pancreas. Serum amylase and lipase were normal. Liver biochemistry showed low albumin but normal aminotransferases, total bilirubin and alkaline phosphatase. Gastroscopy revealed multiple oral ulcers, severe circumferential punched out ulcerations between 21-36 cm from the incisors (Fig 2), gastroduodenitis and multiple duodenal ulcers. Esophageal biopsy showed acute ulcerative esophagitis with detached and acutely inflamed fibrinohemorrhagic detritus. Duodenal biopsy showed acute inflammation. Symptomatic improvement was noted with antacid (Mylanta; Johnson and Johnson, Fort Washington, USA), xylocaine mixture and esomeprazole. She underwent hemodialysis for renal failure, and was discharged.

Fig 1: Erythematous papules evolved into target-like lesions with central necrosis (left) and digital necrosis

Fig 2: Severe circumferential ulceration with areas of punched out ulcers (21-36 cm from the incisors)
home on oral prednisone (60 mg once a day), oral cyclophosphamide (50 mg once a day) and thyroid supplements.

Eight weeks later, she complained of bloody diarrhea and dysphagia. CT scans of abdomen and pelvis with oral contrast revealed bowel wall thickening involving the ascending colon. A 2.5 cm mass was noted near the junction of the tail and body of pancreas. Colonoscopy showed deep ulcerations throughout the colon and rectum. Biopsies showed non-specific focal ulceration. *Clostridium difficile* toxin was positive and oral metronidazole was started.

Repeat gastroscopy done because of dysphagia showed a stricture at 26 cm from the incisors; the stricture was dilated up to 15 mm with through-the-scope (TTS) balloon. Subsequently the patient had multiple hospitalizations for dysphagia and abdominal pain. Multiple esophageal structure dilations using TTS balloon technique were done. Repeat CT scan of the abdomen suggested chronic pancreatitis. Elevation of amylase to 273 IU/L, and lipase to 74 IU/L was noted only once. She lost 18 Kg of weight. She was briefly on tube feedings via percutaneous endoscopic gastrostomy (PEG) tube. Skin, oral and digital lesions improved with immunosuppressive therapy but, she developed “saddle-nose” deformity. Since she developed leucopenia, cyclophosphamide was replaced with azathioprine. At the current time, patient has resumed oral feeding with steady weight gain. She is continued on hemodialysis, prednisone, azathioprine and thyroid replacements.

**Discussion**

Wegener’s granulomatosis is a multi-systemic, pauci-immune systemic vasculitis of unknown etiology. It primarily involves small to medium sized blood vessels of the upper and lower respiratory tract and renal vasculature. It can occur at any age and has no gender preponderance. Frequency of gastrointestinal involvement with WG is about 5-10%. Involvement of different caliber vessels of the intestinal wall results in lesions ranging from patchy focal ischemia with mucosal ulceration to intestinal infarction, gangrene and perforation. Bleeding within the intestinal wall and into the bowel lumen can occur with aneurysmal dilatation and rupture of vessels. Though GI involvement has been detected at autopsy in a number of cases, clinical manifestations of severe intestinal disease have been infrequently reported.

The diagnosis of GI involvement in systemic vasculitis rests on the evidence of extra-intestinal disease. Histologically vasculitis, ischemia and ulceration are the most common findings. Reports of WG involving small and large bowel in the form of hemorrhagic colitis with ulcer formation and intestinal perforation have been previously published. Esophageal and pancreatic involvement with WG is rare. There are only two case reports of clinical acute pancreatitis; vasculitis of pancreatic vessels has been observed in autopsy studies.

Thyroid involvement in WG is also rare. In our patient with no previously documented thyroid disease, elevated thyroid peroxidase and TSH levels, with low T4 and low T3 uptake were noted. The underlying mechanism for increased antithyroid antibodies in WG is unclear. Presence of autoantibodies directed against various human antigens in autoimmune diseases may be a plausible explanation.

**References**


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