

PICTURES IN DIGESTIVE PATHOLOGY

Severe intestinal involvement in Wegener's granulomatosis with negative c-ANCA

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CASE REPORT

A 28-year-old patient with a history of ulcerated lesions in nasal cavity and necrotic skin lesions in legs and arms presented with bouts of diarrhea and recurrent bowel obstruction, which led us to suspect that the patient suffered from Crohn's disease or Wegener's granulomatosis (WG); the patient died before a final diagnosis was reached. A cutaneous biopsy showed leucocytoclastic vasculitis consistent with WG. However, colonoscopy, gastrointestinal X-rays examination, and small-bowel biopsies were all compatible with Crohn's disease, which affected the large bowel and terminal ileum with a stricture at the hepatic angle due to an ulceration; cANCA were negative over the process. The patient received various immunosuppressive agents with no success (cyclophosphamide, methotrexate, infliximab). Upper airway involvement progressed in such a way that the patient needed a percutaneous gastrostomy, a tracheostomy and a masseter myotomy for trismus and dyspnea. During surgical intervention a huge distension of small bowel (ileum and distal jejunum) was seen, with significant wall thickening, and no evidence of stops or perforation (Fig. 1). This is why we only performed a biopsy of terminal ileum. The patient died within 24 hours after surgery while in the critical care unit from multiple organ failure. The autopsy showed a condition consisting of small-vessel vasculitis (arterioles and venules) and occasional histiocytes around vessels forming loose granulomas with scattered multinuclear giant cells (foreign-body type) (Fig. 2). This lesion pattern involving the small and large bowel, trachea, and bronchi, in association with focal and segmentary glomerulonephritis, destruction of the nasal septum with a "saddle nose" (Fig. 1), and oral ulcers are finally diagnostic of WG.

The diagnosis of WG is histologic, and includes: a) acute necrotic granulomas of upper and lower airways; b) focal necrotic vasculitis of small and medium vessels; and c) renal disease in the form of focal or diffuse necrotic glomerulonephritis. Typical clinical features include: persistent pneumonitis with bilateral nodular and cavitory pulmonary infiltrates (95%), chronic sinusitis (90%), mucosal ulcerations at nasopharynx (75%), and evidence of renal disease (80%).

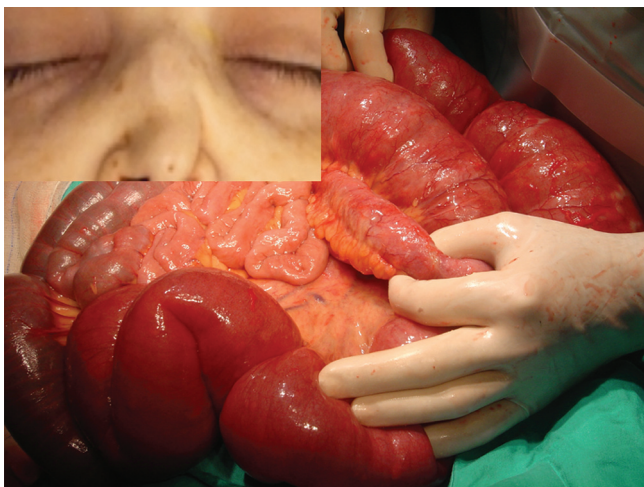


Fig. 1.- A close-up of a saddle nose, and gross involvement of the small bowel.

Detalle de nariz en silla de montar y afectación macroscópica de intestino delgado.

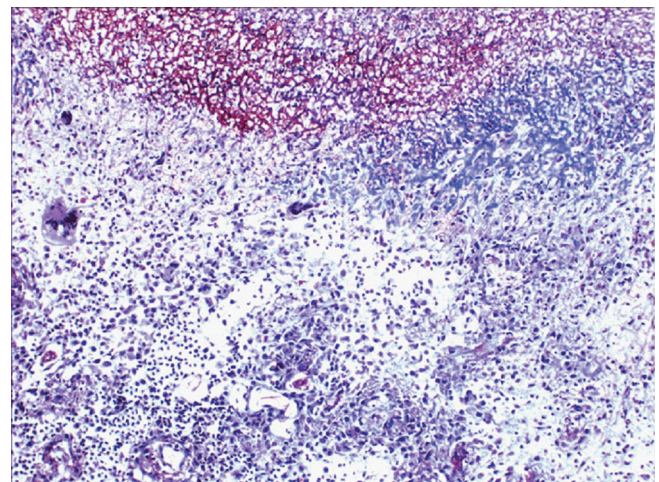


Fig. 2.- A microscopic view of the intestinal wall (trichromic, 20X).

Microscopía pared intestinal (tricrómico 20X).

Bowel affectation as initial or main symptom, though rare, has been described, mainly as bloody diarrhea (1,2), perforation (3,4) or, less frequently, as bowel obstruction (3), which, as was the case with our patient, may lead to death. cANCA_s are positive in more than 93% of patients with active disease; however, this was never the case with our patient.

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