Small intestinal perforation complicating Wegener’s granulomatosis

Sir, Storesund et al. [1] recently reported two patients in whom gastrointestinal vasculitis complicating
Wegener’s granulomatosis required surgical intervention. Severe intestinal involvement of this nature is very rare in this condition and the occurrence of bowel perforation is rarer still with only a handful of cases described in the literature. These reports were reviewed by the authors and all were found to share certain clinical features. First, it was noted that serious gastrointestinal pathology occurred only in patients with extensive involvement of other organs, i.e. pulmonary and renal disease. The second interesting comment related to the extent of underlying disease activity when gastrointestinal symptoms first emerged. The authors considered that intestinal disease was likely to occur during active phases of the disease, and most instances of bowel ischaemia previously described in Wegener’s granulomatosis have occurred within a very short time of diagnosis and the commencement of immunosuppressive therapy.

We have encountered a patient whose clinical course differs considerably from past experience and may suggest that a different perspective be cast on this rare complication. A 56-yr-old female patient presented to the ENT outpatient department with a 4 week history of painful swelling of the nasal bridge with associated rhinorrhea, intermittent fever, arthralgia and weight loss. Investigations confirmed the presence of an acute phase response with raised ESR and CRP. Autoantibody screen was positive for cANCA. Subsequent nasal biopsy showed areas of acute inflammation with focal areas of geographic necrosis surrounded by a macrophage response in the form of palisaded histiocytes and occasional multinucleated giant cells; there were no vasculitic changes. Chest X-ray was normal and renal function, including creatinine clearance and urinalysis for sediment, was entirely normal.

A diagnosis of Wegener’s granulomatosis was made and the patient had an excellent clinical response to prednisolone 40 mg and cyclophosphamide 100 mg daily. Over the following 6 weeks, she presented on two occasions with crampy abdominal pain which was self-limiting; investigations including barium follow-through and enema were normal. On both occasions, she was otherwise well and ESR and CRP were within normal limits. Six weeks later, she was admitted with peritonitis. ESR and CRP had risen to 20 and 3.0, respectively, but she had no clinical evidence of active disease elsewhere. A laparotomy was performed and revealed three discrete areas of perforation, serosal thickening and granulation in the small bowel; one of the areas of perforation was surrounded by a granulomatous reaction, although the blood vessels did not appear inflamed. Wide local resection of the diseased bowel was performed and the patient made an uncomplicated recovery thereafter on continued oral prednisolone and cyclophosphamide.

This patient is notable for being the first patient with limited Wegener’s granulomatosis, i.e. without pulmonary or renal involvement, who has developed gastrointestinal perforation in the course of her disease. Although some patients presenting with limited disease progress to develop multiorgan involvement, previous cases documenting intestinal involvement have exclusively described this complication in the setting of established pulmonary and renal involvement. Furthermore, the patient has been followed up for some 5 yr and has not exhibited disease spread. It is also of interest that gastrointestinal complications arose at a time when her disease was otherwise quiescent on immunosuppressive treatment, in contrast to the previous experience which documented severe gastrointestinal pathology in the setting of active disease elsewhere. Her original gastrointestinal symptoms may have been secondary to small bowel vasculitis, the features of which were modified by immunosuppression. The progression of her small bowel disease to perforation, at a time when her disease was otherwise quiescent, raises two possibilities. First, it may indicate that gastrointestinal vasculitis in these patients pursues a different course and may require more aggressive treatment. The absence of vasculitic changes in the small bowel biopsies mitigates against, but does not exclude, this eventuality. It is more likely that she had suffered small bowel vasculitis early on in her disease course, resulting in a weakened bowel wall with areas of granulomatous change, and continued treatment with immunosuppressive agents may thereafter have contributed to the development of perforation. This case illustrates that gastrointestinal ischaemia may complicate Wegener’s granulomatosis of a limited nature and in the setting of otherwise well-controlled disease.

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