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# Letters to the Editors

## Saddle nose deformity in a patient with Crohn's disease

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Dear Sirs,

We read with interest the report by Merkonidis *et al.* regarding the development of a saddle nose deformity in a patient with Crohn's disease.[1] We would like to offer a different perspective on the data they present and a different message which readers might take from the report.

The patient described had nasal inflammation evident clinically and necrotizing granulomatous inflammation on biopsy of a nasal ulcer, thereby meeting the American College of Rheumatology classification criteria for the diagnosis of Wegener's granulomatosis (WG).[2] That he did not have vasculitis evident clinically at the time of assessment does not exclude the diagnosis, as the manifestations given in this report are those of limited disease in which clinical evidence of vasculitis may be lacking.[3] Similarly, the absence of vasculitis on biopsy of the nasal septum, even in the presence of necrotizing granulomatous inflammation, has been described in an adult WG series.[4] The appearance of the nasal biopsy may also have been modified if the patient was receiving immunosuppressive therapy for his bowel disease at the time of biopsy. The patient also had a positive antineutrophil cytoplasmic antibody test with a predominant specificity for PR3 – an uncommon finding in inflammatory bowel disease (present in just 2.8 per cent of patients with Crohn's disease in the study by Walmsley *et al.*[5] cited by the authors) but found in 65–91 per cent of adult patients with active WG.[6] The other extra-intestinal features described in Merkonidis and colleagues' patient, such as arthralgias and episcleritis, although seen in association with Crohn's disease, are also common in active WG. While the normal urinalysis reported by the authors makes generalized WG unlikely, it does not exclude localized disease. Given these concerns, we feel that it would have been appropriate to investigate further with computed tomography scans of both the sinuses and the chest, the latter looking for manifestations such as nodules and 'ground glass' appearance of the lung parenchyma, which may not be evident on plain radiographs. Even if these were normal, careful follow up with regular monitoring of urinary sediment would be prudent, as patients with localized WG may develop renal involvement over time.

We do not dismiss the possibility that this patient's unusual presentation may have been purely the result of Crohn's disease. We do feel, however, that WG was not completely excluded. Indeed, even with the additional investigations suggested, it may not be possible to absolutely 'rule out' WG in this patient. In light of this and of reports describing the coexistence of Crohn's disease and WG in some patients[7,8] and of WG initially misdiagnosed as Crohn's disease (revealed only later by the onset of renal failure),[9] we urge follow up with these possibilities in mind.

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*Authors' reply*

We thank the above correspondents for their pertinent comments. In presenting this interesting case, we wished to highlight and raise for discussion the inherent difficulties in the diagnosis of this patient's nasal symptoms and signs, in the setting of a longstanding history of Crohn's disease.

In this patient, a diagnosis of Crohn's disease was made on the basis of histological specimens from intestinal biopsies from perianal fistulae and intersphincteric abcesses. However, as we mentioned in the original paper, we certainly could not rule out the possibility that the patient's nasal findings may have been accounted for by Wegener's granulomatosis. We do acknowledge that the absence of vasculitis does not exclude a diagnosis of Wegener's granulomatosis but nonetheless this is not a typical finding in the condition. Furthermore, the co-existence of Crohn's disease and Wegener's granulomatosis in the same patient, whilst described, is in itself an extremely unusual finding.

Nonetheless, in consideration of this possibility, the patient has undergone a magnetic resonance imaging scan of the sinuses that has revealed circumferential mural thickening within the right maxillary sinus and partial destruction of the lateral wall of the nasal cavity and septum. These changes could be consistent with localized Wegener's granulomatosis, but equally they do not

rule out the possibility of nasal Crohn's disease. To date, the patient has had normal renal function and urinalysis as well as unremarkable lung function tests, rendering a diagnosis of generalized Wegener's granulomatosis unlikely.

We appreciate that the diagnosis of both Crohn's disease and Wegener's granulomatosis can often be problematic. Whilst open to alternative perspectives, we feel that our case emphasizes the need to correlate clinical findings with serology, histopathology and radiology, since no single modality is diagnostically definitive.

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