

Saddle nose deformity in a patient with Crohn's disease

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Abstract

Crohn's disease is a chronic idiopathic inflammatory disease of the bowel, and in most cases it involves the small bowel and colon. Extraintestinal manifestations occur frequently and multiple organ systems may be affected. In contrast, nasal manifestations are extremely rare and only a few cases have been reported to date. Saddle nose deformity in a patient with Crohn's disease has not been reported in the English literature. We report a case of such a deformity and the current literature on nasal manifestations in Crohn's is reviewed. The normal diagnostic and therapeutic features are also discussed.

Key words: Crohn's Disease; Nose Deformities; Acquired

Introduction

Crohn's disease is a chronic idiopathic inflammatory disease of the gastrointestinal tract. The diagnosis is made on a constellation of radiological, endoscopic and histological findings in the appropriate clinical setting.¹ Combined radiological and endoscopic findings in conjunction with the anatomic pattern of involvement form the basis of the diagnosis and this is confirmed by histological examination of biopsy specimens - where chronic granulomatous inflammatory changes are found.

Extraintestinal manifestations are common, and have been described in association with almost every organ system. Nasal manifestations are exceptionally rare with only eight cases reported to date. These have included nasal obstruction, rhinorrhoea, recurrent epistaxis, paranasal sinus disease and nasal septal perforation.¹⁻⁸ In this report we describe saddle nose deformity in a patient with Crohn's ileitis, pancolitis, episcleritis and arthralgia. A review of the available literature on nasal involvement in Crohn's disease is also presented.

Case report

A 24-year-old man presented with a three-month history of saddle nose deformity (Figure 1). He also had bilateral nasal obstruction, nasal discharge and intermittent epistaxis for seven months. The nasal obstruction was most marked on the right side and was accompanied by the production of gelatinous casts upon clearing of the nose. Three years earlier he had been diagnosed with Crohn's colitis following analysis of intestinal biopsies from perianal fistulae and intersphincteric abscesses. His symptoms at the time were controlled with systemic corticosteroids and second-line immunosuppressants.

Nine months prior to his presentation he had experienced an exacerbation of his intestinal symptoms and soon afterwards developed right episcleritis, arthralgia and nasal symptoms.

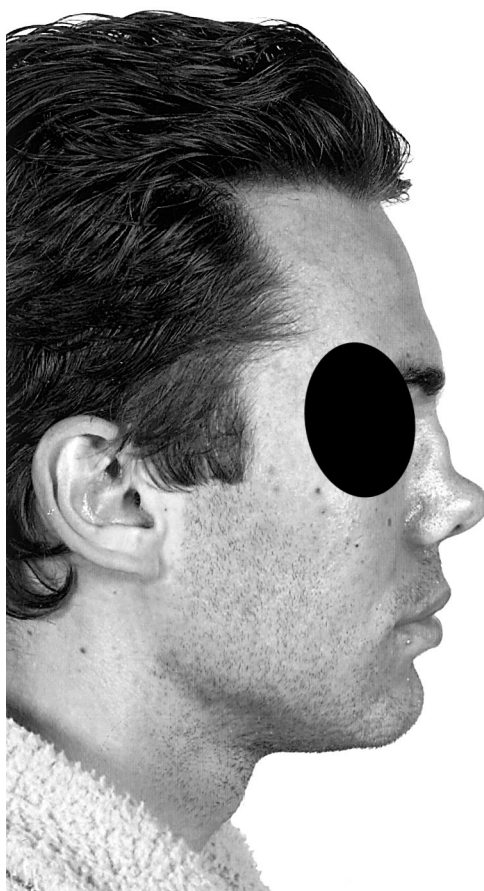


FIG. 1
Nasal collapse resulting in a saddle nose deformity.

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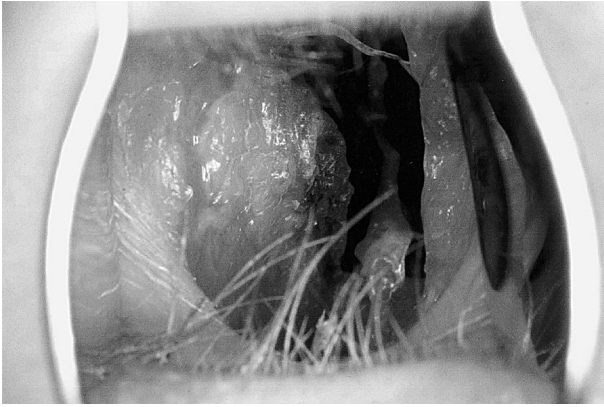


FIG. 2

A view into the right nasal cavity of our patient illustrating crusting and ulceration.

On examination he was afebrile and well nourished. He had acne on the torso but no other skin lesions. There was no history of sinusitis or deafness and the aural cartilages were unremarkable. No oral lesions were found. The cardiovascular, respiratory and abdominal systems were all normal. Laboratory investigations revealed an elevated C-reactive protein of 31 and an erythrocyte sedimentation rate (ESR) of 72. Indirect immunofluorescence of ethanol fixed neutrophils showed his anti-neutrophil cytoplasmic antibody (ANCA) to be positive with proteinase-3 (38) and myeloperoxidase (7) activity. Neutrophil and lymphocyte counts were normal, as were angiotensin converting enzyme (ACE) levels. He had negative fluorescent treponema pallidum antibody (FTPA), and a chest X-ray and urinalysis were unremarkable.

Examination of the nose revealed a highly inflamed nasal mucosa with extensive crusting in both nasal cavities. There was deep ulceration to both inferior turbinates, extending to the aggar nasi on both sides of the nasal septum (Figure 2). A biopsy of the nasal lesions showed acute on chronic granulomatous inflammation composed of neutrophils, eosinophils, lymphocytes and plasma cells (Figure 3). Within this inflammatory granulation tissue there were Langhan's type multinucleated giant cells together with epithelioid cells. In some surface areas there

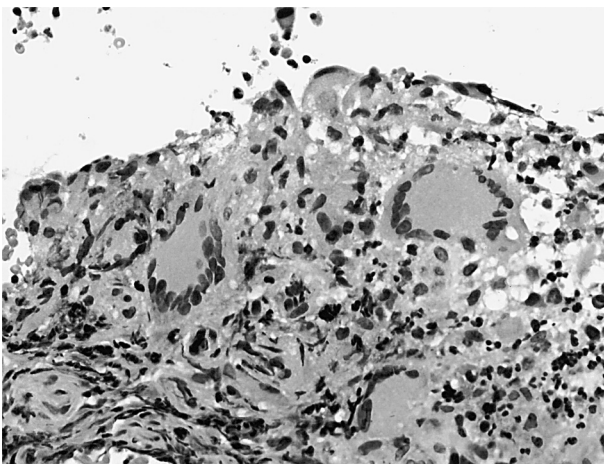


FIG. 3

Photomicrograph of nasal biopsy showing acute on chronic granulomatous inflammation, within which multinucleated giant cells and epithelioid cells may be identified. No vasculitis is evident (H&E, original magnification x100).

was basophilic necrosis. No foreign bodies, fungi or acid-fast bacilli were identified, and no vasculitis was evident.

Topical treatment with corticosteroids and antibiotic ointments led to considerable improvement of his nasal symptoms and the appearance of the nasal cavity. His saddle nose deformity did not progress.

Discussion

From its initial conception as a 'regional enteritis',⁹ Crohn's has expanded to include the entire gastrointestinal tract from the mouth to the anus, and a range of extraintestinal manifestations. These include cutaneous lesions such as erythema nodosum and pyoderma gangrenosum,¹⁰ arthralgias, lung, liver and bone marrow involvement, and virtually every type of eye pathology.¹¹ The most common involvement in the head and neck are oral lesions in the form of oral aphthae and, less frequently, persistent lip swelling, angular cheilitis, and a cobblestone appearance of the oral mucosa.¹² Laryngeal lesions are rarely found too, and include gross oedema and ulceration of the epiglottis and arytenoids.¹³

Extraintestinal manifestations are more prevalent in colonic involvement, and in patients with other manifestations.¹ They usually occur during exacerbations of intestinal disease, but may precede intestinal symptoms by many years.^{3,12,14} Their incidence rises with duration of the disease and has been reported as 33 per cent in one series of 498 Crohn's cases. In contrast, we have reviewed the literature and uncovered only a handful of reports of nasal involvement in Crohn's disease. Table I summarizes these.

In this paper we have described a patient with long-standing Crohn's disease and a saddle nose deformity, which is a malformation that is not usually associated with the disease. We believe that granulomatous inflammation of the nose led to the formation of deep mucosal ulceration and eventually to saddle nose deformity as nasal symptoms of obstruction, discharge and cast production preceded the saddle nose deformity. All but one of the cases reported to date had intestinal Crohn's disease at the time of presentation of nasal symptoms, and all of them had other manifestations as well.^{1,2,5-8} In one case, reported by Ghandour and Issa, nasal symptoms, in the form of crusting of the external nares and therapy resistant hypertrophied right inferior turbinate, developed years after oral manifestation of the disease, but before manifestation of intestinal symptoms.³ All reported cases in which the intestinal symptoms are described exhibit colonic involvement, a feature shown to be associated most frequently with extraintestinal manifestations in previous studies.¹

In view of the paucity of cases in the literature, the diagnosis of Crohn's disease in the nose presents some difficulty. Histopathological evaluation of nasal mucosal specimens may reveal granulomatous inflammatory changes but, as in oral specimens, variable degrees of acute and chronic inflammation may be the only finding. Traditionally, serology plays a minimal role but autoimmunoassay in our patient revealed positivity for the atypical ANCA with PR3 activity and positive ANCA's have been found in up to 28 per cent of Crohn's patients in previous studies, particularly in patients with colonic involvement.¹⁵ However, no single histological or serological finding is diagnostic of the disease.

We also considered the possibility that our patient did, in fact, have Wegener's granulomatosis, given the necrosis evident histologically. Factors mitigating against this were the absence of vasculitis, which normally characterizes the disease, and the rarity of these two uncommon conditions

TABLE I
CHARACTERISTICS OF THE REPORTED CASES OF NASAL MANIFESTATIONS OF CROHN'S DISEASE

Author	Age	Sex	Presenting symptoms	GI findings	Other manifestations	Nasal findings	Treatment and outcome
Kinnear (1985) ²	36	F	Chronic nasal obstruction and post-nasal drip	Anal stricture, quiescent duodenal and colonic involvement	Perineal involvement	Chronic atrophic rhinitis with crusting and hypertrophic tissue in PNS	Topical steroid therapy effective
Ghandour <i>et al.</i> (1991) ³	10	F	Lip swelling, nasal crusting and obstruction	None	Oral ulcers	Crusting of external nares, hypertrophied/ congested right inferior turbinate	No response to topical corticosteroids and sodium cromoglycate. Inferior and middle turbinates trimmed
Ernst <i>et al.</i> (1993) ⁴	17	F	Frontal headache, nasal obstruction and progressive dysphagia	Diarrhoea. Ileocaecal disease diagnosed two years earlier	Arthralgia, pretibial erythema, oral vesicles, perineal disease	Sino-nasal polyposis, severely inflamed and enlarged inferior turbinates	Antibiotics treatment and systemic steroid. Resolution of nasal findings and symptoms within 10 days
Pochon <i>et al.</i> (1995) ⁵	38	M	Nasal obstruction with watery rhinorrhoea and recurrent epistaxis	Diarrhoea	Oral ulcers	Swollen nasal mucosa with diffuse thickening of sinus lining. 1.5 ml of clear fluid evacuated from septum	Oral steroid for one month returned nasal mucosa to normal
Ferjaoui <i>et al.</i> (1999) ⁶	23	F	Purulent rhinorrhoea and saddle nose deformity	Diarrhoea	Nil	Ulcerated nasal mucosa and crusting	Oral corticosteroids and immunosuppressants returned nasal mucosa almost to normal
Kriskovich <i>et al.</i> (2000) ⁷	12	M	Nasal crusting, epistaxis and nasal septal perforation	Severe colitis	Oral ulcers, oesophageal and perineal involvement	1 x 1.5 cm anterior septal perforation with adjacent crusting, and mucosal oedema	Topical and systemic corticosteroids, and topical antibiotic ointments
Ulnick <i>et al.</i> (2001) ¹	45	M	Nasal stenosis and obstruction, intermittent epistaxis	Quiescent	Pyoderma of the scalp. Laryngeal involvement	Nasal stenosis	Laser to nose. Systemic and topical corticosteroids with 6-mercaptopurine achieved limited control
Venail <i>et al.</i> (2002) ⁸	32	M	Nasal obstruction and epistaxis	Previous ileostomy	Nil	Ulcerated nasal mucosa and crusting	Nasal antibiotic lavage
Present study	23	M	Saddle nose deformity, nasal obstruction and cast production on blowing the nose	Active but controlled ileitis and pancolitis	Perineal and eye involvement. Arthralgia	Saddle nose. Inflamed nasal mucosa with deep ulceration of both inferior turbinates and the nasal septum. Marked crusting more so on the right	Topical corticosteroid drops, antibiotic ointments and nasal irrigations with saline effected symptomatic relief

co-existing. In fact, there is only one case in the literature of Crohn's and Wegener's in the same patient.¹⁶

Management of Crohn's disease consists of surgical and medical treatment comprising aminosalicylates, prednisolone and mild cytotoxic drugs such as 6-mercaptopurine and azathioprine.¹⁷ Maintenance therapy after remission is achieved with azathioprine and oral budesonide and, although high-dose oral prednisolone is of proven value in maintaining remission, its use is limited because of systemic side effects. Oral metronidazole for a restricted period of

three months has also been shown to prolong remission.¹⁸ From our experience and our literature review, nasal manifestations, depending on their severity, are best treated with topical corticosteroids in conjunction with conventional systemic therapy. Topical antibiotic preparations may also be of value in more severe cases with mucosal ulceration.

In conclusion, we have highlighted a case of saddle nose deformity in the setting of Crohn's disease. Several features have been discussed that point to the two entities being related and we have reviewed the available literature and

discussed possible diagnostic features and therapeutic interventions.

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- **Crohn's disease has a number of extraintestinal manifestations and in the head and neck these include aphthous ulceration and angular cheilitis**
- **Nasal complications are rare and include obstruction, rhinorrhoea, recurrent epistaxis, paranasal sinus disease and nasal septal perforation**
- **This paper reports the first case in the English literature of saddle nose deformity associated with Crohn's disease and presents a review of the literature on the treatment of this disabling condition**

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