Rosacea fulminans is a rare disorder of unknown cause that mainly affects postadolescent women, with abrupt onset and disfiguring course if left untreated. The simultaneous occurrence of rosacea fulminans and inflammatory bowel disease is rare and has been reported predominantly in the setting of ulcerative colitis. We describe here a case of rosacea fulminans in a patient with Crohn’s disease and discuss a possible association between the two conditions. Key words: rosacea; pyoderma faciale; inflammatory bowel disease.

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Rosacea fulminans, previously called pyoderma faciale, is a disease of postadolescent women, usually without previous acne, which is characterized by the sudden onset of conglobate, nodular lesions on the face. A history of episodic flushing and marked seborrhoea is elicited in most patients. There are only a few articles in the literature on the simultaneous occurrence of rosacea fulminans and inflammatory bowel disease. We recently reported the occurrence of rosacea fulminans in the setting of ulcerative colitis (1). In this report, a woman with both rosacea fulminans and Crohn’s disease is described.

CASE REPORT

In 1991, a 21-year-old woman with recurrent episodes of abdominal pain and diarrhoea was diagnosed with Crohn’s disease on repeated colonoscopy and intestinal biopsies. She had a history of low-grade acne vulgaris during puberty.

In April 1998, she experienced an increase in sebum secretion on her face and shortly thereafter presented with an acute onset of inflammatory papulopustules and nodules on the background of a reddish-eryanotic skin (Fig. 1). The lesions were located mainly in the central part of the face, although a few lesions were also present on the neck and trunk. Purulent material drained spontaneously from abscessing nodules on the cheeks. Marked facial oiliness and oedematous swelling were also evident. Telangiectases, but no comedones, could be observed.

Physically she felt well. There was a history of episodic flushing during the last year. The manifestations of Crohn’s disease had been stationary during the last 8 months and the therapeutic regimen during this period had included azathioprine (50 mg/day), mesalazine (4 g/day), and hydrocortisone (10 mg/day). She had not taken halogen-containing drugs. She had no menstrual abnormalities.

The erythrocyte sedimentation rate was raised (34 mm in the first hour) and there was leukocytosis (14.7/nl). All other laboratory investigations were within normal range. Repeated bacteriological investigation of the papulopustules did not reveal pathogenic organisms. Histopathologic examination of a biopsy from the terminal ileum showed no signs of active inflammation.

A therapeutic approach with dapsone (100 mg/day) and topical fusidic acid cream over a 6-week period showed no improvement. This was followed by treatment with isotretinoin (13-cis-retinoic acid, Roaccutan®) 0.6 mg/kg/day and introduction of an oral contraceptive (2 mg cyproterone acetate and 0.035 mg ethinyl estradiol: Diane®-35) for 3 months without response. Thereafter, the medications for Crohn’s disease were modified: the dosage of hydrocortisone was slowly tapered, mesalazine was withdrawn and the dosage of azathioprine was increased up to 150 mg/day.

After 4 weeks, a marked improvement of the facial lesions could be achieved and the seborrhoea was minimal. During the following months occasional recurrences of the intestinal manifestations were controlled with adjustment of the dosage of azathioprine. Isotretinoin was maintained for 6 months. After healing, no significant scarring was seen. Discrete telangiectatic erythema (Fig. 2) and a tendency to episodic flushing persisted during the next 12 months of follow-up.
DISCUSSION

Pyoderma faciale was originally described by O’Leary & Kierland in 1940 (2). In 1982, Massa & Su (3) described their experience with 29 patients with this condition. Ten years later, our study group reported 20 additional cases (4). We redefined the disease and its nosology and further proposed the term rosacea fulminans, in analogy to the corresponding ferocious process in acne, which has been dubbed acne fulminans.

This disease is neither a pyoderma nor a variant of acne. It is confined principally to the central portions of the face, although extrafacial lesions may rarely be present. It is characterized by a sudden onset of papulopustules and nodules which frequently fuse and form sinus tracts that discharge purulent material (5). In contrast to conventional rosacea, eye involvement (ophthalmic rosacea) is not a feature of the disease. Characteristically, episodes of facial flushing and marked seborrhoea may precede the facial blow-up (5). In contrast with acne fulminans, constitutional symptoms are notably absent.

The simultaneous occurrence of rosacea fulminans and inflammatory bowel disease has been documented only 7 times in the literature (Table I). Six patients had ulcerative colitis (1, 3, 6–8) and 1 had Crohn’s disease (9). This is the second report of rosacea fulminans occurring in the setting of Crohn’s disease. Although the course of rosacea fulminans has been reported to be associated with reactivation of the inflammatory bowel disease (6, 7), there was no direct relationship between the occurrence of facial lesions and intestinal inflammation in our patient.

No significant association between rosacea and gastrointestinal disease has been established so far. A possible relationship has been suggested because of the ease with which rosacea patients flush in response to normal gastric stimuli. Recently, it was suggested that Helicobacter pylori, a spiral-shaped Gram-negative organism living in the gastric mucosa, may be implicated in the pathogenesis of rosacea, possibly by raising gastrin levels that may stimulate flushing (10). Walton et al. (11) described 4 patients with a combination of rosacea and ulcerative colitis. In all 4, ulcerative colitis preceded the onset of severe papulopustular rosacea. In 1, the severity and poor initial response of rosacea to treatment was related to the activity of the ulcerative colitis, and the rosacea improved only after proctocolectomy. The authors suggested that the severity of rosacea could have been due to the associated inflammatory bowel disease.

As in many other conditions of unknown aetiology, immunological factors have been implicated in the pathogenesis of both inflammatory bowel disease and rosacea. There is some evidence that the expression of immune responses, directed against the gut itself or against associated antigens such as enteric bacteria, is responsible for generating inflammation in Crohn’s disease and ulcerative colitis (12, 13). It is likely that immune mechanisms also play a role in the pathogenesis of rosacea. Some investigators have demonstrated anergy to dinitrochlorobenzene sensitization and deposition of immunoglobulins at the dermo-epidermal

Table I. Simultaneous occurrence of rosacea fulminans and inflammatory bowel disease

<table>
<thead>
<tr>
<th>Authors</th>
<th>Age (years/sex)</th>
<th>Inflammatory bowel disease</th>
<th>Extraintestinal manifestations</th>
<th>Medication</th>
<th>Correlation</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Massa &amp; Su (3)</td>
<td>?/?</td>
<td>Ulcerative colitis</td>
<td></td>
<td>Sulfasalazine</td>
<td>?</td>
<td>1982</td>
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<tr>
<td>Sigl &amp; Bauerdorf (6)</td>
<td>33/f</td>
<td>Ulcerative colitis</td>
<td></td>
<td>Sulfasalazine</td>
<td>+</td>
<td>1989</td>
</tr>
<tr>
<td>McHenry et al. (9)</td>
<td>31/f</td>
<td>Crohn’s disease</td>
<td>Erythema nodosum</td>
<td>Mesalazine, sulfasalazine</td>
<td>+</td>
<td>1992</td>
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<tr>
<td>Schmitz &amp; Zouboulis (7)</td>
<td>51/m</td>
<td>Ulcerative colitis</td>
<td></td>
<td>Mesalazine, corticosteroids</td>
<td>+</td>
<td>1995</td>
</tr>
<tr>
<td>Dessoukey et al. (8)</td>
<td>31/w</td>
<td>Ulcerative colitis</td>
<td>Erythema nodosum, arthralgias</td>
<td>Sulfasalazine, prednisolone</td>
<td>?</td>
<td>1996</td>
</tr>
<tr>
<td>Jansen &amp; Plewig (1)</td>
<td>28/m</td>
<td>Ulcerative colitis</td>
<td></td>
<td>Sulfasalazine</td>
<td></td>
<td>1997</td>
</tr>
<tr>
<td>Present case</td>
<td>21/f</td>
<td>Crohn’s disease</td>
<td></td>
<td>Mesalazine, hydrocortisone</td>
<td></td>
<td>2000</td>
</tr>
</tbody>
</table>

Fig. 2. Discrete telangiectatic erythema on the face after 6 months of therapy.
junction, as seen in lupus erythematosus (14). The inflammatory cell infiltrate in rosacea has some of the characteristics of that seen in hypersensitivity reactions (15). Both anticollagen and antinuclear antibodies have been reported to occur in lymphocyte eluates of rosacea patients (16). Manna et al. (14) reported the presence of circulating antinuclear antibodies of IgM class as well as a positive association with autoimmune disorders and signs of immunodeficiency in rosacea patients.

Rosacea fulminans generally responds to a therapeutic regimen, including both isotretinoin and corticosteroids (17). Although a favourable response to dapsone has been reported in a patient with rosacea fulminans (18); our patient did not improve with this medication.

REFERENCES