CASE REPORT

Subcorneal pustular dermatosis and episcleritis associated with poorly controlled ulcerative colitis

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SUMMARY

A man aged 56 years with a history of ulcerative colitis (UC) status postsubtotal colectomy was hospitalised with fevers, dry cough, eye redness and a new bloody, mucoid rectal discharge. 2 months prior to admission, the dermatologist had started him on dapsons for subcorneal pustular dermatosis but did not recognise that he had recently self-discontinued mesalamine enemas, inducing a flare of his UC. After a thorough inpatient evaluation, including flexible sigmoidoscopy, active UC involving the rectal stump was determined to be driving his dermatological and ophthalmological findings. By reinstituting mesalamine enemas, control of his UC was achieved and the extraintestinal manifestations of his inflammatory bowel disease (IBD) resolved. This case illustrates the importance of careful history taking and of early recognition of extraintestinal manifestations of IBD in order to appropriately target treatment and prevent unnecessary morbidity.

BACKGROUND

A new extraintestinal diagnosis in an inflammatory bowel disease (IBD) patient may not represent a primary disorder and should prompt consideration for interplay between the new disease process and active IBD. If prompt correlation between an extraintestinal manifestation and active colonic inflammation goes unrecognised, this may lead to substantial ongoing colonic inflammation and additional extraintestinal manifestations of IBD. This reminder is important as roughly 6–40% of patients with IBD develop extraintestinal manifestations of their disease.^{1 2} Up to 15% of IBD patients develop cutaneous manifestations, whereas 4–10% develop ocular manifestations, which are the systems we focus on in this case.³ While a cutaneous or ocular illness may typically manifest as a primary disorder, careful history taking and understanding of extraintestinal manifestations of IBD remain imperative. Recognising conditions that

Figure 1 Flaccid vesicles, vesicopustules and erythematous erosions on his trunk and legs.

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Figure 2 Bright red episcleral discolouration.

parallel IBD activity early in their course, confirming compliance with a patient's IBD medication regimen and prompt management of active colonic inflammation may result in reduced morbidity from IBD activity and/or subsequent extraintestinal manifestations.

CASE PRESENTATION

A man aged 56 years presented to the hospital with 1 week of fever, dry cough and eye redness. Two months prior to presentation, he had developed flaccid vesicles, vesicopustules and erythematous erosions on his trunk and legs (figure 1), for which he was evaluated by dermatology, diagnosed with subcorneal pustular dermatosis (Sneddon-Wilkinson disease), and treated with dapsone. Relevant medical history included ulcerative colitis (UC) status postsubtotal colectomy 7 years earlier, resulting in a 10 cm rectal stump and a proximal ileostomy. He had been prescribed mesalamine enemas, but further evaluation revealed that he had self-discontinued these 4 months earlier due to retention difficulties. Subsequently, he developed a mild, intermittent, bloody, mucoid rectal discharge not appreciated at the time of his dermatology evaluation.

INVESTIGATIONS

On admission, vital signs revealed a temperature of 37.4°C (99.3°F), respiratory rate of 14 breaths/min, pulse of 85 bpm, blood pressure of 109/61 mm Hg and oxygen saturation of 97% on room air. He appeared to be in no acute distress. Ocular examination revealed bilateral bright red episcleral discolouration, non-tender to palpation (figure 2). Lungs were clear to

auscultation bilaterally. The abdomen was non-tender with a healthy-appearing right lower quadrant ostomy. Areas of post

inflammatory hyperpigmentation were visualised on his trunk and legs. Rectal examination revealed the presence of a rectal stricture. Laboratory analysis was significant for a white cell count of 9.1 (4–10) K/ μ L, haemoglobin 11.1 (13.5–17.0) g/dL, haptoglobin 65 (22–239) mg/dL, lactate dehydrogenase 422 (120–240) IU/L, C reactive protein 12.2 (0–0.6) mg/dL and Westergren erythrocyte sedimentation rate 70 (0–15) mm/hour. His renal function and liver function tests were within normal limits. Chest radiograph revealed patchy left lower airspace opacity. Nasopharyngeal PCR was negative for influenza and respiratory syncytial virus. He had a normal white cell count and haptoglobin, excluding dapsone-related agranulocytosis and haemolytic anaemia. There was no evidence of bacterial infection, and patient's fever and cough were felt to represent a viral upper respiratory tract infection. The patient was diagnosed with episcleritis by ophthalmology after his episcleral erythema blanched with phenylephrine drops. A flexible sigmoidoscopy was performed, revealing a rectal stricture that was dilated, revealing friable sigmoid mucosa consistent with active UC (figures 3 and 4).

OUTCOME AND FOLLOW-UP

Poorly controlled UC involving the rectal stump was determined to be driving our patient's dermatological and ophthalmological findings. Mesalamine enemas were reinstituted for treatment of his active UC. After our patient resumed mesalamine enemas, control of his UC was achieved within 1 month, and a follow-up flexible sigmoidoscopy revealed resolution of his active bowel disease (figure 5). His subcorneal pustular dermatitis continues to be treated with daily dapsone, and his episcleritis resolved with a combination of topical petroleum and ketorolac therapy tapered over several weeks (figure 6).

DISCUSSION

We will discuss the most common cutaneous and ocular manifestations of IBD and provide a brief review on subcorneal pustular dermatosis. The most common cutaneous manifestation, erythema nodosum, is seen in 3–10% of UC patients and 4–15% of Crohn's patients. Pyoderma gangrenosum, the second most common cause, occurs in only ~1% of IBD patients.⁴ Erythema nodosum and neutrophilic dermatoses (such as Sweet Syndrome and subcorneal pustular dermatosis) parallel IBD activity, whereas pyoderma gangrenosum does not. Episcleritis is the most common ocular manifestation of IBD, occurring in 2–5% of patients and is not

Figure 3 Rectal stricture at 5 cm proximal to the anus pre and postdilation.

Figure 4 Normal sigmoid (left) versus friable sigmoid (right) represented by arrows.

Figure 5 Healthy appearing sigmoid colon after resuming mesalamine enema therapy.

Figure 6 Healthy appearing episclera after treatment.

associated with vision loss.⁵ Episcleritis typically presents as abrupt onset of painless redness, irritation and eye watering. The diagnosis is confirmed on ocular examination. Blanching with phenylephrine drops helps confirm the presence of episcleritis and rule out scleritis (conjunctival and episcleral vessels blanch with phenylephrine drops, which are unable to penetrate deeper scleral vessels). Scleritis and uveitis are less commonly associated with IBD than episcleritis. They do not parallel IBD activity, and are associated with vision loss. Urgent ophthalmological evaluation is indicated if there is concern for either condition. Scleritis typically presents as a severe, constant boring pain that is worse at night or the morning often causing patients to have difficulty with sleep.

Uveitis presents with reduction in visual acuity and is associated with a ciliary flush on examination. Subcorneal pustular dermatosis typically presents as recurrent crops of sterile pustules in flexural surfaces and skin folds, and generally affects women. This condition may be associated with paraproteinemias (IgA monoclonal gammopathy), haematological cancers, solid tumours, IBD or rheumatological disease.⁶ Diagnosis is based on clinical history, physical examination and confirmed with a positive skin biopsy (including histopathology and culture to exclude infectious aetiologies) revealing subcorneal vesicles or pustules comprised of mainly neutrophils with occasional eosinophils. First-line treatment is dapsone, though more aggressive immunosuppression may be required for treatment of refractory or relapsing disease.⁷ All patients prescribed dapsone must be screened for glucose-6-phosphate dehydrogenase (G6PD) deficiency to avoid profound haemolytic anaemia. The side-effect profile of dapsone requires frequent examinations and laboratory monitoring (including complete blood count and comprehensive metabolic panel) to avoid the risk of cutaneous side effects, including erythema multiforme, erythema nodosum and Stevens-Johnson syndrome, and other complications, including abdominal pain, pancreatitis, aplastic anaemia, agranulocytosis, haemolysis and rare risks of cholestatic jaundice/toxic hepatitis, suicidal ideation and reversible peripheral

Learning points

- ▶ Extraintestinal manifestations of inflammatory bowel disease (IBD) are common, and can be associated with unnecessary morbidity if not recognised early.
- ▶ Erythema nodosum, neutrophilic dermatoses (most commonly Sweet syndrome) and episcleritis parallel IBD activity, while pyoderma gangrenosum, scleritis and uveitis do not.
- ▶ Scleritis and uveitis are associated with vision loss (unlike episcleritis), and require prompt ophthalmology evaluation.
- ▶ A diagnosis of subcorneal pustular dermatosis (Sneddon-Wilkinson disease) should prompt a clinician to search for a secondary disorder as this condition is frequently associated with paraproteinaemias, haematological cancers, solid tumours, IBD and other rheumatological diseases.
- ▶ All patients prescribed dapsone must be screened for glucose-6-phosphate dehydrogenase (G6PD) deficiency to avoid profound haemolytic anaemia.

motor neuropathy. Were dapsone therapy to fail or should our patient have experienced adverse reactions to dapsone, phototherapy or systemic corticosteroids would be considered as second-line therapy. Topical corticosteroids were also used in our case as an adjunct and for symptom management. Since this is a relapsing and chronic condition, more aggressive therapies may be indicated in the future. Biological agents such as infliximab, adalimumab and etanercept, or oral immunosuppressant medications such as ciclosporin, acitretin, colchicine and mycophenolate have been observed in case reports to offer successful treatment options.^{8–13}

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