CASE REPORT

Florid urticarial vasculitis heralding a flare up of ulcerative colitis

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SUMMARY

A 75-year-old man with ulcerative colitis (UC) and diet controlled diabetes mellitus presented with a 3-week history of slightly itchy, red plaques on both lower limbs ascending gradually to cover the trunk and arms. One week later, he developed a flare up of his UC. Routine blood tests showed modest drop in haemoglobin (122 g/L) and C reactive protein (85 mg/L). Serology was remarkable for high antiproteinase 3 (c-ANCA). Serum electrophoresis showed a mildly positive paraprotein band (γ region). Stool culture was negative. Urine analysis showed proteinuria. Skin biopsy showed features of urticarial vasculitis (UV). He underwent a flexible sigmoidoscopy after the flare up showed mildly active UC. The patient was given hydrocortisone for 7 days and then prednisolone. Both rash and UC subsided. Electrophoresis was repeated 4 weeks later showing normal pattern. Prednisolone has been gradually reduced. Although rare, UV can be considered as one of the skin manifestations of UC.

BACKGROUND

Ulcerative colitis (UC) is characterised by recurring episodes of inflammation limited to the mucosal layer of the colon and rectum. Approximately 42% of patients with UC have extraintestinal complications,¹ the skin being one of the most frequently affected organs. The two most common skin disorders associated with UC, erythema nodosum and pyoderma gangrenosum, appear to be reactive response that are not specific to inflammatory bowel disease.² Although rare, urticarial vasculitis (UV) should be considered as one of the skin manifestations of UC and in some cases it actually preceded the intestinal symptoms.

CASE PRESENTATION

A 75-year-old man with ulcerative colitis, on mesalazine 2400 mg daily and diet-controlled diabetes mellitus (non-insulin-dependent diabetes mellitus), presented with a 3-week history of rash (figure 1) on both lower limbs associated with bilateral knee swelling. The rash started as mildly itchy, sore, raised red plaques on his lower limbs and then started to ascend gradually up his body, covering the trunk and arms.

One week after the appearance of the rash, he had a flare up of his UC with diarrhoea, more than 10 times per day, abdominal pain and rectal bleeding.

INVESTIGATIONS

Routine blood tests at presentation showed normal white cell count (WCC) including neutrophils,

Figure 1 Rash at presentation 3 weeks after initial onset. There are urticated plaques that were initially erythematous evolving into purpuric areas particularly centrally.

haemoglobin (Hb) 122 g/L, C reactive protein 85 mg/L, liver function test normal but with a slightly low albumin level 32 g/L (range 35–50 g/L), and normal renal function. Serology was remarkable for high antiproteinase 3 (anti-PR3 ANCA=11.3), positive antinuclear antibodies (1/320) and negative anti-double stranded DNA antibodies. Serum electrophoresis showed a faintly positive paraprotein band (γ region). Stool culture was negative. Urine analysis showed proteinuria (urine protein 0.24 g/L).

A skin biopsy was performed demonstrating normal epidermis with a patchy inflammatory infiltrate focused on upper to mid-dermis predominantly associated with vessels and also seen around the skin appendages (figure 2A, B).

There was no evidence of dysplasia or malignancy. Features present in the biopsy were more in keeping with necrotising vasculitis, for example, leucocytoclastic vasculitis.

DIFFERENTIAL DIAGNOSIS

- UV
- Borrelia

CrossMark

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Figure 2 (A and B) Skin biopsy: inflammatory infiltrate composed of neutrophils with some eosinophils. Vessels showing numerous neutrophils within their walls associated with swelling of endothelial cells and focal fibrinoid necrosis. There is red blood cells extravasation.

TREATMENT

The patient was initially started on doxycycline for possible Lyme disease but this was stopped when borreliosis was excluded by serology. At the same time the patient was given hydrocortisone 100 mg intravenously three times daily for 7 days and then prednisolone 40 mg daily.

OUTCOME AND FOLLOW-UP

Both the rash and UC subsided on corticosteroids. The rash had started to resolve within 4 days of starting the steroids and the inflammatory components were completely clear after 10 days.

The patient underwent flexible sigmoidoscopy after the flare up; this showed patchy superficial colitis throughout without ulceration, representing UC attenuated by the steroid therapy. Colonic biopsies demonstrated normal to mildly disturbed architecture with mucin cell depletion and acute or chronic inflammatory infiltrate, including focal cryptitis. This was consistent with mildly active UC (figure 3A, B).

Electrophoresis was repeated 4 weeks after presentation showed a normal pattern. The association between UV and IgG monoclonal gammopathy has previously been reported^{3 4} and in our patient this was transient.

Prednisolone had been reduced by 5 mg every week for the monitoring of UC symptoms and patient returned back to his regular medications (mesalazine 2400 mg).

DISCUSSION

UV is an eruption of erythematous wheals that clinically resemble urticaria but histologically show changes of leucocytoclastic vasculitis.^{5 6} The cutaneous features may include painful, burning or pruritic skin lesions. The persistence of individual

Figure 3 (A and B) Colon biopsy: normal to mildly disturbed architecture with mucin cell depletion and acute on chronic inflammatory infiltrate including focal cryptitis. This was consistent with mildly active ulcerative colitis.

lesions for longer than 24 h is unlike ordinary urticaria. There is palpable purpura, pronounced central clearing of lesions and residual hyperpigmentation following resolution. However, because clinical characteristics of UV may overlap with those of allergic urticaria, confirmation of the diagnosis requires a lesional skin biopsy.⁷

UV is idiopathic in up to 70% of cases,⁸ but it has been associated with autoimmune diseases, such as Sjögren syndrome and systemic lupus erythematosus, immunoglobulin disorders and leukaemia. Certain viral diseases, including hepatitis B and hepatitis C, and infectious mononucleosis are also associated with this type of vasculitis. UV can also be secondary to some medications, particularly antibiotics and non-steroidal anti-inflammatories.⁸ The association of a neutrophilic UV with an IgG λ paraprotein has been described before in the context of Schnitzler syndrome where there is also a significant risk of haematological malignancy.⁴ In our case, the paraprotein band was mild and transient, a recognised occasional feature of some autoimmune disorders, mycobacterial infections and some skin diseases, including neutrophilic dermatoses.⁹ It can be speculated that it results from a response directed against either an exogenous antigen, for example, microbial agent, or an autogenous antigen uncovered by the acute inflammatory process rather than having a pathogenic role per se.

There are case reports of UV in association with UC. Swaminath *et al* reported a case of a man with a history of UC, complicated by end-stage liver disease secondary to sclerosing cholangitis, who developed vasculitis and diarrhoea refractory

from therapy with standard immunosuppression. He was successfully treated with rituximab.¹⁰

There are reports of leucocytoclastic vasculitis without urticaria. A 22-year-old man presented with a 2-year history of the lower limb rash. A skin biopsy confirmed leucocytoclastic vasculitis which did not respond to cyclosporin (3 mg/kg/day) and oral steroids. A year and half later, the patient presented with symptoms of abdominal pain and diarrhoea with rectal bleeding. An endoscopic evaluation and biopsy led to a diagnosis of UV. His UC and vasculitis responded to oral sulfasalazine (3 g/day) and rectal steroids.¹¹ Two other patients with cutaneous leucocytoclastic vasculitis (but without urticaria) preceding intestinal symptoms had been reported. In these patients, skin lesions appeared 1 and 5 months before UV, respectively.¹²

Learning points

- ▶ Urticarial vasculitis can be considered as one of the skin manifestations of ulcerative colitis (UC).
- ▶ Urticarial vasculitis can precede intestinal symptoms of UC.
- ▶ Urticarial vasculitis in association with UC can be treated by corticosteroids.

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Competing interests None.

Patient consent Obtained.

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