Sweet's syndrome: a rare extraintestinal manifestation of ulcerative colitis

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DESCRIPTION

A 50-year-old man presented with mild proctitis, biopsies of which indicated chronic inflammatory bowel disease. Despite mesalazine suppositories, he progressed to severe colitis affecting the descending colon to the rectum. He proved refractory to steroids and was started on azathioprine and infliximab; however, he failed to improve and underwent a subtotal colectomy with ileostomy 3 months after initial presentation. Pathology showed active, chronic inflammation present from the caecum distally with cryptitis, crypt abscesses and extensive mucosal ulceration down to and in some areas through the muscularis propria. Typical features of Crohn's disease such as granulomas were absent, and the condition was thought more in keeping with ulcerative colitis. The patient was readmitted 2 months later with an exacerbation of his colitis affecting his rectal stump and fever. An elevated neutrophil count and marked extraintestinal manifestations were noted including multiple tender skin papules (figure 1) affecting his trunk, face and extremities, mouth ulcers, arthralgia and uveitis (figure 2). Biopsies of the skin lesions demonstrated primarily an infiltration of mature neutrophils, which, in conjunction with his clinical presentation, was indicative of Sweet's syndrome. Human leucocyte antigen (HLA) typing revealed the patient was homozygous for HLA-A, HLA-B and HLA-C, specifically indicating presence of HLA-A*11, HLA-B*35, HLA-C*04, HLA-DRB1*01, HLA-DRB4*01, HLA-DQB1*03. To the authors' knowledge, this HLA type has not previously been associated with Sweet's syndrome. The patient responded well to an intensification of his immunosuppression, with intravenous hydrocortisone, mesalazine and azathioprine, and he was again discharged home with a view to elective completion proctectomy.

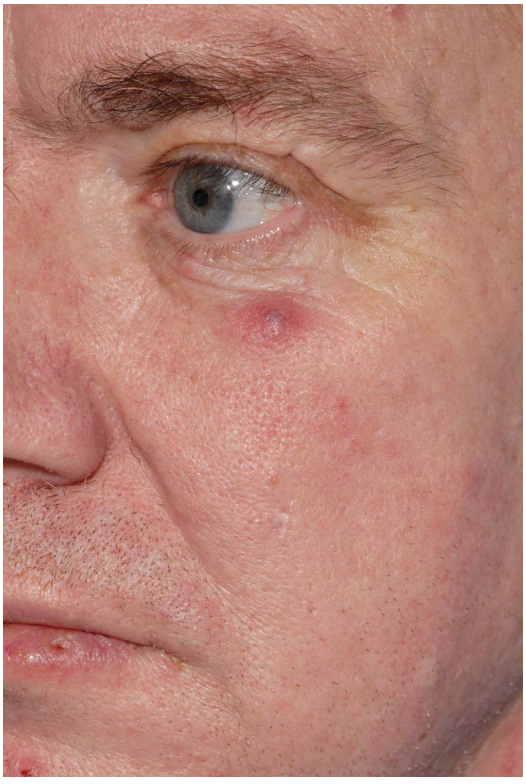

Figure 1 Facial skin papules, biopsy of which demonstrating Sweet's syndrome.

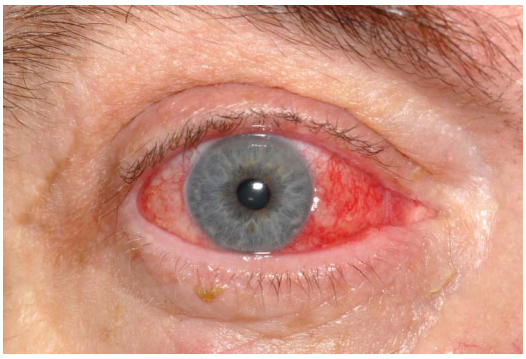

Figure 2 Ocular manifestation indicative of uveitis.

Learning points

- Extraintestinal manifestations (EIM) of inflammatory bowel disease (IBD) are common, affecting 25–40% of patients with IBD.[1]
- The precise aetiology underlying the association between IBD and EIM remains unclear, with many advocating that IBD is a systemic autoimmune disease with the most marked symptoms apparent in the intestine.[2]
- The most commonly affected organs outside of the gastrointestinal tract are musculoskeletal, ocular and dermatological; however, almost every other system can be affected, including the hepatobiliary, genitourinary, vascular, cardiac, pulmonary and endocrine.[3]

Contributors RS and JH were responsible for the diagnosis and management of this patient. RS coordinated the photography and coauthored the case report with JH.

Competing interests None declared.

Patient consent Obtained.

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REFERENCES

1 Orchard T. Extraintestinal complications of inflammatory bowel disease. *Curr Gastroenterol Rep* 2003;5:512–17.
2 Danese S, Semeraro S, Papa A. Extraintestinal manifestations in inflammatory bowel disease. *World J Gastroenterol* 2005;11:7227–36.
3 Baumgart DC, Sandborn WJ. Inflammatory bowel disease: clinical aspects and established and evolving therapies. *Lancet* 2007;369:1641–57.