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Behcet's Syndrome resembling complex perianal Crohn's Disease

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Abstract

Behcet's syndrome is a systemic inflammatory disorder that involves several organ systems and is exceptionally rare in the Western world. The diagnosis is frequently difficult as it resembles several other disease processes. A 23-year-old male with a previous presumptive diagnosis of Crohn's disease presented to our unit with genital ulceration. This is on a background of recurrent perianal abscesses requiring surgical drainage and seton placement. He subsequently developed a complex perianal fistula extending from the rectum to the perineum and left groin. After drainage and an unsuccessful trial of biologic immunosuppressive therapy, he developed several papulopustular cutaneous lesions and oral ulcerations. The diagnostic criteria for Behcet's syndrome was met and he was referred to a rheumatologist for ongoing management.

Keywords

Behcet syndrome, Crohn's disease, rectal fistula, anus diseases, abscess

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Introduction

Behcet's syndrome (BS) is a rare systemic inflammatory condition with multiorgan involvement. Its estimated prevalence is less than 1 to 2 per million in the Western world, though it is more commonly encountered in parts of the Middle-East and East Asia.[1] Typically, it is characterised by the presence of recurring orogenital ulceration. However, we now know there is a spectrum of presentations reflecting the underlying organ involvement.[2] This together with the absence of a hallmark diagnostic test has meant that the diagnosis is frequently difficult. We present a case that further adds to this complexity, whereby the condition resembled perianal Crohn's disease requiring recurrent surgical management. Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Case

A 23-year-old male from Shanghai presented with a 6-month history of worsening painful perineal ulcers. This is on a background of Crohn's disease (CD) diagnosed 12 months earlier on endoscopy at an institution overseas. Although he was prescribed mesalazine at the time of diagnosis, he was frequently non-compliant with therapy. A predominant feature of his Crohn's was the presence of recurrent perianal abscesses and fistulas requiring drainage and seton placement. The patient was otherwise a non-smoker, denied excess alcohol consumption and had no known allergies.

On examination, there were multiple clean based ulcerations ranging from 2 to 5 mm in the perineum (Figure 1). Keloid scars marking areas of previous ulceration were also evident. There was however no adjoining induration to suggest a perianal fistula at this instance. Biopsy of the lesions demonstrated inflammatory infiltrate but no cell atypia. Tissue cultures for bacteria, acid fast bacilli and fungi were negative. Serological markers for HIV and Syphillis were within normal limits. In light of the stated history of CD and the appearance of the ulcers, the working diagnosis at the time was pyoderma gangrenosum. After the period of investigation, the patient noted some improvement in his perineal pain without intervention and elected to continue with expectant management.

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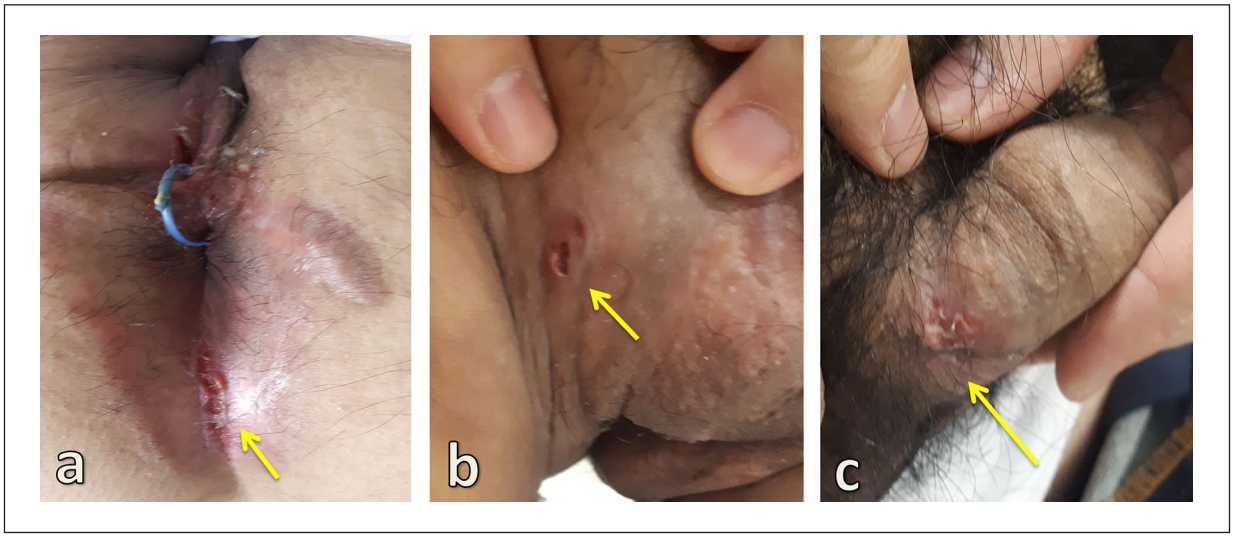

Figure 1. (a) Photograph of the perineum with the patient positioned in lithotomy. Clean based ulceration in the natal cleft without any evidence of underlying fluctuance to suggest a pilonidal abscess (arrow). Note the adjacent seton facilitating drainage of a previous perianal fistula. Note also a keloid scar formed from a previous ulcer site at the 3 o'clock position – there is a propensity for ulcers to leave residual scars in Behcet's syndrome. (b) A 3 × 2 mm ulcer near the junction of the right hemiscrotum and medial thigh, again with no evidence of underlying fluctuance. (c) Ulceration with adjacent induration at the lateral end of the proximal shaft of the penis.

Three months later, he presented to our unit with a left groin abscess that spontaneously ruptured. Examination revealed a punctum inferomedial to the left inguinal ligament with associated purulent discharge. An MRI of the pelvis was organised to establish whether there was any evidence of atypical fistulising disease associated with Crohn's. On T2-weighted imaging, there was evidence of a tract adjoining the left groin to the perineum, along with a medial branch extending towards the rectum (Figure 2). The tract was drained via the perineal opening and a seton inserted to allow drainage and healing of the complex fistula.

In the coming months, he began to experience intermittent PR bleeding and lower abdominal pain despite compliance with mesalazine. He was commenced on infliximab for a presumed flare of CD at a dose of 5 mg/kg. There was an improvement in his gastrointestinal symptoms and incidentally in the appearance of the ulcers in the coming days. Prior to his second dose of infliximab at the 2-week interval, ulcerations in the perineal region recurred. At this time, he also developed papulopustular lesions over his arms and inner thighs alongside oral ulcerations. He was referred to a rheumatologist with investigations revealing HLA B51 and HLA B27 positivity. The diagnostic criteria for BS was subsequently met and a plan for tailored therapy was initiated.

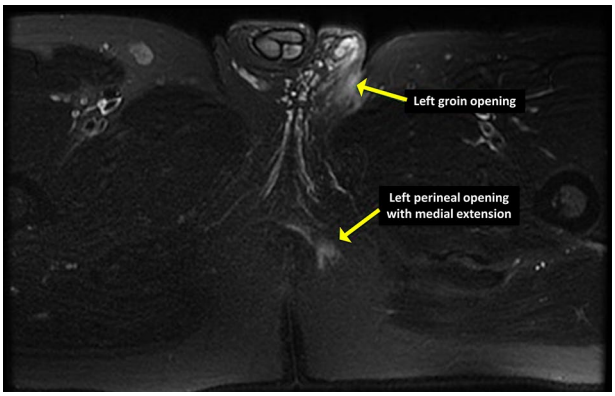

Figure 2. T2-weighted fat-suppressed MRI sequence in the coronal plane demonstrating a fistulous tract between openings in the left groin and the perineum (arrows). The perineal end of the tract demonstrates a component extending medially towards the rectum.

Discussion

BS was first described in 1937 in Turkey as a condition with hallmark features of orogenital aphthous ulceration and ocular inflammation. Since then, it has become increasingly apparent it is a multisystem disorder affecting the mucocutaneous, ocular, gastrointestinal, musculoskeletal, pulmonary and neurological systems.[3] BS is several fold more prevalent in regions along the 'Old silk road' which is consistent with our patient's demographic.[2] Unfortunately, no definitive serological markers or cardinal histological features exist for the condition. Diagnosis of BS is made using clinical criteria in the revised International Criteria for Behcet's Disease (ICBD); although the system has its limitations owing to the heterogeneity of the syndrome.[2,4]

The aetiology of BS remains incompletely understood, though our current evidence suggests it is an interplay between genetic, immunological and environmental factors.

HLA-B51 is the most common genetic risk factor and its presence in this case lent weight to the final diagnosis.[5] Histologically, it is characterised by the presence of a lymphocyte-mediated vasculitis, though these features are not exclusive to BS.[3] Furthermore, these changes more commonly present in deeper layers of involved tissue that may not accessible with superficial biopsies.[6]

There are several reports in the literature on the similarities between CD and BS and our case adds further evidence to this frequent diagnostic dilemma.[6–9] Specifically, the term 'Intestinal Behcet' was coined as gastrointestinal involvement is encountered in up to 40% of cases of BS in East Asia.[7] Symptoms of intermittent lower abdominal pain, bloody diarrhoea, fevers and the endoscopic finding of ileocolic ulceration are pathognomonic for both disease processes. Moreover, oromucosal ulceration and cutaneous lesions may represent the extraintestinal manifestations of CD or side-effects of therapy.[8] Despite the clinical similarities, the two are considered as separate entities each with its own natural course and treatment options.[2,8]

A number of studies have attempted to explore the differentiating factors and devise algorithms to distinguish between CD and BS. In BS, gastrointestinal ulcers on endoscopy are more commonly round and focal as opposed to the longitudinal ulcers segmentally distributed in CD.[9,10] Histologically, CD is characterised by non-caseating granulomas compared with the venulitis in BS; although as mentioned – these features may not be evident on superficial biopsies.[6] The case we describe is unusual in that oral ulceration occurred more than 18 months after the initial onset of symptoms, whereas it usually manifests early in BS.[8] Furthermore, the presence of perianal fistulizing disease is strongly suggestive of CD and its appearance in BS is exceptionally rare (<1% of cases).[9,11]

The treatment options in BS reflect the underlying organ involvement, although there is a notable overlap with CD which further obscures the distinction between the two entities.[8] Anti-TNF-α therapy has been used in intestinal Behcet, which may explain our patient's partial response to therapy.[12] Thus, in cases of isolated intestinal BS, differentiating from CD may not be obligatory as the therapeutic options are concordant. Topical corticosteroids are used in the preliminary stages of cutaneous outbreaks.[13] Anti-interleukin therapy appears to show promise as a multisystem target, although there is a lack of quality randomised-control data to support their routine use.[14] Further studies are needed to better characterise the condition's pathophysiology and appraise these emerging treatment options.

Conclusion

We hope to present this case as a gentle reminder to consider alternate diagnoses in atypical cases that present to our units. BS in particular can mimic several other disease processes as it manifests with non-specific findings in several organ systems. The constellation of findings put together ultimately leads to the diagnosis. When faced with a case of refractory CD in the right demographic group, the diagnosis of BS should be considered.

Declaration of conflicting interests

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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