Vulval Crohn's Disease: A Case Series

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Introduction

Up to 44% of patients with gastrointestinal Crohn's disease (GCD) develop cutaneous symptoms. Rarely this may present as vulvar Crohn's disease (VCD), with only 130 cases reported in the literature.

Signs of VCD vary greatly between patients but classically it presents with labial oedema and knife-like ulcerations. This may result from a direct extension of gastrointestinal involvement or be non-contiguous. In these "metastatic" cases, 25% will precede a diagnosis of GCD.

Here we present two cases of VCD in two 35 year-old women.

Patient One: Case History

The first patient was diagnosed with GCD at the age of 16, affecting the mouth, distal ileum and colon. Her case proved difficult to treat with multiple relapses despite the use of medical therapies including steroids, azathioprine (AZA) and anti-TNF agent infliximab.

Month 1 Although she initially responded well to treatments with her Harvey Bradshaw score improving from 9 to 0, she developed an ileocaecal stricture requiring bowel resection.

Month 2 Her case was complicated by iatrogenic adrenal insufficiency and neutropaenic sepsis for which her AZA and infliximab were suspended.

Month 4 Her medications were recommenced on resolution of her sepsis. However this didn't achieve remission and suffered further Crohn's relapses, requiring a hemicolectomy. Following surgery, she redeveloped active disease. Due to the risk of early stricturing, adalimumab was started and she responded well to this.

Month 6 Adalumimab was later stopped as she developed a reaction to the medication. Her disease progressed, developing perianal fistulae and significant weight loss due to significant pain. Methotrexate and ustekinumab were later commenced but this was not beneficial.

Month 9 Further small bowel resection and ileostomy were complicated by sepsis and ITU admission from which she recovered. Both surgical and medical management had been exhausted. Subsequently she was referred for consideration of trial medications at a quaternary centre.

Alongside this she developed VCD postnatally 6-7 years following her diagnosis of GCD. Two biopsies were inconclusive before a third biopsy showed non-necrotising epithelioid granulomas with multinucleate giant cells, in keeping with VCD. Examination revealed extensive oedema of the labia majora and labia minora with knife-cut like fissures in the inter-labial sulci and natal cleft.

She was commenced on topical steroids and emollients, dermol 500 as a soap substitute and 5% lidnocaine as an adjust to her GCD therapies. Later she was referred to a quaternary dermatology centre where she was commenced on a 6 month course of lymecycline and topical fucibet.

Patient Two: Case History

A 35 year-old woman, with no past medical history but with a family history of inflammatory bowel disease, presented to gynaecology clinic with new vulval lesions after the birth of her first child.

Examination revealed significantly thickened vulval wall skin, labia majora oedema and deep symmetrical knife-cut like transverse fissuring affecting the labia majora. She had active cheilitis of her lips, although buccal mucosa was unaffected, and inflammation extending from the perineum to the natal cleft.

Histopathology showed non-caseating granulomatous inflammation with associated necrosis and abundant Langhan's type giant cells consistent with VCD.

She was referred to dermatology where a diagnosis of VCD was made. Topical and oral steroids were commenced and she was referred to gastroenterology for PR bleeding and diarrhoea. Steroids began to fail and she was commenced on AZA.

After presenting acutely with perianal pain to gastroenterology, investigations confirmed GCD and she was treated with oral ciprofloxacin for a suspected perianal abscess. MRI pelvis showed multiple intersphincteric fistulae but no abscess formation. AZA was stopped and examination under anaesthetic (EUA) confirmed multiple fistulae which were treated with seton insertions.

AZA was recommenced however medical management began to fail and the patient had ongoing rectal bleeding with severe pain and swelling affecting the vulva and anus. An end colectomy with a permanent stoma was offered to the patient however she declined this on the grounds of the risk of treatment failure.

Later she developed perianal sepsis, AZA was suspended and she was treated with antibiotics. On resolution, she was commenced on Humira. She had a short lived improvement on this. Repeat EUA showed that her entire vulva was replaced by inflamed indurated necrotizing multi-sinus lesions, a fenestrated right labia and entire clitoris involvement.

Her disease led to significant weight loss and difficulty mobilising due to severe pain. Humira was stopped, ustekinumab and later mercaptopurine were started alongside intra-lesional steroid injections into the vulva, with a plan to perform a defunctioning colostomy if this failed. This led to marked improvement in her vulval symptoms.

Several months into treatment, she became pregnant with her second child. As per international guidance, ustekinumab and mercaptopurine have been continued during pregnancy. She is for an elective c.section in view of her vulval disease and remains under follow up.

Discussion

VCD creates several clinical dilemmas. Firstly, owing to its variable presentations, it is often challenging to establish a diagnosis in the absence of GCD. Histological examination may show non-caseating granulomas, however this is often missed due to the lesions localising nature. Even if demonstrated, this is non-specific with wide differentials including tuberculosis and sarcoidosis. Secondly, the evolution of VCD is unpredictable and there is no established standard of care.

A lack of standardised treatment strategies and its rarity means that VCD is often clinically challenging. A multidisciplinary approach is required in order to minimise significant physical, social and mental morbidity.

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