

Use of Lymphoedema service for severe extraintestinal vulval Crohn's: a multidisciplinary approach

J Mann¹, N Mann¹, R England², M Sharma¹

¹Department of Dermatology, University Hospitals of Derby and Burton NHS Foundation Trust

² Department of Lymphoedema, University Hospitals of Derby and Burton NHS Foundation Trust

Introduction

Extraintestinal vulval Crohn's disease (CD) is a rare entity, often misdiagnosed. Patients experience pain, psychological distress and sexual dysfunction as a result of disfigurement.¹ Mechanisms of the disease are poorly understood, though it is thought to be linked to impairment of lymphatic drainage in intestinal CD, possibly from direct extension into the affected skin.²

We present a case of extraintestinal vulval Crohn's managed with multidisciplinary (MDT) approach and use of lymphoedema service.

Case

Our patient was a 24-year-old female with an extensive 8-year history of ileo-colonic and perianal Crohn's disease, being managed on Ustekinumab. She suffered with significant extra-intestinal involvement of the vulva with cobblestone appearance, knife cut erosions, induration, and discharge consistent with lymphorrhoea (see figure 1). There had been no response to local treatment with topical steroids and oral antibiotics. The appearance and symptoms had a significant impact on the patient's psychological wellbeing and her relationship with her partner.

Management

She was referred to the local lymphoedema clinic and commenced on prophylactic Penicillin V antibiotic as per the British Lymphology Society (BLS) guidelines. She was taught Manual Lymphatic Drainage (MLD) techniques (see figure 2) and provided compression pads to be worn inside support undergarments to improve her discomfort and symptoms (see figure 3).



Figure 1: vulval lymphoedema



Figure 2: MLD technique



Figure 3: compression pads

Six months after treatment, she noticed a dramatic improvement in her genital swelling, with a reduction in episodes of cellulitis and improvement in her psychological distress and sexual function.

Genital Lymphoedema Management Principles

- **Skin Care:** Soap substitute, moisturiser
- **Compression:** In the form of supportive undergarments or compression pads/garments e.g. Comfizz lycra shorts or foam inserts.
- **Exercise:** Pelvic floor exercises, swimming, yoga/pilates .
- **Lymphatic drainage:** Manual lymphatic drainage to stimulate the lymphatic system.

Conclusion

Patients with extraintestinal vulval Crohn's benefit from the MDT approach between surgery, gastroenterology, dermatology and in particular, the lymphoedema service to optimise disease management.

References:

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Figure 1 taken from DermNetNZ and Figure 2 and 3 taken from Google images