Unusual association of diseases/symptoms

Leukocytoclastic vasculitis presenting with acute haemorrhagic oedema of the scrotum

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Summary

Fournier's gangrene is a form of rapidly progressive necrotising fasciitis involving the soft tissues of the male genitalia. The treatment for this urologic emergency is immediate surgical debridement. Misdiagnosis of Fournier's may lead to unnecessary surgical interventions, making careful recognition of systemic signs of illness crucial. The authors present a case of a 48-year-old patient who presented emergently with massive scrotal oedema, erythema and pain suspicious for Fournier's gangrene and systemic signs of illness, including palpable purpura and leukocytoclastic vasculitis on biopsy.

BACKGROUND

Fournier's gangrene is a life-threatening emergency that requires immediate surgical debridement. We present this case to stress the importance of recognising systemic signs of illness that may account for scrotal pathology so as to prevent unnecessary and potentially harmful surgical interventions.

CASE PRESENTATION

A 48-year-old patient was transferred to our tertiary care centre from an outside hospital following the rapid overnight development of massive scrotal oedema, erythema and pain worrisome for Fournier's gangrene. On physical examination, the scrotum was grossly oedematous, tender and purple and approximately the size of a baseball (figure 1). The discolouration and swelling involved the penile shaft and perineum, which were also tender to palpation. The glans was spared. The patient also complained of suprapubic tenderness along the inguinal canal bilaterally.

Figure 1 Marked scrotal oedema.

There was no foul odour, crepitus or lymphadenopathy. Further findings on physical examination were multiple scattered violaceous palpable purpura on both legs, abdomen, flanks and buttocks (figure 2). In addition, a 3 cm bulla on the dorsum of the left foot was observed (figure 3).

The patient's history of present illness was notable for several weeks of unspecific gastroenterological complaints

Figure 2 Palpable purpura covering the right abdomen and flank.

Figure 3 3 cm bulla on dorsum of left foot.

Figure 4 LCV (100X periodic acid-Schiff (PAS) stain). There are superficial and mild dermal perivascular and interstitial infiltrates comprised mainly of neutrophils and nuclear dust of neutrophils. Fibrin is present in the walls of the blood vessel. Images provided by Dr Russo in dermatopathology.

for which the patient was prescribed ciprofloxacin, sucralate and metronidazole. After developing a painful and erythematous swelling of his left ankle, he was switched from ciprofloxacin to trimethoprim-sulfamethoxazole. Admission to the outside hospital was prompted by the development of acute bilateral lower extremity cellulitis

Figure 5 LCV (200X PAS stain). Higher magnification view of figure 4, demonstrating an inflammatory neutrophilic infiltrate in the upper dermis. There is purpura with red blood cells extravasated into the upper dermis. Images provided by Dr Russo in dermatopathology.

of unknown aetiology, which was treated in the intensive care unit setting with intravenous vancomycin and clindamycin for a presumed infectious aetiology. In addition, he developed a bulla on the dorsum of his left foot as well as a palpable but non-tender purpuric rash on his back, buttocks and lower extremities.

INVESTIGATIONS

Initial laboratory studies revealed an elevated white blood cell count of 13 900. Stool cultures, blood cultures, liver function tests, basic metabolic panel, hepatitis A, B and C panels were all normal. Erythrocyte sedimentation rate (ESR) was 80 H (0–10) and C-reactive protein (CRP) 90.6 H (0.0–4.9). A CT scan of the abdomen and pelvis showed marked scrotal oedema and inflammation but without subcutaneous emphysema. A biopsy of the rash from his right thigh revealed histopathologic features of leukocytoclastic vasculitis (LCV) with extension deep into the dermis (figures 4 and 5). Immunofluorescence staining was negative for IgA.

TREATMENT

With a suspicion of Henoch Schönlein purpura (HSP) given the findings of LCV in the dermis, antibiotics were discontinued and the patient was started on a 4-day course of 125 mg of intravenous methylprednisone every 6 h. Topical triamcinolone cream was applied daily over the lesions on his back, buttocks and legs. The patient was discharged with a tapered dose of steroids for 6 weeks.

OUTCOME AND FOLLOW-UP

The patient's scrotal oedema had improved at discharge but remained significant. An autoimmune investigation was negative. He had an uncomplicated recovery at home and returned to clinic 1 month later with complete resolution of his scrotal oedema.

DISCUSSION

Fournier's gangrene is a form of rapidly progressive necrotising fasciitis involving the soft tissues of the male

genitalia caused by a polymicrobial infection. This urologic emergency has a mortality estimated at 7.5%,¹ prompting immediate broad-spectrum antibiotic coverage and emergent surgical debridement to remove all areas of necrotic skin. Early diagnosis of Fournier's is based on clinical and radiologic findings. Classically, patients with Fournier's present with genital pain, crepitus, oedema, erythema, an odour and evidence of soft tissue necrosis. CT findings may demonstrate air in subcutaneous perineal tissues.² Recognising systemic signs of illness that may account for scrotal pathology is crucial to prevent the misdiagnosis of Fournier's, which puts the patient at risk of unnecessary surgical intervention.

In this patient, systemic signs of disease included scattered palpable purpura, subcutaneous non-pitting oedema, bullae and pathologically confirmed cutaneous leukocytoclastic vasculitis. LCV is a histopathologic term that encompasses a wide differential. The presentation is most consistent with hypersensitivity vasculitis, a small vessel immune complex mediated vasculitis secondary to a drug reaction, infectious process, vaccination or other stimulus. In this form of vasculitis, the skin is the predominant organ system involved, with palpable purpura and macules typically evident on the lower extremities.³ Our patient took two antibiotics and sucralfate prior to developing non-pitting oedema and a single bulla. He developed an oedematous scrotum on hospital day 7. Acute haemorrhagic oedema of the scrotum secondary to leukocytoclastic vasculitis was first described in 1913 by Snow and has been reported primarily in the paediatric population.⁴ Diagnosis is based on clinical presentation, including purpura, arthralgias and non-pitting oedema and a biopsy-proven LCV.⁵ Laboratory findings are generally non-contributory but may demonstrate an elevated CRP and ESR.³

HSP was initially on the differential given the findings of palpable purpura, abdominal pain and arthritis.⁶ Our patient did not likely have HSP given a negative IgA stain on direct immunofluorescence, a common diagnostic feature of this disease.⁷ Scrotal oedema has been reported in patients with HSP, but this association is not sensitive or

specific to HSP, and has mostly been reported in the paediatric population.^{8 9}

Learning points

- ▶ Patients with leukocytoclastic vasculitis may present with significant scrotal oedema, erythema and pain that closely mimics Fournier's gangrene.
- ▶ Vasculitis must be considered in the differential for scrotal pathology when systemic signs such as palpable purpura are present.
- ▶ Early recognition of systemic illness is crucial in order to avoid unnecessary surgical intervention.

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Competing interests None.

Patient consent Obtained.

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