Juvenile gangrenous vasculitis of the scrotum - a rare entity

Ferreras C[1], Gorito V[1], Coelho AR[2], Valente D[3], Amoedo P[4], Nogueira A[4], Rodrigues M[5], Brito I[5]

ACTA REUMATOL PORT. 2021;46:82-84

INTRODUCTION

Juvenile gangrenous vasculitis of the scrotum (JGVS) was first described in 1973, with less than 20 cases in the literature. It is characterized by an acute painful scrotal ulcer of unclear etiology observed in healthy young men[1].

CASE REPORT

A previously healthy 17-year-old male adolescent was admitted to the emergency department with painful scrotal ulcers occurring in the last 24 hours. Three days before he reported odynophagia and fever which resolved spontaneously. He denied local trauma, product application, sexual risk behaviors, as well as previous genital or oral ulcers. Physical examination revealed 3 necrotic ulcers in the right hemiscrotum with well-defined borders without suppuration, associated with local edema and erythema (Figure 1), and inguinal lymphadenopathy. Laboratory workup showed leukocytosis (22.5×10^9/L) with neutrophilia (87%) and elevated C-reactive protein (191mg/L). Microbiological and immunological studies were normal. Scrotal US showed subcutaneous thickening, without testicular changes. He received empirical intravenous ceftazidime and clindamycin. Biopsy revealed necro-hemorrhagic changes of the superficial skin, associated with extensive polymorphic, predominantly mononuclear inflammatory infiltrate and a recent vascular thrombus in the reticular dermis. There were no immune deposits in the vascular walls seen with direct immunofluorescence (Figure 2). These findings are compatible with JVGS. The lesions resolved within a month without sequelae.

1. Department of Pediatrics, Centro Hospitalar Universitário de São João; Faculdade de Medicina, Universidade do Porto;
2. Department of Pathology, Centro Hospitalar Universitário de São João
3. Department of Pediatrics, Centro Hospitalar Universitário de São João
4. Department of Dermatology and Venereology, Centro Hospitalar Universitário de São João
5. Pediatric and Young Adult Rheumatology Unit, Centro Hospitalar Universitário de São João; Faculdade de Medicina, Universidade do Porto

DISCUSSION

JVGS is characterized by a sudden onset, commonly preceded by an episode of pharyngitis with fevers up to 3 weeks before. It exclusively affects scrotal skin, with 1–5 round to polycyclic, sharply demarcated ulcers, which are initially pruritic and burning. The lesions rapidly progress to painful necrotic ulcers with black eschars. This is accompanied by an elevation of acute-phase reactants (ARF). Other laboratory findings including microbiological investigations are usually un-

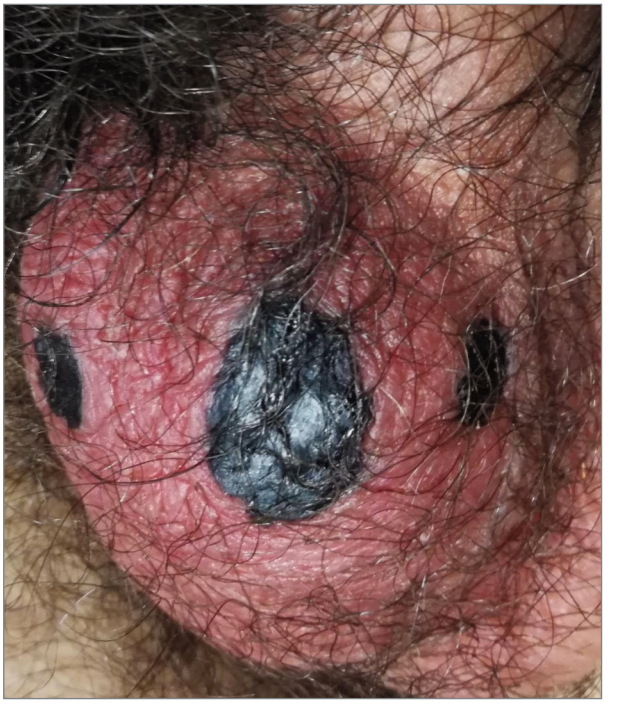

FIGURE 1. Necrotic scrotal ulcers in the hemiscrotum.

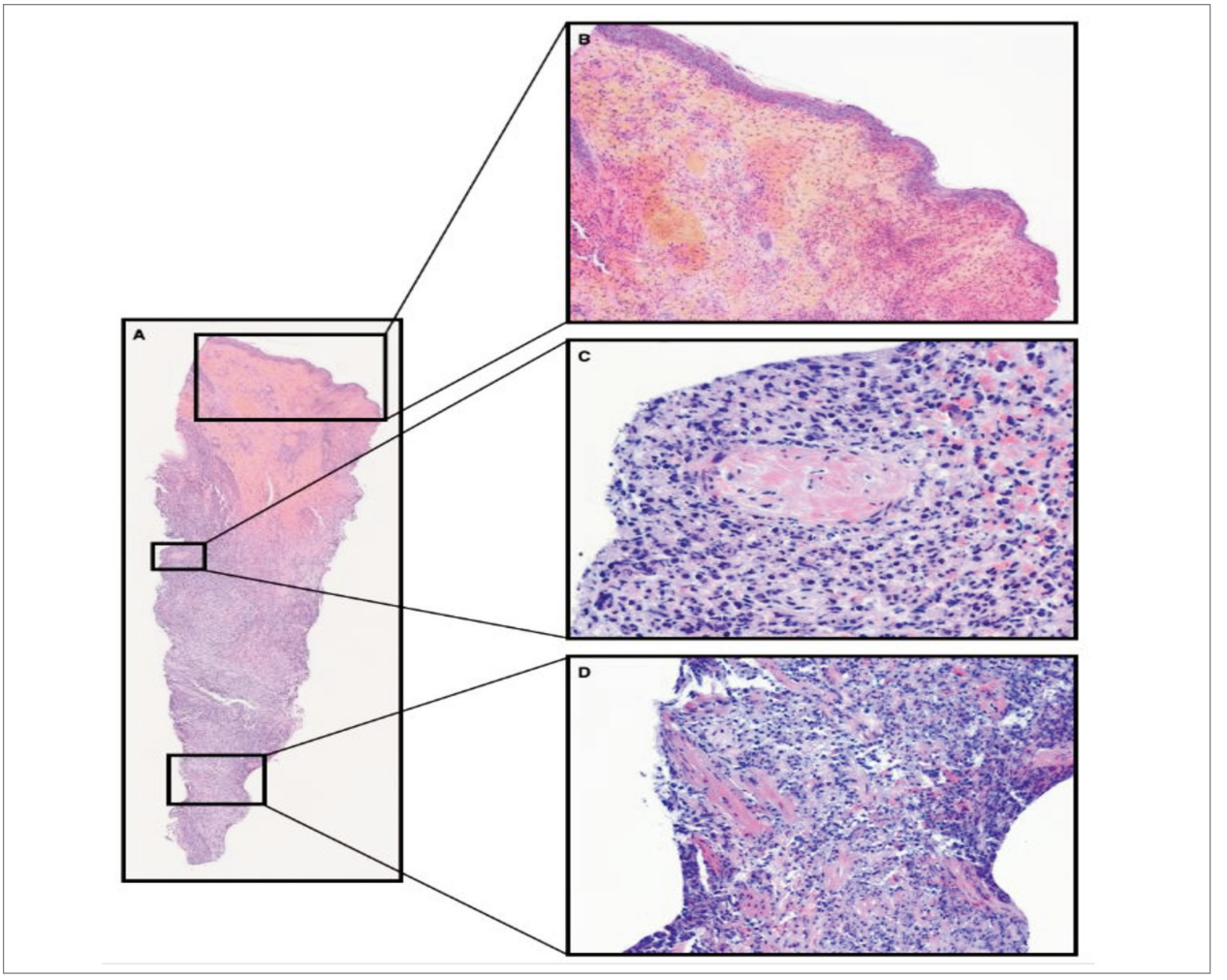

FIGURE 2. Scrotal wall biopsy (A, low magnification) disclosing superficial necro-hemorrahgic changes (B, HE 100x), dermal vessels with recent thrombus (C, HE 400x) and extensive polymorphic infiltrate that reach the cremasteric muscle level (D, HE 200x)

remarkable[2,3].

The differential diagnosis of JGVS includes numerous ulcerative scrotal diseases, such as sexually transmitted infections, Fournier's gangrene, or ecthyma gangrenosum. Recently, Chen W et al. proposed that given the overlap between JGVS and *ulcus vulvae acutum,* they may be counterparts of the same type of Lipschutz genital ulcer[4,5].

Histopathology of the ulcer is usually unspecific, including fibrinoid necrosis of small vessels, perivascular lymphocytic infiltrates, numerous extravasated red cells, and abundant neutrophilic infiltrate in the deep dermis, consistent with vasculitis[2,4].

The course is usually self-limited with resolution within 2–3 weeks without relapse. Therapeutic response to a short course of systemic antibiotics and steroids was reported to be effective in most cases, but spontaneous healing is likely[4].

Despite accurate initial clinical suspicion, given the presence of high ARF, the severity of other diagnostic hypothesis, and since diagnostic confirmation is usually retrospective through histopathology, initial antibiotic treatment and steroid delay are frequently unavoidable, such as in the present case.

In conclusion, although very few cases have been described, this entity should be kept in mind whenever healthy young individuals present with abrupt scrotal ulcerations, which otherwise seem well.

Despite its benign course, the acute presentation is often distressing and alarming for both patients and

clinicians.

These patients may present to pediatricians, urologists, dermatologists, infectious disease specialists, or rheumatologists, further hindering a correct diagnosis.

CORRESPONDENCE TO
Cristina Ferreras
Department of Pediatrics
Centro Hospitalar Universitário de São João
Alameda Prof. Hernâni Monteiro, 4200-319, Porto
E-mail: cristinaferreras87@gmail.com

REFERENCES

1. Pinol Aguade J, Castells Mas A, Lecha M, Ferrando J, Castells Rodellas A. A Propos de cinq Observations d'une forme Particuliere de Vascularite Gangreneuse Juvenile du Scrotum. Medecine et Hygiene, Geneve, 1974: 112–128.
2. Nuno Gomes N, Costa-Silva M, Nogueira A, et al. Juvenile gangrenous vasculitis of the scrotum – a diagnosis not to forget. Int J Dermatol. 2020 Mar;59(3):e49-e50
3. Nettleton J, Crawford-Smith H, Adimonye A, McMeekin F. Scrotal necrosis and no Fournier's in sight: a rare case of juvenile gangrenous vasculitis. BMJ Case Rep 2019;12:e226530.
4. Chen W, Plewig G. Lipschutz genital ulcer revisited: is juvenile gangrenous vasculitis of the scrotum the male counterpart? J Eur Acad Dermatol Venereol 2019; 33: 1660–1666.
5. Jimenez-Cauhe J, Gil-Redondo R, Dominguez-Santas M, et al. Juvenile gangrenous vasculitis of the scrotum: the male counterpart of Lipschutz ulcer? J Eur Acad Dermatol Venereol 2019 Dec; 33(12):e459-e461