## Purpura Fulminans and *Rickettsia Conorii* Israeli Tick Typhus Infection

## Púrpura Fulminante e Infeção por Tifo da Carraça Israelita *Rickettsia Conorii*

**Keywords:** Purpura Fulminans; Rickettsia conorii; Rickettsia Infections; Spotted Fever Group Rickettsiosis
**Palavras-chave:** Infecções por Rickettsia; Purpura Fulminante; Rickettsia conorii; Rickettsiose do Grupo da Febre Maculosa

Purpura fulminans (PF) is a rare, life-threatening thrombotic disorder marked by sudden cutaneous hemorrhage, skin necrosis, and disseminated intravascular coagulation.[1] It is commonly associated with severe bacterial infections, especially *Neisseria meningitidis* and *Streptococcus pneumoniae*, and can also result from rickettsial diseases.[2] Spotted fever, caused by *Rickettsia* species, is endemic in Mediterranean countries, including Portugal, and presents with fever, rash, and an inoculation eschar. If untreated, it can lead to PF, multi-organ failure, and septic shock. These conditions are due to failure of the anticoagulant protein C pathway and uncontrolled microvascular clotting.[3] Early diagnosis and treatment are essential, even though PF often has high morbidity and mortality rates up to 32.3% in hospitalized patients, despite therapy.[1,2]

We describe the case of a 67-year-old male presenting with hypertension, dyslipidemia, and obesity, with fever (40°C), abdominal pain, and myalgias lasting for eight days. He lived in an urban apartment, with no recent travel or animal contact, except for his domestic dog and cat. Physical examination revealed hypotension and non-blanching erythematous-violaceous macules lasting 24 hours, compatible with PF, and a necrotic plaque on the left thigh, consistent with a *tache noire* (Fig. 1). This raised suspicion for Mediterranean spotted fever, acute infectious PF by other infectious agent and idiopathic or acquired PF. Lab results showed thrombocytopenia, acute kidney injury and rhabdomyolysis. Empirical treatment with doxycycline, piperacillin-tazobactam, and vasopressors was started. Despite improvement from septic shock after treatment for 21 days with doxycycline, the thrombotic nature of PF, combined with vasoconstriction induced by the vasopressor therapy, caused extensive cutaneous necrosis on the lower limbs and scrotum, bullae formation and secondary bacterial infections, resulting in bilateral below-knee amputations and scrotal reconstruction. Skin biopsy confirmed the initial hypotheses, revealing thrombotic vasculopathy. Furthermore, polymerase chain reaction on the skin biopsy confirmed *Rickettsia conorii* Israeli tick typhus.

This case demonstrates a rare yet severe complication of Mediterranean spotted fever, resulting in PF and multi-organ failure due to endothelial injury, disseminated intravascular coagulation and tissue necrosis.[3] The patient's progression from nonspecific symptoms to septic shock and PF emphasizes the challenge of diagnosing rickettsial infections, particularly in non-endemic urban areas. The presence of a *tache noire* is a critical diagnostic clue in febrile patients with a rash.[4] Despite prompt treatment with doxycycline, rapid deterioration can occur once endothelial damage and coagulation are widespread. This case underscores the importance of an early diagnosis and a multidisciplinary care in managing PF's complications.[5]

### AUTHOR CONTRIBUTIONS

FM, MA, PF: Study design, data acquisition and analysis, writing and critical review of the manuscript.

ITA, PV: Data acquisition, writing and critical review of the manuscript.

All authors approved the final version to be published.

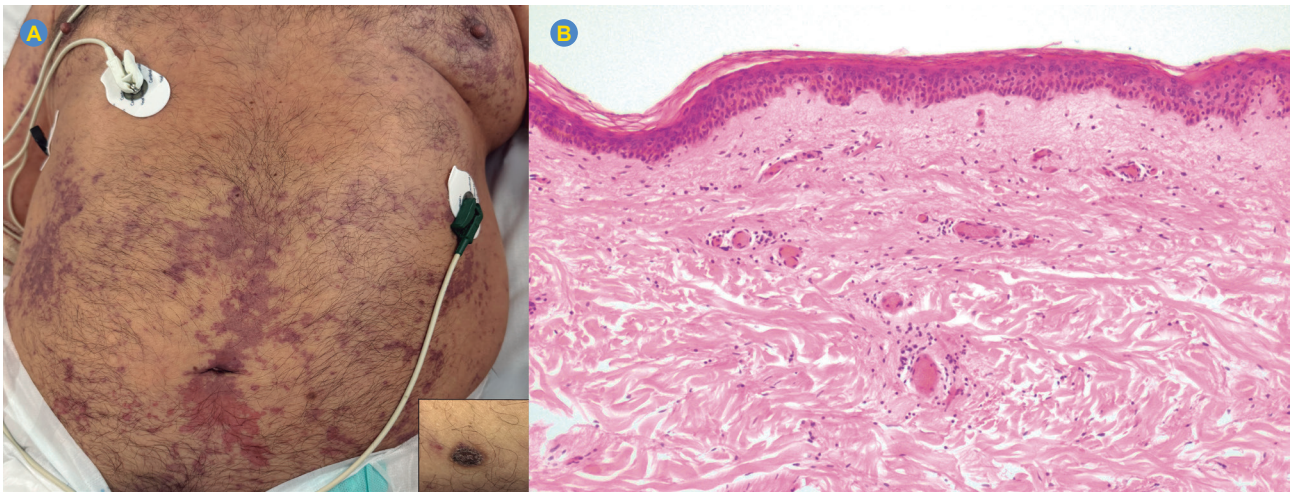


Figure 1 – Physical examination revealing non-blanching erythematous-violaceous macules and patches distributed on the abdomen and chest, and a black eschar on the medial region of the left thigh compatible with a *tache noire* (inset) (A). Intravascular hyaline thrombi in the dermis, accompanied by a sparse perivascular neutrophilic infiltrate and no epidermal changes on histopathological exam (B).

## PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

## DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

## PATIENT CONSENT

Obtained.

## COMPETING INTERESTS

The authors have declared that no competing interests exist.

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