Púrpura retiforme secundaria a *Rickettsia conorii*

Retiform purpura secondary to Rickettsia conorii

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ABSTRACT

Retiform purpura consists of branching purpuric lesions caused by a complete blockage of blood flow in the dermal/subcutaneous vasculature. It is an acute life-threatening disorder characterized by intravascular thrombosis and hemorrhagic infarction of the skin complicated with disseminated intravascular coagulation. It is commonly seen in acute infections following meningococcal and streptococcal infections. Few cases have been described of retiform purpura following rickettsial infections in the literature and rarely with this magnitude.

The purpose of this report is to highlight the possibility of this situation in critically ill patients. We report a case of a 62-year-old woman admitted in the intensive care unit with septic shock from *Rickettsia conorii*. She developed quickly an intense retiform purpura requiring several surgical interventions to control the necrosis. The dermatologic situation was only completely controlled with treatment of underlying situation. This is a severe complication of a critical infectious disease.

Keywords: Retiform purpura, vasculopathy, vasculitis, *Rickettsia conorii*.

RESUMEN

La púrpura retiforme consiste en lesiones purpúricas ramificadas causadas por un bloqueo completo del flujo sanguíneo en la vasculatura dérmica/subcutánea. Es un trastorno agudo potencialmente mortal caracterizado por trombosis intravascular e infarto hemorrágico de la piel complicado con coagulación intravascular diseminada. Se ve comúnmente en infecciones agudas después de infecciones meningocócicas y estreptocócicas. Se han descrito pocos casos de púrpura retiforme tras infecciones por rickettsiosis en la literatura y rara vez de esta magnitud.

El propósito de este informe es resaltar la posibilidad de esta situación en pacientes críticamente enfermos. Presentamos el caso de una mujer de 62 años ingresada en la unidad de cuidados intensivos con shock séptico por *Rickettsia conorii*. Rápidamente desarrolló una intensa púrpura retiforme que requirió varias intervenciones quirúrgicas para controlar la necrosis. La situación dermatológica solo se controló por completo con el tratamiento de la situación subyacente. Esta es una complicación grave de una enfermedad infecciosa crítica.

Palabras clave: Púrpura retiforme, vasculopatía, vasculitis, *Rickettsia conorii*.

INTRODUTION

Branching purpuric lesions, also known as retiform purpura, are a net-like pattern of cutaneous hemorrhages reflecting the anatomy of the underlying blood supply to the dermis and subcutaneous tissue[1]. Retiform purpura occurs when intravascular thrombosis and hemorrhagic infarction of the skin that is rapidly progressive and accompanied by vascular collapse and disseminated intravascular coagulation.[1]

The commonest origin is the acute infections, most notably with meningococcal, staphylococcal, and streptococcal infection[1]. Retiform purpura caused by rickettsial infection is very rarely reported in the literature[1]. If the etiology of purpura is promptly diagnosed as a rickettsial infection and treated accordingly, the outcome will be excellent. We report a case of a 62-year-old woman with rickettsial infection in septic shock evolving with retiform purpura.[2]

CASE DESCRIPTION

A 62-year-old woman was admitted to the emergency department of our hospital with a fever, generalized myalgia, anorexia and a recent rash that was erythematosus initially, starting over her bilateral lower limbs and hands and progressing rapidly to involve her back and trunk. Her medical history included hypertension, dyslipidaemia, obesity, and depression. The patient lived in a rural area and had been exposed to 2 dogs at home but had no recollection of tick exposure. On examination, she was in severe distress with a high fever of 39.4ºC, taquicardia. hypotensive and had a disseminated, purpural rash over her body with areas of hemorrhage over both legs, hands, and the trunk. Her higher mental functions were intact. Laboratory findings included leukocytosis, severe thrombocytopenia, acute renal failure, increased levels of liver enzymes, creatine kinase and lactate dehydrogenase.

She was admitted to the intensive care unit (ICU) in a state of septic shock and given broad-spectrum antimicrobial drug therapy (4th day of onset symptoms). During the next 24h she evolved in to multiorgan failure, severe jaundice, liver failure, and acute lung injury developed, disseminated intravascular coagulation (DIC) and the rash became black, necrotic with a few hemorrhagic blebs and muscle exposure in the lower extremities (Figure 1). Further investigation was preformed upon admission in the ICU with skin biopsy of the purpuric lesions. From the instigation initially preformed upon admission in the emergency room, the serologic testing resulted in the identification of antibodies of *Rickettsia* by indirect immunofluorescence (*Rickettsia conorii* - IgM 303, IgG 80) and positive hemocultures for *Rickettsia conorii*. The antimicrobial drug therapy was switched to intravenous doxycycline. The skin biopsy, that was sent to another center, revealed

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Figure 1. Purpura over her body upon 48h of hospital admission. Muscle tissue visible.

Figure 2. Exposure on the calcaneal tendon in the right ankle.

Figure 3. Near-total recovery on the 22nd day

a prominent thrombosis with neutrophilic inflammation and positive detection of *Rickettsia conorii* DNA by polymerase chain reaction. Serologic testing for *Rickettsia* was not repeated.

As time progressed, it was possible to observe clinical recovery. The skin lesions required several surgical interventions to control the necrosis and exposure on the calcaneal (Achilies) tendon in the right ankle (Figure 2). Antipyretics, antiallergy drugs, and topical emollients were also applied. She recovered almost completely, with regression of the rash at follow-up after 22 days and transferred to the surgical ward (Figure 3).

DISCUSSION

Retiform purpura can occur in a life-threatening disorder characterized by sudden progressive cutaneous hemorrhage and necrosis[3]. It often affects the distal extremities, especially when patients are on vasopressors and have an additional component of peripheral gangrene related to pressor use[3]. Cutaneous involvement with DIC is also characteristically symmetric[4]. Because of the high mortality associated with DIC, prompt recognition and rapid treatment of the underlying cause is essential[4]. While the underlying etiology is identified and corrected, hemodynamic support, including optimization of fluid status, blood pressure, temperature, and pH is critical.[3]

The clinical presentation of rickettsial infection has a wide spectrum ranging from undifferentiated fever to multiorgan involvement leading to fatal outcomes[1]. It can present with various cutaneous manifestations, typically features a skin rash including discrete macular-papular lesions distributed mainly in the hands, limbs, chest, abdomen, and the soles of the feet[1]. However, there are many other variations, including fern leaf pattern skin necrosis, patchy necrotic lesions, and cutaneous edema. Identification of cutaneous lesions in rickettsial infections plays a pivotal role in making a diagnosis early.[1]

In the literature, rickettsial infections are very rarely reported to cause retiform purpura. Treating the infection is fundamental in reversing the situation. The first drug of choice for rickettsial disease is doxycycline[5]. Alternatively, chloramphenicol can be used[5]. Some strains of *Rickettsia conorii* are poorly responsive to standard antirickettsial drugs, including tetracycline or doxycycline, and alternative treatment should be issued[5]. The treating physician should consider the possibility of rickettsial infection in the etiological diagnostic workup of any patient who presents with a severe purpuric and hemorrhagic rash.

CONFLICT OF INTEREST
The authors declare that there is no conflict of interest in this work.

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ETHICAL ASPECTS
All participants submitted a consent form to be included in this study.

REFERENCES

1. Dalugama C, Gawarammana IB. Rare presentation of rickettsial infection as purpura fulminans: a case report. J Med Case Rep. 2018;12(1).
2. Wysong A, Venkatesan P. An approach to the patient with retiform purpura: Approach to retiform purpura. Dermatol Ther. 2011;24(2):151–72.
3. Georgesen C, Fox LP, Harp J. Retiform purpura: Workup and therapeutic considerations in select conditions. J Am Acad Dermatol. 2020;82(4):799–816.
4. Sheikh AB, Javed N, Stoltze K. Diffuse Dermal Angiomatosis Manifestation of Underlying Severe Peripheral Vascular Disease. EJCRIM. 2020;7.
5. Biggs HM, Behravesh CB, Bradley KK, Dahlgren FS, Drexler NA, Dumler JS, et al. Diagnosis and management of tickborne rickettsial diseases: Rocky Mountain spotted fever and other spotted fever group rickettsioses, ehrlichioses, and anaplasmosis - United States: A practical guide for health care and public health professionals. MMWR Recomm Rep. 2016;65(2):1–44.