Manchas Eritematosas Policíclicas de Instalação Aguda e Rapidamente Progressivas

Pedro Andrade

Assistente Hospitalar de Dermatovenereologia/Consultant of Dermatovenereology; Serviço de Dermatologia, Hospital Pedro Hispano - ULS Matosinhos EPE – Matosinhos, Portugal

PALAVRAS-CHAVE – Angioedema; Eritema; Idoso.

Dermatology Quiz

Rapidly Progressive Acute Polycyclic Erythematous Patches

KEY-WORDS - Aged; Angioedema; Erythema.

75-year-old male with known diagnosis of hypertensive chronic kidney disease, low-risk prostate adenocarcinoma and type 1 hereditary angioedema, was admitted in the hospital for intravenous treatment of left basilar acute pneumonia and later referred to the Dermatology Department for evaluation of an acute skin rash.

The patient presented large serpiginous and polycyclic erythematoviolaceous patches, with rapid centrifuge progression, located on the trunk (Fig. 1); these lesions became apparent on the day before his visit – five days after starting treatment with meropenem, initiated after piperacillin/tazobactam-induced pancytopenia. No relevant symptoms were reported apart from dyspnea and cough, related with the lung infection.

A cutaneous biopsy was taken after infusion of a single unit of fresh-frozen plasma, revealing a very sparse perivascular cell infiltrate and disproportionate dermal vascular dilation and oedema, underlying an atrophic epidermis, compatible with the clinical diagnosis of erythema marginatum-like eruption (Fig. 2).



Figure 1 - Rapidly evolving polycyclic erythematoviolaceous patches on the trunk.

Correspondência: Pedro Andrade Serviço de Dermatologia - Hospital Pedro Hispano, ULS Matosinhos EPE R. Dr. Eduardo Torres - 4464-513 Senhora da Hora, Portugal Tel: +351 229391000 E-mail: pedrodesousaandrade@gmail.com Recebido/Received 5 Outubro/5 October 2016 Aceite/Accepted 11 Outubro/11 October 2016

Qual o Seu Diagnóstico?



Figure 2 - Histopathological examination [H&E, (A) 40x; (B) 100x]: very sparse perivascular cell infiltrate and disproportionate dermal vascular dilation and oedema, underlying an atrophic epidermis.

DIAGNOSIS: ERYTHEMA MARGINATUM-LIKE ERUP-TION IN HEREDITARY ANGIOEDEMA

Erythema marginatum-*like* eruption is a rare condition described in patients with hereditary angioedema, and usually arises as a preceding or concomitant sign in acute angioedema crisis.¹⁻⁴

This patient regularly attended Allergology consultations for the diagnosis of type 1 hereditary angioedema, established after confirmation of low seric levels and functional activity of C1-esterase inhibitor (7.1mg/dL and 40%, respectively) in the context of recurrent abdominal pain episodes, diarrhoea and cutaneous angioedema. His brother, father and several paternal relatives also presented symptoms. The patient was not undergoing continuous prophylactic treatment due to the inexistence of previous glotic edema or respiratory crisis, and the low intensity, short duration and spontaneous regression of the recurrent abdominal symptoms; brief treatment courses with aminocaproic acid were sporadically prescribed with rapid symptomatic resolution.

In this case, erythema marginatum-like eruption might have been triggered by the acute pneumonia, intravenous

antibiotic treatment or secondary immune disturbance. Treatment was promptly initiated with aminocaproic acid (3 gb.i.d) and, as a result, a complete resolution of the dermatosis was observed in less than 36 hours, without residual post-inflammatory skin changes. Interestingly, no symptoms or lesions of acute angioedema were reported, and treatment was interrupted after 5 days. No recurrence of the dermatosis was observed in the following months.

The interest of this case is justified not only by the rare presentation of erythema-marginatum-*like* eruption with its typical evanescent figurate pattern, but also by the significant scarcity of the dermal inflammatory infiltrate contrasting with a rather intense vascular dilation and dermal edema, curiously matching the classical histopathological features of acute angioedema plaques.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse. Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa. Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes. Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

Conflicts of interest: The authors have no conflicts of interest to declare. **Financing Support:** This work has not received any contribution, grant or scholarship. **Confidentiality of data:** The authors declare that they have followed the protocols of their work center on the publication of data from patients. **Protection of human and animal subjects:** The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

REFERENCES

- 1. Bygum A. Hereditary angio-oedema in Denmark: a nationwide survey. Br J Dermatol. 2009; 161:1153-8.
- Ergin H, Baskan M, Akalin N, Gürses D. A case of hereditary angioedema with recurrent arthritis, erythema marginatum-like rash and chest pain. Turk J Pediatr. 2003; 45:261-4.
- Farkas H, Harmat G, Fáv A, Fekete B, Káradi I, Visy B, et al. Erythema marginatum preceding an acute oedematous attack of hereditary angioneurotic oedema. Acta Derm Venereol. 2001; 81:376-7.
- Starr JC, Brasher GW. Erythema marginatum preceding hereditary angioedema. J Allergy Clin Immunol. 1974; 53:352-5.