Perniosis

Manuel Mendes Gomes, Lèlita Santos

Serviço de Medicina Interna,
Centro Hospitalar
e Universitário de Coimbra,
Coimbra, Portugal

Correspondence to
Dr Manuel Mendes Gomes,
manuelmgomes@gmail.com

Accepted 20 April 2014

DESCRIPTION

This 39-year-old woman came to our clinic because of severe cyanotic fingers (figure 1), toes, nose and ears, with wounds in the back of both hands but without necrosis (figure 2). These were longstanding and very disabling symptoms, in intimate relation with cold exposure, persisting all winter (temperatures around 5–10°C) and disappearing in the summer. There were no other identified triggering factors. She did not have a classic three-phase Raynaud's phenomenon or other accompanying signs and symptoms, such as fever or arthralgias. She was a non-smoker and her medical history was unremarkable, and so was her familial history. There was no chronic or occasional medication intake. Blood tests were unremarkable, including autoimmunity (ANA, ENA, anti-ds-DNA antibodies and antiphospholipid antibodies), cryoglobulins, complement, rheumatoid factor, lupus anticoagulant, Venereal Disease Research Laboratory (VDRL), HIV and hepatitis C serologies. Nailfold capillaroscopy was unremarkable (no sclerosis, no giant capillaries and no major tortuosities). Skin biopsy showed dermal and hypodermal perivascular, perifollicular and perieccrine lymphocytic infiltrate, without interface changes, thus ruling out lupus and confirming the diagnosis of perniosis (despite the absence of oedema).

Furthermore, the lack of systemic symptoms or thrombotic events (especially in the extremities) made it less likely to be cryofibrinogenaemia¹; keratolytic winter erythema was improbable since there was no familial history, other triggering factors (such as stress or menstruation), centrifugal peeling of the skin on the palm and skin biopsy did not show typical compacted parakeratotic layer.² The absence of concomitant fever, arthralgia and joint

Figure 2 Large wound on the back of left hand. Cyanotic fingers and desquamative lesions. Lesion probably in relation with continuous cold exposure and minor traumatic events.

swelling ruled out familial cold autoinflammatory syndrome. Biopsy and blood tests ruled out lupus.

Treatment with vasodilators was moderately effective.

Perniosis (or pernio) is a common condition, within the spectrum of acrosyndromes and Raynaud's phenomenon.³ It manifests as painful erythematous or violaceous papules on the extremities. Most cases resolve in a few weeks. Severe lesions, like those found in our patient, are very rare.

Figure 1 Severe cyanotic fingers, with desquamative lesions and wounds in the back of both hands. Both palms were free of lesions and there was no evidence of livedo reticularis or necrosis. Picture taken after 30 min in a room with a temperature of 23°C

Learning points

- ▶ Perniosis is a relatively common condition which can be very disabling.
- ▶ Severe lesions, namely wounds, are rare but may occur in this setting.
- ▶ Ruling out other conditions is essential, since most cases of perniosis respond well simply to symptomatic treatment.

Contributors MMG is the responsible physician for this patient and respective diagnosis and for writing the article. LS revised the article.

Competing interests None.

Patient consent Obtained.

To cite: Gomes MM, Santos L. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-203732

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Rico MF, Brusco JE, *et al.* Criofibrinogenemia esencial. A propósito de un caso. *Arch Argent Dermatol* 2013;63:17–22.
- 2 Danielsen AG, Weismann K, Thomsen HK. Erythrokeratolysis hiemalis (keratolytic winter erythema): a case report from Denmark. *J Eur Acad Dermatol Venereol* 2001;15:255–6.
- 3 Jacob JR, Weisman MH, Rosenblatt SJ, *et al.* Chronic pernio. A historical perspective of cold-induced vascular disease. *Arch Intern Med* 1986;146:1589–92.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow