Acute Perniosis in Elderly People: A Predictive Sign of Systemic Disease?

Massimo Guadagni and Guido Nazzari
UOC Dermatologia La Spezia, Ospedale Civile S. Andrea, Viale Vittorio Veneto n°196 La Spezia (Sp), Italy. E-mail: massimo.guadagni@asl5.liguria.it
Accepted March 25, 2010.

Perniosis is a cold-induced vasculopathic disease characterized by tender, painful, pruritic erythematous, livid papular or nodular acral lesions, with bullous and ulceronecrotic lesions sometimes present in severe cases. The most frequently affected areas are the hands, feet, fingers, nose and ears. The occurrence of acute perniosis (AP) in elderly people is unusual. Correlation with a wide range of extracutaneous diseases has been reported. This preliminary study indicates that the sudden appearance of perniosis in an adult must be evaluated carefully, as it may be a predictive sign of an associated systemic disease.

MATERIALS AND METHODS
Consecutive office-based patients over the age of 65 years, presenting with an acute or previously unevaluated episode of perniosis were enrolled in a preliminary study during the period September 2006–June 2008 in Eastern Liguria, North Italy. Exclusion criteria were: positive anamnesis for acrocyanosis, atherosclerosis, previous and recurrent perniosis, recognized connective tissue or neoplastic disease, and significant (not occasional) use of perniosis-related drugs (1–5).

A complete check-up, consisting of clinical, blood, urine and instrumental investigations (Table I), was applied to assess the general health status of every patient. Any abnormal signs or symptoms were investigated further with appropriate laboratory studies. Follow-up visits were scheduled monthly until September 2007.

RESULTS
Seven consecutive out-patients, age range 65–87 years were enrolled in the study. All patients manifested AP in the autumn–winter period, with persistence of the lesions for more than 2 months and complete or partial resolution in the summer months. A careful clinical and drug history was taken. Histological examination of perniosis specimens revealed epidermal hyperkeratosis, a dense perivascular lymphomonocytic infiltrate in the superficial dermis, and small vessel damage. Immunofluorescence studies were negative. One patient did not complete the study (Table I). In only one patient was no associated disease detected. In all other cases diagnosis of an underlying disease was made during the study period.

DISCUSSION
Perniosis (chilblains) occurs predominantly in the acral areas. It must be distinguished from acrocyanosis (6, 7), acral vascular syndrome (8) and perniotic eruptions (9).

Although the pathogenetic mechanism for AP has not yet been determined, the presence of an abnormal vascular response to cold and minimal trauma is strongly suspected. Crowson & Magro (9) hypothesized an endothelial cell injury due to pathological circulating substances (fibrin, antibodies, abnormal cells, etc.) as an inducer of AP. The presence of positive immunofluorescence, especially for SSA(Ro) and an interface dermatitis or vasculopathic reaction favourably support this mechanism (9–11).

A wide range of diseases causing an increase in serum viscosity, coagulation abnormalities or pathological cell circulation has been reported in association with perniosis, such as proliferative blood cell lines disorders, viral infections, cytoproteinaemia, connective tissue disease and diseases causing weight reduction (12–17).

For these reasons the unexpected appearance of perniosis in elderly people, especially male patients,
without evidence of vascular, autoimmune disorders or use of suspected drugs, can represent an important sign that further clinical investigation is necessary. Despite the limited number of published cases, this preliminary study confirms this hypothesis and strongly supports the need for more extensive studies.

The authors declare no conflict of interest.

REFERENCES

1. Maaron SM, Hansley D. Pernio. In e-medicine. 2005. [cited 2009 August 30]. Available from: URL: http://emedicine.medscape.com/article/1087946-overview.

2. Devos SA, Van Den Bossche N, De Vos M, Naeyaert JM. Adverse skin reactions to anti-TNF-alpha monoclonal antibody therapy. Dermatology 2003; 206: 388–390.

3. Richez C, Dumoilin C, Schaeverbeke T. Infliximab induced chilblain lupus in a patient with rheumatoid arthritis. J Rheumatol 2005; 32: 760–761.

4. Gocal R, Dornan TL, Ledinghan JGG. Peripheral skin necrosis complicating beta-blockade. BMJ 1979; I: 721–722.

5. Hoffbrand BJ. Peripheral skin necrosis complicating beta-blockade. BMJ 1979; I: 1082.

6. Nousari HC, Kimyai-Asadi A, Anhalt GJ. Chronic idiopathic acrocyanosis. J Am Acad Dermatol 2001; 45: S207–S208.

7. Del Giudice P, Durant J, Dellamonica P. Hand edema and acrocyanosis: “puffy hand syndrome”. Arch Dermatol 2006; 142: 1084–1085.

8. Poszepczynska-Guigné E, Viguier M, Chosidow O, Orcel B, Emmerich J, Dubertret L. Paraneoplastic acral vascular syndrome: epidemiologic features, clinical manifestations, and disease sequelae. J Am Acad Dermatol 2002; 47: 47–52.

9. Crowson AN, Magro CM. Idiopathic perniosis and its mimics: a clinical and histological study of 38 cases. Hum Pathol 1997; 28: 478–484.

10. Magro CM, Crowson AN. The cutaneous pathology associated with seropositivity for antibodies to SSA (Ro): a clinicopathologic study of 23 adult patients without subacute cutaneous lupus erythematosus. Am J Dermatopath 1999; 21: 129–137.

11. Viguier M, Pinquier L, Cavelier-Balloy B, De La Salmoniere P, Cordoliani F, Flageul B, et al. Clinical and histopathologic features and immunologic variables in patients with severe chilblains. A study of the relationship to lupus erythematosus. Medicine 2001; 80: 180–188.

12. Dreno B, Gandon P, Bureau B, Milpied N, Barriere H. Skin lesions from hypersensitivity to cold during chronic myelomonocytic leukaemia. Br J Dermatol 1986; 115: 607–609.

13. Kelly JW, Dowling JP, Pernio. A possible association with chronic myelomonocytic leukaemia. Arch Dermatol 1985; 121: 1048–1052.

14. Marks R, Lim CC, Borrie PF. A perniotic syndrome with monocytosis and neutropenia: a possible association with a preleukaemic state. Br J Dermatol 1969; 81: 327–332.

15. Sun NCJ, Bowie E JW, Kazmier FJ, Elveback LR, Owen CA. Blood coagulation studies in patients with cancer. Majo Clin Proc 1974; 49: 636–641.

16. Yazawa H, Saga K, Omori F, Jimbow K, Sasagawa Y. The chilblain-like eruption as a diagnostic clue to the blast crisis of chronic myelocytic leukemia. J Am Acad Dermatol 2004; 50: S42–S44.

17. Goette DK. Chilblains (perniosis). J Am Acad Dermatol 1990; 23: 257–262.