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

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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 Easthern 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, cyoproteinaemia, connective tissue disease and diseases causing weight reduction (12–17).

For these reasons the unexpected appearance of perniosis in elderly people, especially male patients,

Table I. *Case collection*

Pat. No./sex/ age (years)	Symptoms	Check abnormalities[a]	Related diseases
1/M/87	Acute onset on hands, ears and nose	Anaemia, Lymphocytopaenia, Granulocytopaenia	Myelodysplastic disease
2/M/80	Acute onset on hands, ears and nose	Anaemia, Lymphocytopaenia, Granulocytopaenia	Myelodysplastic disease
3/M/87	Acute onset on hands and ears 2 years previously	Lymphocytosis	Chronic myelomonocytic leukaemia
4/M/76	Hand and ear perniosis	Anaemia	Colorectal adenocarcinoma, Ofuji papuloerythroderma
5/F/83	Hand perniosis	RF positive, ANA 1:640, ENA (Ro-SSA+ La-SSB+)	Sjögren's syndrome
6/F/67	Hand perniosis for 2 years	NK-T cells reduction, β2 microglobulinaemia, Raynaud's phenomenon in infancy	None
7/M/65	Hand perniosis	Diabetes, RF positive	No follow-up

[a]*Check panel*: complete blood cell count, sedimentation rate, ANA, ENA, anti-DNA, anti-phospholipid antibody panel, cryoglobulin, cryofibrinogen, cold agglutinins, serum protein electrophoresis, quantitative immunoglobulins, Bence-Jones proteins, screening test for hepatitis virus B,C and for HIV, rheumatoid factor, C3/C4 levels, ECG, chest-X ray, abdominal ultrasonographic examination, and serological markers of cancer.
NK-T: natural killer T cells; ANA: anti-nuclear antibody; ENA: extractable nuclear antigens; RF: rheumatoid factor; ECG: electrocardiography.

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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.

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