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PHOTO QUIZ

Skin lesions as a first presentation

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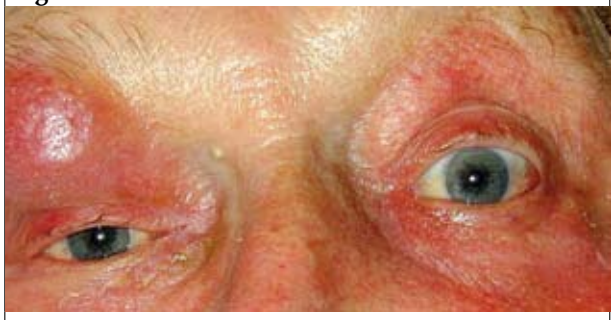
CASE REPORT

A healthy 79-year-old man presented to the Department of Dermatology in September 2003 with purple-red, swollen lesions involving the whole body for a period of five weeks (*figure 1*). The eyebrows of the right eye were swollen, the upper more than the lower. At that time laboratory results were: haemoglobin 7.2 mmol/l, platelet count 162 x 10^9/l, white blood cell count 6.9 x 10^9/l with a monocytosis of 20.9% and C-reactive protein of 255 mg/l. Skin biopsy is shown in *figure 2*.

WHAT IS YOUR DIAGNOSIS?

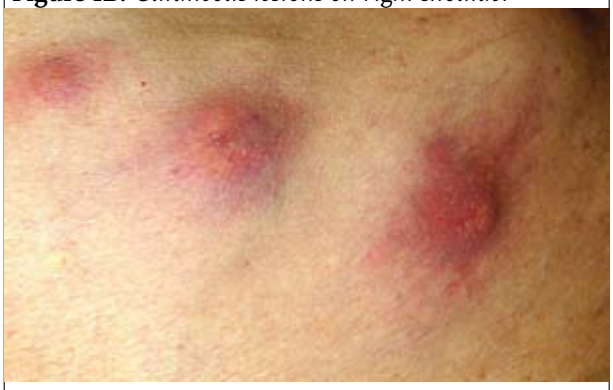
See page 265 for the answer to this photo quiz.

Figure 1A. *Cutaneous lesions*



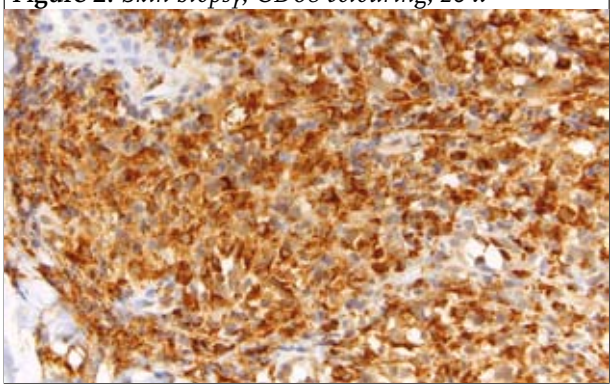
Pictures made after starting treatment, but hardly changed compared with the moment of presentation.

Figure 1B. *Cutaneous lesions on right shoulder*



Pictures made after starting treatment, but hardly changed compared with moment of presentation.

Figure 2. *Skin biopsy, CD68 colouring; 20 x*



ANSWER TO PHOTO QUIZ (ON PAGE 264)
SKIN LESIONS AS A FIRST PRESENTATION

DIAGNOSIS

Bone marrow aspiration showed a hypercellular bone marrow with an increase of atypical myeloid blasts. The skin and bone marrow biopsy were negative for CD79a CD20, CD 3 and S-100 and positive for Lysozyme, CD43, CD13 and CD68. The diagnosis of chronic myelomonocytic leukaemia (CMML) was made and he was treated with etoposide 50 mg daily. Due to toxicity, the etoposide treatment had to be stopped and the patient died in November 2003 of respiratory insufficiency.

CMML is a haematological malignancy characterised by a wide heterogeneity of clinical presentations and course of disease. In 2001 the World Health Organisation (WHO) re-classified acute myeloid leukaemia (AML), including myelodysplastic syndrome (MDS), chronic myeloproliferative disease (CMPD) and the myelodysplastic/myeloproliferative (MDS/MPD) clonal haematopoietic stem cell diseases.[1] The features of MDS/MPD overlap with those of MDS and CMPD, and CMML was re-classified into MDS/MPD by the WHO. The incidence of CMML is 0.46/100,000. The natural course of CMML is variable, with a median three-year survival of 29%.[2]

The most common presenting symptoms in CMML are caused by bone marrow dysfunction and the resultant peripheral blood cytopenia. CMML has been associated with various dermatological conditions, but rarely involves the skin directly.[3] Skin involvement is often a late stage of the disease. The clinical appearance of leukaemia cutis is highly variable and described as an erythematous maculopapular rash, numerous widespread skin nodules of less than 1 mm in diameter, an unusual localised bullous lesion, and a widespread itchy rash. The most important treatment for CMML is systemic chemotherapy. For cutaneous lesions, however, the addition of whole-body electron-beam irradiation followed by consolidation chemotherapy may be advised.[4]

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