

Two patients with recurrent fever and wine red discolouration of the eyelids

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CASE REPORT

A 65-year-old woman was seen in our outpatient clinic because of recurrent febrile attacks since the age of six years. The medical history revealed appendectomy and diagnostic laparotomy during childhood because of unexplained fever and abdominal pain. The attacks are characterised by spiking fever up to 40°C twice a day, conjunctivitis, abdominal pain, erythematous skin lesions, myalgia, arthralgia especially of hands and legs and an intense acute phase response. During the last episode the erythrocyte sedimentation rate was 100 mm/h, C-reactive protein serum concentration 219 mg/l and leucocyte count $34.2 \times 10^9/l$. The attacks generally last two to three weeks and recur two to five times a year. In between two episodes there are no symptoms. Extended clinical observation, CT scanning of the thorax and abdomen, indium 111-IgG scanning, multiple cultures of body fluids and microscopic examination and cultures of bone marrow did not reveal a diagnosis. Treatment with prednisone (20 mg/d orally) and NSAIDs during attacks alleviated symptoms but had no influence on the duration and recurrence rate of the attacks. The family history revealed that father, brother, brother's daughter and her own daughter experienced similar attacks. Over the past two decades she developed a progressive, slightly elevated wine red discolouration of the skin surrounding the eyes and the eyelids (*figure 1* left panel), which became more prominent and itched during attacks.

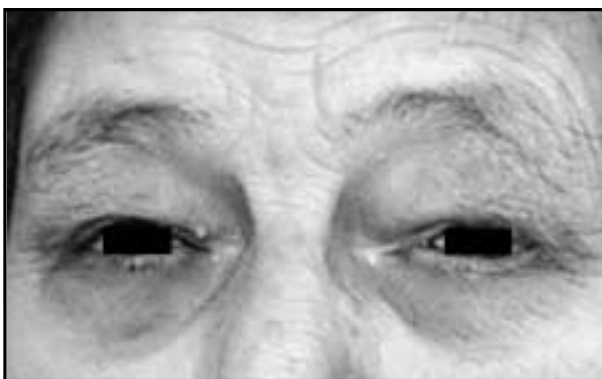


Figure 1

Wine red discolouration of the eyelids and the skin surrounding the eyes (left panel mother, right panel daughter)

A colour version of this photo quiz can be found on our website www.njmonline.nl.

Her now 35-year-old daughter has had similar attacks since the age of four, characterised by spiking fever up to 40°C, myalgia, arthralgia in hands and legs, migrating guirlande-like erythematous cutaneous lesions on the trunk and an acute phase response. These attacks last for two to three weeks and recur two to three times a year. During the most recent visit to our outpatient clinic we observed a wine red discolouration of the eyelids and the skin surrounding the eyes resembling that of her mother (*figure 1*, right panel).

WHAT IS YOUR DIFFERENTIAL DIAGNOSIS?

See page 139 for the answer to this photo quiz.

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