Interactive case report

A 66 year old woman with a rash: case progression

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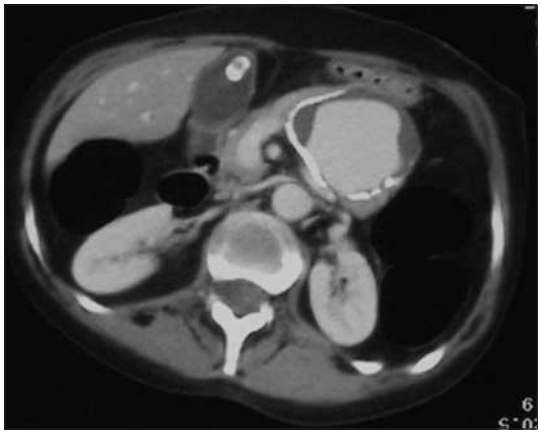
Last week (p 588) we presented the case of Ruth, a 66 year old housewife who presented to her general practitioner with a two to three month history of feeling "out of sorts" and an erythematous papular rash affecting the fingers, dorsum of the hands, knees, elbows, and neck. We invited responses on the likely diagnosis, further investigation, and what to tell the patient. To look at the rapid responses and discussion of the case so far go to bmj.com (http://bmj.com/content/full/326/7389/588).

Ruth was referred to a consultant dermatologist, who agreed that the most likely diagnosis was dermatomyositis. A skin biopsy showed a sparse lymphocytic inflammatory infiltrate and mild upper dermal oedema. Immunofluorescence gave non-contributory and non-specific results: IgG and C3 tested negative, IgA showed spotty coarse intra-epidermal positivity, and there was mild IgM positivity in the basement membrane.

Because of the known association between dermatomyositis and malignant disease (odds ratio 4.4; (95% confidence interval 3.0 to 6.6), Ruth was investigated further.[1] A pelvic examination found some fullness in the right adenexa but no other abnormality. Chest radiography showed no abnormality, and her blood tests, including analyses for urea and electrolytes, thyroid stimulating hormone, cortisol, complement (C3 and C4), and cardiac troponin I, gave normal results. Her creatinine kinase concentration was over 1000 IU/l (reference range 24-273 IU/l) and her C reactive protein concentration was also raised (26.6 mg/l; reference <5 mg/l).

Ruth was told that she had dermatomyositis on the basis of the clinical findings and laboratory results and that in some cases the underlying cause is cancer.[1] She was started on oral prednisolone 30 mg/day and ranitidine cover at 150 mg/day. A mammogram was taken and showed no abnormality. She had computed tomographic colography to assess the colon and check for extracolonic disease in the pelvis. This found no colonic abnormality or pelvic masses but showed a large enhancing lesion continuous with the splenic artery (figure). This was considered to be a splenic artery aneurysm. There was no history of previous trauma or pancreatitis.



Computed tomogram with intravenous contrast of upper abdomen showing large aneurysm of the splenic artery with calcification and some thrombus in the wall

Questions

1 What are the possible risks and benefits related to surgery?

2 Should Ruth have surgery and if so when?

3 What are the long term implications for Ruth from taking steroids and azathioprine at this dose?

Please respond through bmj.com

In the absence of any overt underlying malignancy, Ruth was started on azathioprine for its steroid sparing effect at a dose of 150 mg a day. She was given information about the systemic disturbances that azathioprine can cause and what to look out for and a telephone number to ring. Monthly full blood count and liver function tests were also arranged. A week later she was feeling much better and within a month her creatinine kinase concentration was within normal limits. Her dose of prednisolone was then gradually reduced.

A month later her liver function test results, which were previously normal, showed a raised alanine aminotransferase concentration at 277 IU/l (reference 10-36 IU/l) and alkaline phosphatase at 219 IU/l (reference 40-145 IU/l). Azathioprine was therefore stopped, but she continued taking prednisolone 20 mg daily. Ruth was referred to a vascular surgeon to assess her splenic artery aneurysm. He arranged splenic artery angiography and warned her of the symptoms of a possible aneurysm leak. The risk of rupture was judged to be less than 10%.

Competing interests: None declared.

1 Zantos D, Zhang Y, Felson D. The overall and temporal association of cancer with polymyositis and dermatomyositis. *J Rheumatol* 1994;21:1855-9.

This is part 2 of a 3 part case report where we invite readers to take part in considering the diagnosis and management of a case using the rapid response feature on bmj.com. In three weeks' time we will report the outcome and summarise the responses

Endpiece

A choice

Medicine is my lawful wedded wife and literature my mistress. When one gets on my nerves, I spend the night with the other.

Chekhov

Submitted by Julian Sheather, researcher, London