IMAGE CHALLENGE

A woman with painful lesions of the legs

CLINICAL INTRODUCTION

A 78-year-old woman presented to the emergency department because of weakness and rapidly evolving painful skin lesions. Her medical history was unremarkable, and she was not taking any medications. Physical examination showed irregular violaceous lesions of the right lower limb (figure 1A), knees (figure 1B) and small bullae on the legs (figure 1C). Blood tests were normal; urinalysis showed gross haematuria. An ultrasound of the urinary tract showed an invasive tumour of the bladder.

QUESTION

What is the most likely diagnosis?

A. Sweet syndrome
B. Anthrax
C. Pyoderma gangrenosum
D. Pemphigus foliaceus

For the answer see page 422

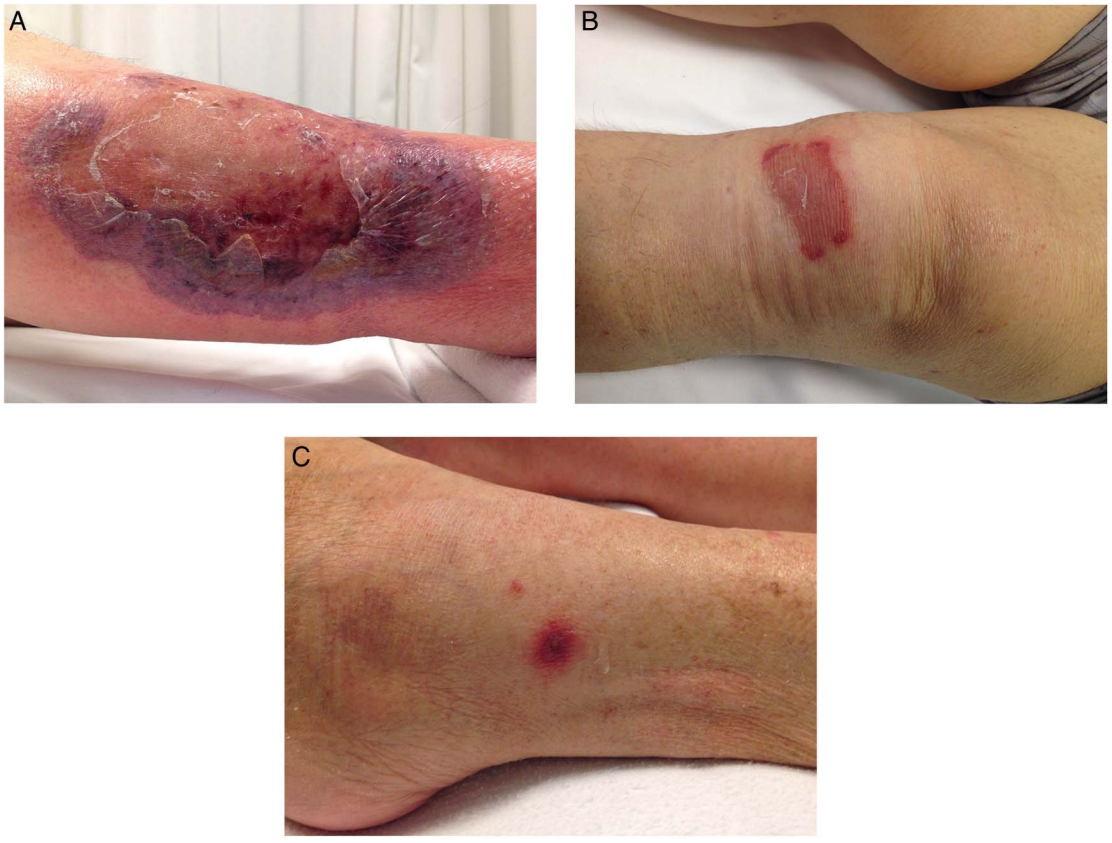

Figure 1 (A) Violaceous and irregular skin lesion of right lower limb. (B) Violaceous and irregular skin lesion of knee. (C) Bullous lesion (ankle).

IMAGE CHALLENGE

A woman with painful lesions of the legs

ANSWER: C

Biopsies of the lesions showed predominantly neutrophilic infiltrate compatible with a diagnosis of *paraneoplastic pyoderma gangrenosum* secondary to *urothelial carcinoma*.

Sweet syndrome would demonstrate fever and cutaneous lesions, usually occur on the upper extremities, face or neck. Cutaneous anthrax is painless, and occurs on upper extremities, head and neck; so, these two diagnoses can be excluded. Pemphigus is unlikely since it usually begins on the face, scalp and later erupts on the chest, back and usually are not painful.

Pyoderma gangrenosum is a rare (incidence has been estimated to range between 1 and 3.3 in 330 000) neutrophilic dermatosis.[1] Violaceous and irregular skin lesions evolve to become pustular, bullous or vegetative. Usually found on the inferior limbs, peristomal or genital variants exist. The aetiology seems to be immunological; more than 70% of cases are secondary to an underlying condition,[2] such as haematological diseases (multiple myeloma, acute myeloid leukaemia, myelodysplastic syndromes or hairy cell leukaemia), inflammatory bowel disease, rheumatoid arthritis or PAPA (pyogenic arthritis, pyoderma gangrenosum and acne) syndrome, which is an autosomal dominant, hereditary auto-inflammatory disease. Paraneoplastic cases are more often multifocal and rarely associated with solid tumours. Prognosis is good, but influenced by the underlying disorders. In mild cases, local treatment with topical steroids and tacrolimus is sufficient. For severe forms, systemic therapy with corticosteroids, mycophenolate mofetil or azathioprine is indicated.[3] The underlying disease has to be treated as well.

The patient was treated with topical steroid, colchicine and surgical resection of the bladder carcinoma. After 3 months' follow-up, the lesions significantly improved in size and depth.

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Contributors HY and FD contributed equally to the diagnosis, management of the patient and writing of this submission.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; internally peer reviewed.

To cite Yildiz H, Dupriez F. *Emerg Med J* 2016;**33**:422.

Accepted 25 August 2015

Emerg Med J 2016;**33**:422. doi:10.1136/emermed-2015-205055

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