Anterior abdominal wall ischaemia in association with multiple myeloma

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INTRODUCTION

This article reports a case of anterior abdominal wall ischaemia in a 52-year-old woman with an underlying diagnosis of multiple myeloma. The patient underwent emergency surgical debridement of the affected area. The abdominal wall was subsequently resurfaced with a split skin graft. Histological examination of the excised tissue revealed the cause to be a paraneoplastic vasculopathy with immunoglobulin light chain (lambda-type) deposition in the blood vessels of her abdominal apron secondary to multiple myeloma. Literature review has failed to show previous reports of this condition. The differential diagnosis and aetiology are discussed.

DISCUSSION

The patient was systemically unwell with clinical evidence of a spreading cellulitis over her anterior abdomen. The rapid progression of the cellulitis prompted early diagnosis of a necrotizing fasciitis and the patient underwent urgent surgical debridement of her anterior abdominal wall. A high index of suspicion is required in such cases, as it is well-recognized that mortality from necrotizing fasciitis is contributed to by failure of early diagnosis and inadequate surgical debridement (Kanoh, 1996).

Radical excision of the ischaemic tissue was the correct treatment. However, histological examination of the excised tissue showed the ischaemia was caused by immunoglobulin light chain deposition in blood vessels of the skin and subcutaneous tissues because of the multiple myeloma, and not resulting from a necrotizing fasciitis as initially diagnosed. The histopathological features absent in this case but seen in necrotizing fasciitis include necrosis of subcutaneous fat and fascia, inflammation of the dermis and subcutaneous fat with perivascular inflammatory infiltrates and intravascular coagulation with surrounding vasculitic changes (Umbert et al, 1989).

There were also certain clinical features and positive laboratory investigations not seen in this case, mitigating against a diagnosis of necrotizing fasciitis. The patient was not in severe pain from her abdomen, whereas pain associated with necrotizing fasciitis tends to be out of proportion to the area of insult. An elevated white cell count is usually seen in the early stage of necrotizing fasciitis but was not raised here, and despite a multitude of microorganisms which have been demonstrated in necrotizing infections, no microorganisms were found in this case.

Patients affected with necrotizing fasciitis often have some underlying risk factors such as malignancy, chronic disease or immunodeficiency, although necrotizing fasciitis has only been reported on one occasion in association with multiple myeloma, in an immunosuppressed patient following chemotherapy (Urschel, 1999).

The skin lesion developed 4 days after re-initiation of warfarin therapy for a deep vein thrombosis in an obese woman and the clinical course is suggestive of warfarin-induced skin necrosis (Ad-El et al, 2000). However, in this case the histological picture was inconsistent with such a diagnosis as there was no evidence of thrombosis within the vessels and no infarction or

CASE REPORT

A 52-year-old woman with widespread multiple myeloma was referred with an 8-hour history of spreading cellulitis with a central dusky purple area over her lower anterior abdominal skin. There was no crepitus or foul watery discharge although the central area was starting to blister. She was awaiting orthopaedic assessment of cervical spine stability because of tumour infiltration at C5 and C6 levels. She was also being treated for a deep vein thrombosis with warfarin. However, the warfarin had been stopped in error 2 weeks previously, and then restarted 4 days before this episode. She had been commenced on warfarin originally because of a pulmonary embolus following the first course of chemotherapy for treatment of her multiple myeloma.

On examination she was noted to be febrile (38.3°C), tachycardic (110 beats per minute) and hypotensive (98/42 mmHg) with spreading cellulitis and a central dusky purple area over her abdominal apron. The area was not exquisitely tender to palpation. Haematological investigations revealed a normal white cell count and an international normalized ratio of 4.1. In view of the clinical picture a provisional diagnosis of necrotizing fasciitis was made.

The patient was already receiving intravenous cefuroxime and vancomycin for a chest infection and this was continued. She was given fresh frozen plasma to restore clotting function, and underwent wide surgical debridement of the area of ischaemic tissue under general anaesthetic.

Microbiological examination of the debrided tissue failed to demonstrate any causative organisms. Histology revealed epidermal spongiosis, dermal oedema and red blood cell extravasation, but no tissue necrosis. A mild perivascular inflammatory infiltrate with no vessel wall damage was also observed. Extensive deposits of diastase-resistant acid Schiff-positive material were present in blood vessel walls and within the lumens (*Figures 1* and *2*). These deposits represent immunoglobulin light chain (lambda-type) deposition caused by multiple myeloma and are responsible for the abdominal wall ischaemia.

Postoperatively the patient remained well with no extension of the cellulitis and subsequently underwent split skin grafting to the abdominal defect from which she made an uneventful recovery.

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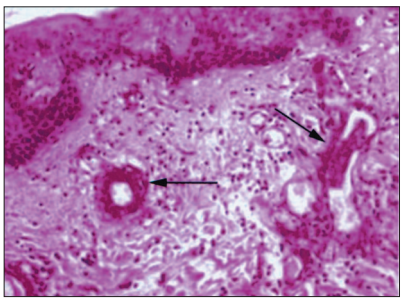

Figure 1. Hyaline deposition in the lumen and wall of small and intermediate blood vessels (diastase periodic acid–Schiff (D-PAS) x200 at 14.5 cm width).

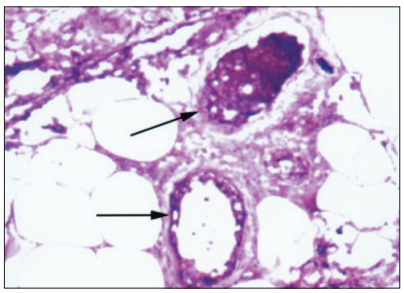

Figure 2. Immunohistochemical staining for immunoglobulin light chain diastase periodic acid–Schiff (D-PAS) positive hyaline material (arrows) in the blood vessel lumen and wall (avidin-biotin complex peroxidase, x400 at 14.5 cm width).

loss of tissue architecture, as one would expect to see in a warfarin-induced skin necrosis.

The clinical picture of an ischaemic anterior abdominal wall may have been exacerbated by a transient type 1 cryoglobulinaemia, which can occur in patients with multiple myeloma (Winfield, 1983), although no cryoglobulins could be seen on repeated assay. Amyloidosis and vasculitis may also result in tissue ischaemia, but histology showed no evidence of these. The ischaemic changes in this case (with a history of multiple myeloma and warfarin therapy) are probably multifactorial in origin. However, the extensive vascular deposition of immunoglobulin light chain (light chain vasculopathy) is the key element in the pathogenesis of the ischaemia observed.

The diagnosis was made on histological examination of the debrided tissue, although an appropriate high index of suspicion of a necrotizing fasciitis led to the correct treatment for an ischaemic area of tissue and a successful outcome for the patient. HM

Ad-El DD, Meirowitz A, Weinberg A, Kogan L, Arieli D, Neuman A, Linton D (2000) Warfarin skin necrosis: local and systemic factors. *Br J Plast Surg* **53:** 624–6

Kanoh T (1996) Multiple myeloma complicated by necrotizing fasciitis. *Rinsho Ketsueki* **37**: 1309–13

Umbert IJ, Winkelmann RK, Oliver GF et al (1989) Necrotizing fasciitis: a clinical, microbiologic and histopathologic study of 14 patients. *J Am Acad Dermatol* **20:** 774–81

Urschel JD (1999) Necrotizing soft tissue infections. *Postgrad Med J* **75:** 645–9

Winfield JB (1983) Cryoglobulinaemia. *Hum. Pathol* **14:** 350–4

In the Public's View...

What a performance!

In September, the government had its hands full of A-levels. Students and teachers suspected that the Department of Education and the Qualifications and Curriculum Authority colluded to lower the grades. The newspapers were full of pretty girls (and less full of spotty boys) complaining that their expected 'A' grades had not materialized. This was probably a good thing for the Department of Health. A day before, a public committee reported a minority of health managers had been fiddling waiting lists, but A-levels took over the front pages.

The only surprise about waiting list fiddles is surprise when they happen. Anyone who has read 'On the unintended consequences of publishing performance data in the public sector' (Smith, 1995) won't be surprised. If you set managers difficult targets, then threaten their businesses with loss of money and them with the sack, they need the ethics of a saint not to apply a bit of imagination. Their desperation is likely to sour relations with the people who do the work, who already feel that their best efforts are merely winning them the chance to work even harder. The situation in hospitals is worsened by factors outside the control of both the workforce and managers. We must meet our surgical targets, but surgical beds are filled with medical patients. We can't get routine operating done, and staff are admonished for not wanting to do extra paid lists at weekends. The government's response is to fly in operating teams from overseas. They tell us that we don't have to worry about the competence of these teams, but no doctor in continental Europe has to go through the appraisal and revalidation hoops that we are. Our Trust seems quite keen on bidding for one of these 'diagnostic and treatment centres', which sounds like a pretty good definition of 'a hospital'.

Smith describes eight consequences of using performance indicators, some of which are seen in our hospitals. Tunnel vision occurs when the organization's objectives (satisfying local health needs) diverge from the measurement scheme (2-week cancer waits). Misinterpretation and gaming occur when the performance data (patients cancelled on the day of surgery) are not processed properly. Smith does not object to indicators, but cautions that they must allow for unintended consequences. Four strategies he suggests include involving staff, which I don't think means pressurizing staff to work even harder.

Sadly, evidence of fiddling is likely to provoke more layers of costly checks (but no balances) from our control-obsessed masters. They ought, but are unlikely, to ask if winding down and accepting some imperfection could be more effective than cranking up.

Widgets per month is a simple measure; total hip replacements per month sounds the same but isn't. Using a simple model for a complex output makes unpleasant perturbations inevitable. And is there any chance of getting my B in Physics (Northern Matriculation Board, 1966) upgraded? HM

Smith P (1995) On the unintended consequences of publishing performance data in the public sector. *Int J Pub Admin* **18:** 277–310

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