CASE REPORT

Severe soft tissue infection masquerading as necrotising fasciitis in a 31-year-old woman with a background of right thigh arteriovenous malformation

Pennylouise Hever,¹ Naveen Cavale,¹ Paul Harnett²

¹Department of Plastic and Reconstructive Surgery, King's College Hospital NHS Foundation Trust, London, UK

²Department of Trauma and Orthopaedic Surgery, King's College Hospital NHS Foundation Trust, London, UK

Correspondence to

Dr Pennylouise Hever, phever@nhs.net

Accepted 9 October 2016

SUMMARY

A 31-year-old woman with a history of a right thigh arteriovenous malformation presented with an acute history of right thigh pain and swelling. This rapidly progressed to gross sepsis. She was intubated, requiring inotropic support and renal replacement therapy. She was considered disproportionately unwell, with the impression one of necrotising fasciitis (NF). She was taken to theatre for emergency exploration and debridement. There was no evidence of NF to note in theatre. Tissue samples cultured group C Streptococcus, with histopathological examination confirming group C Streptococcal soft tissue infection. Thereafter, she was treated with high-dose intravenous antibiotics, use of a negative pressure wound therapy system, and multiple returns to theatre for repeat debridement. Her condition gradually improved, and she was stepped down to a surgical ward 3 weeks after admission. Nine weeks after initial presentation, she underwent skin grafting for wound closure. She returned to work 7 months later.

BACKGROUND

Necrotising fasciitis (NF) is a life-threatening condition requiring prompt diagnosis and urgent debridement to prevent the ongoing sequelae of sepsis, multisystem organ failure and ultimately death. We report a case of severe group C Streptococcus soft tissue infection with accompanying toxic-shock syndrome in a 31-year-old woman presenting with a NF-like clinical picture. The case was complicated by her background of right thigh arteriovenous malformation (AVM), which presented a difficult clinical dilemma—to operate and face the associated risks of massive haemorrhage or to forego urgent debridement in favour of conservative management and risk continued clinical deterioration. The case is the first to describe the management of a severe soft tissue infection presenting on a background of a large AVM.

CASE PRESENTATION

The patient was a 31-year-old woman with a history of a right thigh AVM with vascular and lymphatic components for which she had undergone multiple surgical procedures since childhood. Other significant medical history included Von Willebrand disease (VWD).

She initially self-presented to her local emergency department with an acute history of right

thigh pain and swelling. This rapidly progressed to sepsis, and on transfer to the tertiary centre she was intubated, requiring inotropic support and renal replacement therapy for cardiovascular collapse. Her right hip was warm to touch, erythematous, tense and extremely oedematous (figure 1). There was no crepitus or blistering of the skin and no tenderness to passive stretch. Biochemical and haematological investigations are documented in table 1. Urgent MRI showed signs of deep soft tissue infection (figure 2A, B). Blood cultures were sent, and STAT doses of vancomycin, linezolid and clindamycin were administered after discussion with microbiology, dosed according to the patient's renal function.

TREATMENT

The patient was considered disproportionately unwell, with the impression one of NF. The decision was made to take her to theatre for emergency exploration of her thigh in view of the overwhelming signs of sepsis and likely diagnosis, on the same evening

Figure 1 Clinical examination findings. Black marker demarks area of extension of erythema.

CrossMark

To cite: Hever P, Cavale N, Harnett P. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-216951

Table 1 Significant blood results

Biochemistry	
CRP	163
CK	2263
eGFR	25
Haematology	
WCC	5.6
	Neu 3.9
	Lym 0.3
Hb	89
ABG	
pH	7.34
BE	−11
Lactate	4.1

ABG, arterial blood gas; BE, base excess; CK, creatinine kinase; CRP, C reactive protein; eGFR, estimated glomerular filtration rate; WCC, white cell count.

as presentation. She underwent aggressive wound debridement and wash out with fasciotomy of the thigh as a joint procedure involving the orthopaedic, vascular and plastic surgery teams (figure 3A, B). While we were expecting to see necrotic fascia and/or 'dishwater fluid' in the subcutaneous fat and fascial layers, there was no evidence of NF to note in theatre. The surgery was complicated by postoperative bleeding requiring 4 units of packed red cells and 4 units of fresh frozen plasma. She remained ventilated, receiving ongoing cardiopulmonary support with high doses of norepinephrine. Two days later, she was taken back to theatre for wound re-exploration further to worsening sepsis (lactate of 7) and spreading cellulitis extending to the anterior abdominal wall and perineum. She underwent repeat wound debridement and wash out, with a new incision performed to explore the erythematous suprapubic inflammation. There was still no evidence of NF. In view of ongoing bleeding from her wounds, haematology advice was sought considering the patient's history of VWD. They advised that as all results of VWD screen were high (FVIII level 226, VW Ag 211, VW activity 196.1), VWD was unlikely to be contributing to her bleeding. The extent of her bleeding was therefore felt to be due to her AVM as anticipated rather than inherited coagulopathy. Tranexamic acid was started, and the patient received further transfusions of cross-matched blood.

OUTCOME AND FOLLOW-UP

Tissue samples taken at the time of deep debridement cultured group C *Streptococcus*, with histopathological examination

confirming group C *Streptococcal* soft tissue infection. She continued to be treated with high-dose intravenous antibiotics, use of a negative pressure wound therapy system (NPWT) to minimise tissue oedema and prevent wound expansion and multiple returns to theatre for repeat debridement, wash out and dressing changes. Subsequent tissue samples cultured candida, staphylococcus A and anaerobic streptococci, and antibiotic therapy was adjusted to tazocin, clindamycin and amidulafungin as advised by microbiology. This was later altered to meropenem in line with sensitivities. Her condition gradually improved, and she was stepped down to a surgical ward 3 weeks after admission with 48-hourly NPWT dressing changes. Nine weeks after initial presentation, she underwent skin grafting for wound closure as her wound cavities were too deep, and tissues too oedematous to close directly. She was discharged home after 10 weeks with ongoing NPWT in the community. With ongoing physiotherapy, she has returned to her baseline level of activity, as well as returning to work 7 months after initial presentation. Her wounds continue to heal well (figure 4A, B). The patient signed a consent form authorising this report and use of images.

DISCUSSION

NF is a life-threatening localised infection of the subcutaneous tissues and fascia that may rapidly progress to septic shock and multiorgan failure. It usually begins at the site of trauma or previous surgery, spreading directly across fascial planes, and most commonly presents with pain out of proportion to clinical findings—though presentation is variable which may contribute to delayed diagnosis. Skin changes such as palpable crepitus and blistering are the key clinical diagnostic features; however, these often take days to develop at which time the patient has often progressed to a state of septic shock. Treatment is based on clinical diagnosis and entails radical debridement with the aim to remove all necrotic tissues, arrest the progression of infection and reduce systemic toxicity, with intravenous antibiotics as adjunctive therapy.

The most commonly isolated causative organism is group A *Streptococcus* (type 1 NF), though several organisms have been identified to contribute including non-group A *Streptococci* (including Groups B, C and G), MRSA, aerobic organisms such as *Escherichia coli* and *Pseudomonas aeruginosa* and anerobic organisms such as *Clostridium* (type 2 NF).^{1 2} While group A *Streptococcal* infection has been found to affect individuals with no underlying comorbidities, non-group A *Streptococcal* infection more commonly affects the elderly or chronically ill patient.^{3 4} In our patient—an otherwise well young woman—

Figure 2 (A and B) Sagittal slices of MRI femur (thigh) with Gadolinium Rtc showing a large lobulated infiltrative mass within the right thigh in keeping with a large vascular malformation. Features present which suggest some blood products within and superadded infection as a complication.

Figure 3 Longitudinal incision of the (A) anterior and (B) posterior thigh with oedematous fascia being peeled off the muscles.

we believe it was the patient's AVM which provided a direct nidus for infection, as no cutaneous port of entry was noted. Indeed, vascular malformations have previously been reported as a risk for streptococcal infection in otherwise healthy children^{5 6} and adults.⁷

AVMs are rare congenital lesions, which when peripherally located may present with a range of symptoms including cosmetic symptoms, tissue ulceration, pain, swelling and functional limitations. Our patient presenting with severe soft tissue infection with a clinical picture of NF required urgent surgical debridement. The presence of her AVM raised a difficult clinical dilemma. The AVM's high risk for local haemorrhage, combined with our patient's background of inherited coagulopathy and severe haemodynamic instability at presentation, could have been fatal. Following multidisciplinary discussion however, the decision was made that surgical management was the only viable option for this patient.

In the case of severe soft tissue infection with a potential diagnosis of NF, we therefore propose the best management plan of urgent debridement in spite of concurrent morbidity. However, we urge that this is based on an isolated case report, centred on our experience of events alone. Although there was no evidence of NF in our patient in theatre, the diagnosis of NF is often initially based on clinical diagnosis alone. As a life-threatening condition, we propose aggressive treatment in all patients with a potential diagnosis of NF to maximise outcomes and avoid mortality. We highlight this case in order to make fellow clinicians aware of our experience of a severe soft tissue infection presenting on a background of a severe soft tissue abnormality, which mimicked NF.

Figure 4 (A) Anterior and (B) posterior thigh wounds 5 months after split thickness skin grafting.

Learning points

- ▶ An arteriovenous malformation (AVM) and associated lymphoedema may provide a port of entry for bacteria, leading to severe soft tissue infection.
- ▶ When a patient with a cellulitis or soft tissue infection appears disproportionately unwell, be wary of a diagnosis of necrotising fasciitis (NF).
- ▶ When suspected, NF should be treated aggressively with extensive surgical debridement despite comorbidity.
- ▶ High clinical suspicion, prompt diagnosis and urgent surgical debridement in conjunction with supportive therapy offer the only chance of survival for patients presenting with signs of severe soft tissue infection.
- ▶ The presence of an AVM may confound the diagnosis of a severe soft tissue infection.

Contributors All listed authors made individual contributions to the writing of this article in addition to being involved with the patient's care. PeH was the main contributor responsible for the reporting of the case and will serve as guarantor for the report. NC and PaH supported the planning and development of the initial paper and advised regarding the editors/reviewers comments for the revised draft.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Andreasen TJ, Green SD, Childers BJ. Massive infectious soft-tissue injury: diagnosis and management of necrotizing fasciitis and purpura fulminans. *Plast Reconstr Surg* 2001;107:1025–34.
- 2 Wong CH, Chang HC, Pasupathy S, *et al*. Necrotizing fasciitis: clinical presentation, microbiology, and determinants of mortality. *J Bone Joint Surg Am* 2003;85-A:1454–60.
- 3 Cox NH. Streptococcal necrotizing fasciitis and the dermatologist. *Br J Dermatol* 1999;141:613–14.
- 4 Hakkarainen TW, Kopari NM, Pham TN, *et al*. Necrotizing soft tissue infections: review and current concepts in treatment, systems of care, and outcomes. *Curr Probl Surg* 2014;51:344–62.
- 5 Russell S, Watts RG, Royal SA, *et al*. Group A streptococcal infection of an intramuscular venous malformation: a case report and review of the literature. *Pediatr Emerg Care* 2008;24:839–41.
- 6 Rausch J, Foca M. Necrotizing fasciitis in a pediatric patient caused by lancefield group G streptococcus: case report and brief review of the literature. *Case Rep Med* 2011;2011:671365.
- 7 Binyamini S, Ergas D, Herskovitz PI, *et al*. Pulmonary arteriovenous malformation associated with necrotizing fasciitis of the thigh. *Am J Med Sci* 2008;336:527–9.

Copyright 2016 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow