CASE REPORT

Purpura fulminans in a patient with mixed connective tissue disease

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SUMMARY

A 43-year-old lady was admitted to the intensive care unit with sepsis. She had a history of mixed connective tissue disease, Raynaud's syndrome and hypothyroidism. 2 days later, she developed a purpuric rash on her face and extremities with a livedoid background. Few days later, her distal fingers and toes became gangrenous which then had to be amputated. Laboratory investigations showed that she was coagulopathic and had multiple organ dysfunctions. Antiphospholipid antibodies were negative; however, protein C and antithrombin III levels were low. A skin biopsy showed fibrinoid necrosis in the vessel wall with microthrombi and red-cell extravasation. A diagnosis of purpura fulminans was made.

DIFFERENTIAL DIAGNOSIS

- ▶ Disseminated intravascular coagulation
- ▶ Necrotising fasciitis
- ▶ Cutaneous vasculitis

TREATMENT

This patient was treated with protein C concentrate (100 IU/kg initial dose followed by 60 IU/kg twice a day) in addition to her treatment for sepsis. Her sepsis improved and there was no further spread of the tissue necrosis on her limbs. She was also vaccinated against encapsulated bacteria following the discovery of her hyposplenism.

OUTCOME AND FOLLOW-UP

She recovered fully apart from her gangrenous digits that had to be amputated.

BACKGROUND

Purpura fulminans is a medical emergency that requires early recognition and targeted treatment. Delay in therapy would lead to necrosis of the affected body. Multidisciplinary approach in the management of these patients is recommended for a better outcome.

CASE PRESENTATION

A 43-year-old lady was admitted to intensive care unit with sepsis. She had a history of mixed connective tissue disease, Raynaud's syndrome and hypothyroidism. Two days later, she developed a purpuric rash with a livedoid background on her face (figure 1) and extremities. A few days later, her distal fingers and toes became gangrenous (figures 2 and 3).

INVESTIGATIONS

Laboratory investigations showed that she was coagulopathic and had multiple organ dysfunctions. Protein C and antithrombin III levels were low. Antiphospholipid antibodies were negative.

A skin biopsy showed fibrinoid necrosis in the vessel wall with microthrombi and red-cell extravasation (figure 4). Direct immunofluorescence showed deposition of fibrinogen and C3 in the vessels (figure 5).

An active search for the cause of septicaemia revealed a positive PCR to pneumococcal antigen. On further investigation, she was found to have hyposplenism following the detection of Howell Jolly bodies on her blood film. Her clinical presentation and histological features were consistent with purpura fulminans secondary to pneumococcal sepsis.

DISCUSSION

Purpura fulminans is a rare syndrome of intravascular thrombosis and haemorrhagic infarction of the skin that is rapidly progressive and is accompanied by vascular collapse and disseminated intravascular coagulation. It was first described by Guelliot in 1884. It usually occurs in children but this syndrome has been described in adults and especially in those who are immunocompromised. Neonatal presentation is associated with hereditary deficiency of protein S, protein C and antithrombin III, whereas the idiopathic subtype is commonly associated with sepsis. Pneumococcus, meningococcus and staphylococcus aureus¹ are common microorganisms that have been associated with purpura fulminans. It has also been reported to be a rare complication after chickenpox infection.²

Figure 1 Non-blanching, purpuric rash on the cheeks, nose and periorally.

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Figure 2 Gangrenous distal fingers with oedema and erythema at the proximal margins.

The balance of anticoagulant and procoagulant endothelial cell activity is altered in purpura fulminans. This is precipitated by bacterial endotoxin and mediated by various factors that include the inflammatory cytokines interleukin-1, interleukin-12, interferon- γ and tumour necrosis factor- α , which consume antithrombin III as well as proteins S and C. Microemboli and direct bacterial damage to vessels have also been linked with this process. Protein C, when activated, is a vitamin K-dependent serine protease that has anticoagulant and profibrinolytic activities. Secondary protein C deficiency occurs during disseminated intravascular coagulation and sepsis. Small-sized and medium-sized blood vessels may be occluded with thrombus and cause multiorgan failure. Purpura fulminans can also represent a cutaneous marker of protein C deficiency.³ Sometimes, a fulminant pneumococcal sepsis may be the first clinical manifestation of an unknown splenic hypoplasia,⁴ as in our patient's case. A fatal pneumococcal sepsis has been reported in a patient with mixed connective tissue disease due to unsuspected hyposplenism.⁵ Therefore, regular monitoring of blood film in high-risk patients is important to detect early hyposplenism and start appropriate prophylaxis against infection. Early recognition and treatment of purpura fulminans are essential to reduce mortality and to prevent long-term sequelae.

Figure 3 Gangrenous distal feet with oedema and erythema at the proximal margins.

Figure 4 Skin histology showing fibrinoid necrosis in the vessel wall with microthrombi and red cell extravasation.

Figure 5 Direct immunofluorescence showing deposition of fibrinogen and C3 in the vessels.

Management is by treating the underlying cause and the coagulopathy. Surgical intervention is sometimes required.

Learning points

- ▶ Early recognition and treatment of purpura fulminans are essential to reduce mortality and to prevent long-term sequelae.
- ▶ Regular monitoring of blood film in high-risk patients is important to detect early hyposplenism and start appropriate prophylaxis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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