Purpura Fulminans and Disseminated Intravascular Coagulation secondary to Invasive Pneumococcal Disease in an Immunocompetent Individual.

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Background

Purpura fulminans is a rare, life-threatening, rapidly progressive condition of microvascular thrombosis leading to purpuric lesions and haemorrhagic necrosis often in the setting of DIC. Despite aggressive management of sepsis, this condition still carries a 43% mortality rate[1] and represents a dermatological emergency. It is typically associated with encapsulated endotoxin producing bacteria. This case represents a severe example, secondary to Invasive pneumococcal disease.

Case

A 44-year-old female presented to the emergency department with a three-day history of headache, vomiting and pyrexia. She presented in septic shock with a temperature of 40°C, tachycardia of 110bpm and hypotension of 78/55mmHg and had no signs of meningism on exam. She was a current smoker with a 15 pack year smoking history. She had a CRP of 148mg/L and WCC of 10x10^9/L and was started on broad-spectrum antibiotics. She also had refractory hypoglycaemia, (lowest value 1.9 mmol/L), requiring multiple rounds of treatment, and a rising lactate (highest 5.9 mmol/L).

A reticular/reticuloform purpuric rash developed on her legs with mottling of skin on the tips of her nose and ears.

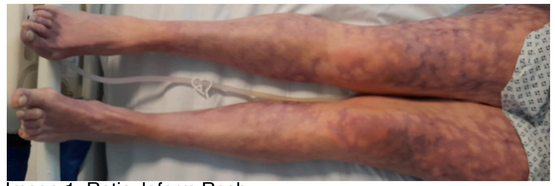

Image 1: Reticuloform Rash

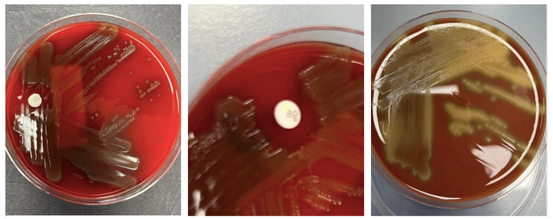

Image 2: Patients' plates - Gram positive Cocci on blood (x2 images) and chocolate agar, with sensitivity to optochin disk.

Within six hours blood cultures flagged positive for Gram Positive Cocci. This was later confirmed to be *Streptococcus Pneumoniae* (Serotype 22F). Clinical features of the rash, combined with histological findings of microthrombi on skin biopsy, confirmed a diagnosis of purpura fulminans. This occurred in the context of a developing Disseminated Intravascular Coagulation (DIC), diagnosed by Isth's criteria on day 2 of admission (See table below). She had a comprehensive coagulopathy, immunological and vasculitic screen which showed acute phase changes consistent with sepsis and DIC including a transient low C3 but no underlying predisposition to severe disease was found.

Marker	Day 0	Day 1	Day 2
Platlets x10^9/L	219	91 ↓	33 ↓
D-Dimer	2,466 ↑	> 5,250 ↑	> 5,250 ↑
PT	16.9	21.5	10.1
INR	1.4	1.8 ↑	0.9
Fibrinogen g/L	2.7	2.2	1.4↓

Table 1: Isths criteria for Disseminated Intravascular Coagulopathy.

Management

Concurrent meningitis was not definitively excluded. As such, her invasive pneumococcal disease was she was treated with meningitic dose Ceftriaxone 2g BD, Vancomycin and Dexamethasone 10 mg QDS IV for 4 days in line with guidelines[2]. Antibiotic treatment was rationalised to Ceftriaxone monotherapy once sensitivities were available. She received supportive care in the Intensive Care unit with stringent skin care and pain management, and did not require vasopressors. A DOAC was started for microvascular thrombosis when platelet counts recovered under the guidance of the Vascular team. She recovered well from the acute phase of her illness; however, she developed critical limb ischaemia with bilateral lower limb dry gangrene requiring a left transmetatarsal amputation.

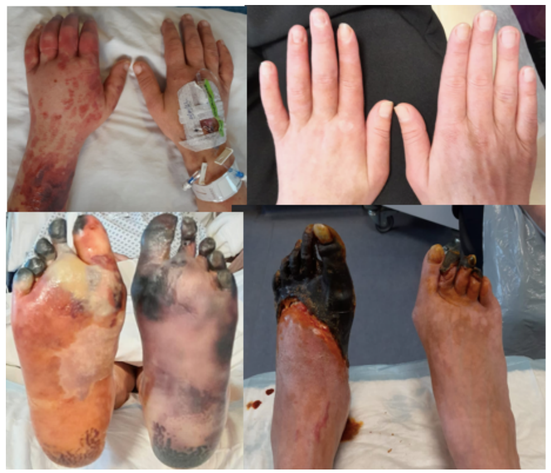

Image 3: Clinical photography at 7 days (left) and 3 months (right).

Discussion

This case demonstrates the devastating sequela of sepsis including, septic shock, purpura fulminans, DIC, hyperlactatemia, and hypoglycaemia. Recent sinusitis in the preceding two weeks was the likely source of initial *Streptococcus Pneumoniae* infection, with smoking the only risk factor identified for invasive pneumococcal disease. Research suggests that cigarette smoke can have a key role in the pathogenesis of invasive pneumococcal disease by affecting both biofilm formation and the bioactivity of pneumolysin which may promote both colonisation and persistence[3]. The serotype identified in this case was 22F which is vaccine preventable with the Pneumococcal Polysaccharide Vaccine (PPV23). The Centre for Disease Control has recently expanded their recommended recipients of the PPV to include current smokers. However, this has not yet incorporated into Irelands recommendations and warrants future consideration.

References:

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