Purpura Fulminans: Not Always Sepsis

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Background:

Purpura fulminans (PF) is a rapidly progressive, potentially fatal, thrombotic disorder. Three categories of PF are recognised: acute infectious (usually associated with sepsis), congenital and idiopathic (1). Meningococcal sepsis was the commonest cause until the introduction of vaccination. The differential diagnosis is wide and a multidisciplinary approach to management is required, including paediatric haematology and rheumatology, microbiology and plastic surgery.

Case Summary

An 8-year-old boy presented with rapidly extending PF lesions (Figure 1) following four days of being generally unwell with fever and diarrhoea. He was treated for suspected sepsis, intubated and ventilated and transferred to paediatric intensive care. He was not shocked.

Laboratory tests demonstrated consumptive coagulopathy (PT 22s, APTT 40s, Fibrinogen 0.37g/L, D-Dimer 47000ng/mL), acquired protein C deficiency (30 U/dL, ref range > 60.0 U/dl), and normal protein S antigen (94 U/dL, normal > 73U/dl) and antithrombin levels. He was treated with antibiotics and analgesia, fresh frozen plasma (FFP), cryoprecipitate and a heparin infusion.

After ten days new PF lesions developed along with compartment syndrome in the left forearm (Figure 2). He underwent emergency fasciotomy. He remained anticoagulated and was treated with high dose methylprednisolone, plasmapheresis with FFP replacement, rituximab and mycophenolate mofetil. He made a good recovery.

He was investigated for haematological, microbiological and immunological causes. The final diagnosis was idiopathic PF secondary to a streptococcal infection with thrombotic storm, possibly immune mediated (ASOT titre 1600 units/mL, Anti DNAs B 1200 units/mL * high level*).

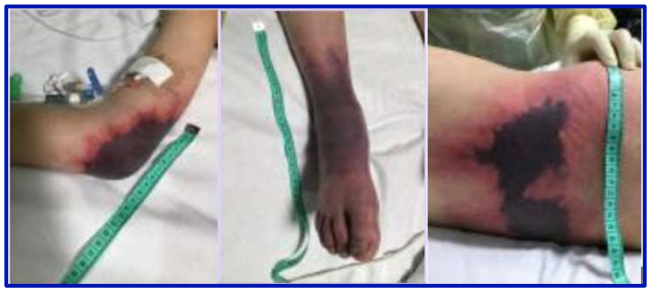
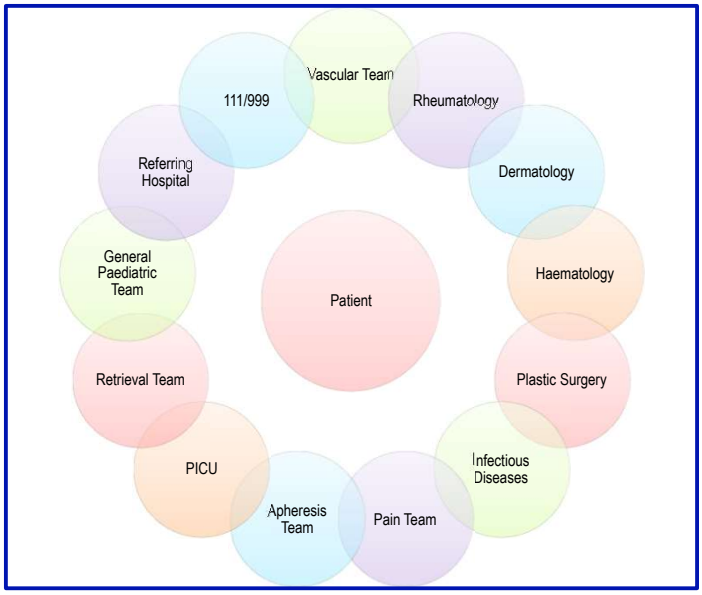
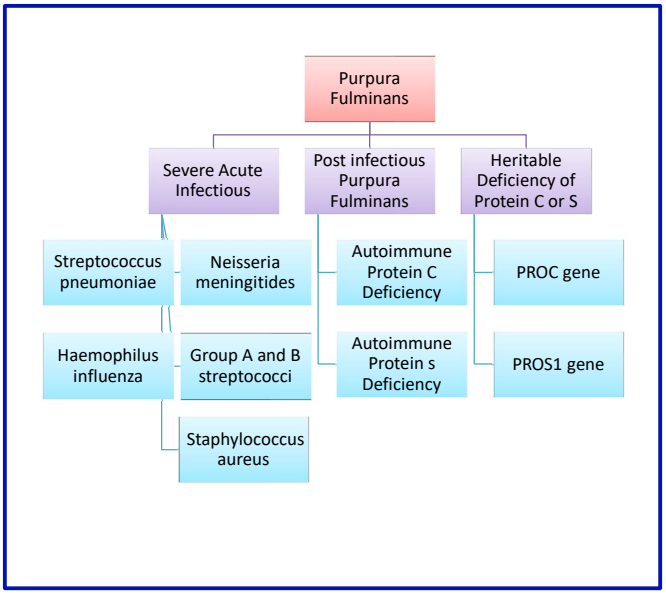


Figure 1. A: right elbow, B: right foot, C: lumbar sacral region

Figure 2. A: left elbow, B: left elbow, C: left elbow pre-op



Teams involved in clinical journey of the patient



Differentials for Purpura Fulminans (*adapted from Chalmers et al., 2011*)

Conclusion:

Idiopathic PF is extremely rare and often preceded by infection with varicella or streptococcus (1). Diagnosis can be challenging. Good supportive care, replacement of coagulation factors, anticoagulation and immune modulation are the mainstays of treatment.

Take-home Message: It can be difficult to immediately exclude sepsis in patients with purpuric lesions, a multi- specialty approach to care is essential to ensure appropriate baseline/on-going investigations and treatments

References: 1. Chalmers E, Cooper P, Forman K, Grimley C, Khair K, Minford A, Morgan M, Mumford AD. Purpura fulminans: Recognition, diagnosis and management. *Arch Dis Child*. 2011; 96:1066-71.

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