

Purpura on the truncus and extremities

CLINICAL INTRODUCTION

A 36-year-old woman with Down's syndrome and spleen hypoplasia presented with a fever of 40°C lasting for 12 h. She had taken 1 mg prednisolone/day for mild graft-versus-host disease after allogeneic stem cell transplantation 4 years previously. Her BP was 91/44 mm Hg, HR 110 beats/min and RR 30 breaths/

min. Her heart sound, lung sound and skin appearance were unremarkable. Laboratory findings showed leucocytosis ($13.12 \times 10^9/L$) and normal platelet count ($269 \times 10^9/L$). She was discharged home, but 16 h after fever onset, experienced cardiopulmonary arrest.

QUESTION

What does the image show?

- A. Henoch–Schönlein purpura
- B. Steroid purpura
- C. Postmortem lividity
- D. Purpura fulminans

ANSWER: D

DIAGNOSIS: acute infectious purpura fulminans (PF) caused by *Streptococcus pneumoniae*.

Figure 1 shows systemic impalpable purpura that rapidly emerged on the patient's truncus gravity-independently. Gram staining of her peripheral blood revealed gram-positive diplococci (figure 2, arrows), indicative of acute infectious PF caused by *S. pneumoniae*. Despite strenuous resuscitation and broad-spectrum antibiotic administration, she died. Her blood culture results subsequently tested positive for *S. pneumoniae*.

PF is a rare, fatal syndrome accompanied by skin necrosis and disseminated intravascular coagulation, caused by protein C-deficiency or overwhelming sepsis,¹ typically caused by *Neisseria meningitidis* and *S. pneumoniae*.² This patient had



Figure 1 The patient's appearance on readmission.

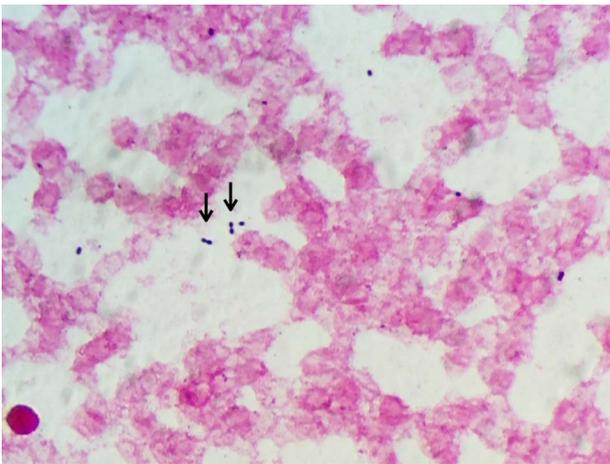


Figure 2 Gram-stained sample of the patient's peripheral blood. The arrows suggest typical gram positive diplococci.

hyposplenism, a risk factor for invasive *N. meningitidis* and *S. pneumoniae* infection.³ Systemic purpura and a rapid,

progressive clinical course is indicative of PF, but obtaining blood culture results to confirm infection requires several days. Gram staining of a peripheral blood sample may be useful for identifying causative bacteria, guiding an early decision to use empirical antibiotics.

Henoch-Schönlein purpura is rare and usually benign in adults, and is typically characterised by elevated palpable purpura and abdominal pain. Long-term corticosteroid administration causes steroid purpura, but never causes cardiopulmonary arrest (CPA). Postmortem lividity generally appears 15 h after death; this patient was immediately taken to the hospital upon CPA; therefore, postmortem lividity was not a consideration.

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