

# Pneumococcal Waterhouse-Friderichsen syndrome despite a normal spleen

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**Summary:** A previously fit 56 year old female presented with a rapidly progressive and fatal pneumococcal septicaemia with disseminated intravascular coagulation. Post-mortem studies confirmed a Waterhouse-Friderichsen syndrome and revealed an anatomically normal spleen; intracellular diplococci were seen within splenic macrophages providing evidence of normal splenic function. This appears to be only the second case of Waterhouse-Friderichsen syndrome due to pneumococcal septicaemia in a patient with a normal spleen.

## Introduction

The association between pneumococcal septicaemia and disseminated intravascular coagulation (DIC) in patients with asplenia or abnormal splenic function is well recognized, with post-mortem evidence of a Waterhouse-Friderichsen syndrome being reported in some patients.<sup>1</sup> We report a case of rapidly overwhelming pneumococcal septicaemia and DIC with bilateral adrenal and ovarian haemorrhages in a patient with a histologically normal spleen.

## Case report

A 56 year old female nurse without any significant previous medical history was admitted with a 24 hour history of malaise, nausea, vomiting, and diarrhoea. On the morning of admission she complained of myalgia and occipital headache.

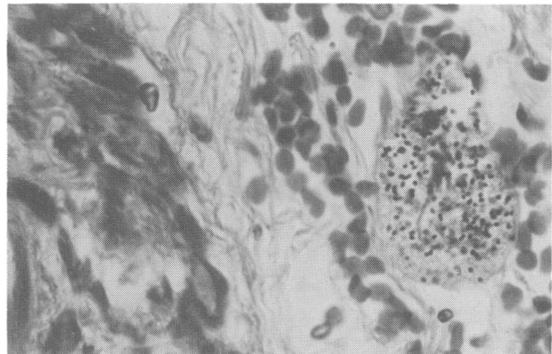
On examination, she was severely ill, pyrexial (38.2°C), dehydrated, with purpuric spots on her abdomen and livedo reticularis on the limbs.

Initial investigations showed: haemoglobin 14.0 g/dl, white cell count  $10.0 \times 10^9/l$ , platelets  $40 \times 10^{12}/l$ , plasma sodium 129 mmol/l, potassium 3.8 mmol/l, bicarbonate 16.9 mmol/l, urea 10.0 mmol/l, creatinine 233  $\mu\text{mol}/l$ , amylase 82 units/dl (normal = 38–162), blood glucose 4.0 mmol/l, prothrombin ratio 4.7 (normal < 1.2), partial thromboplastin time 135 seconds (normal = 30–45), thrombin time 70 seconds (control 14), fibrinogen degradation products 200 mg/l (normal < 10), fibrinogen 0.89 g/l (normal 1.5–4.0).

Blood gases and chest X-ray were normal. A diagnosis of septicaemia with consumption coagulopathy was made and treatment with high dose intravenous benzyl penicillin, hydrocortisone and fresh frozen plasma was commenced.

Over the next hour rapid deterioration occurred with the rash spreading to cover the trunk and face and the patient became centrally cyanosed. Despite intensive treatment the patient became hypotensive and her conscious level deteriorated. Focal and then generalized convulsions followed, with death occurring 4 hours after initial presentation.

Ante-mortem blood cultures, taken prior to antibiotic therapy, were all positive for *Streptococcus pneumoniae*. Post-mortem examination demonstrated widespread visceral petechiae. The adrenal glands showed the classical appearances of the Waterhouse-Friderichsen syndrome. There were extensive bilateral



**Figure 1** Section of spleen ( $\times 500$ , haematoxylin and eosin) showing intracellular diplococci within a macrophage.

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ovarian haemorrhages. The spleen (170 grams) appeared grossly normal. Microscopy of the spleen showed an increased number of polymorphonuclear leucocytes within the red pulp, and diplococci of *S. pneumoniae* were seen within macrophages (Figure 1). Phagocytosis provided contributory evidence of normal splenic function. Liver histology showed multiple microabscesses within the liver parenchyma.

### Discussion

This case illustrates the high mortality associated with *S. pneumoniae* septicaemia complicated by DIC. Early diagnosis and treatment are essential in order to alter the poor prognosis of these patients.

The association of Waterhouse-Friderichsen syndrome with pneumococcal septicaemia is well described

in patients with previous splenectomy,<sup>1</sup> congenital asplenia,<sup>2</sup> acquired hyposplenism<sup>3</sup> and immunosuppression. However, only one previous case has been reported where a histologically normal spleen has been found in fulminant pneumococcal infection with proven Waterhouse-Friderichsen syndrome.<sup>4</sup>

This case presents other unusual histological features: bilateral ovarian haemorrhages, multiple microabscesses within the liver parenchyma and the demonstration of intracellular diplococci in the spleen. It was later discovered that the patients' mother had died of pneumococcal pneumonia 10 days previously. Unfortunately the pneumococcal type was not identified in the mother, so we can only speculate that the exposure to a large antigen load as the patient nursed her mother might have been significant in this case.

### References

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