Lesson of the Week

Chest infection associated with the Waterhouse-Friderichsen syndrome

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The Waterhouse-Friderichsen syndrome is well recognised in association with meningococcal septicaemia and is characterised by an acute pyrexial illness, petechial skin rash, peripheral circulatory failure, and bilateral adrenal haemorrhage.¹ We report on three patients who had had chest infections and collapsed and died unexpectedly. Each fulfilled the diagnostic criteria of the Waterhouse-Friderichsen syndrome.

Case reports

Case 1—A 47-year-old woman attended her general practitioner with a short history of a cough and purulent sputum and was prescribed oxytetracycline 250 mg to be taken four times daily. The following day the patient collapsed while visiting a local art gallery and was dead on arrival at hospital. Necropsy examination showed scattered petechial haemorrhages on the legs and early pneumonic consolidation, from which a moderate growth of pneumococcus was cultured. Culture of a "splenic stab" was sterile. Both adrenal glands showed massive intracortical haemorrhage, and histological studies showed fibrin thrombi in the small vessels of the adrenal capsule, renal cortex, and skin.

Case 2—A 69-year-old man, complaining of pleuritic chest pains and mild dyspnoea, requested a home visit from his general practitioner. A "chest infection" was diagnosed and treatment was started with ampicillin 500 mg four times daily. Later that night the local deputising service was asked to attend because his condition suddenly deteriorated. The patient was pyrexial (temp 37.1°C), with a pulse rate of 180/min and a systolic blood pressure of 75 mm Hg. The respiratory rate was 41/min. Hospital admission was arranged, but the patient died 45 minutes later. Results of the emergency investigations that were done after death included a platelet count of $110 \times 10^9/l$ ($110\,000/mm^3$) and fibrin degradation products of 64 ng/ml. Blood culture was sterile. Necropsy examination showed occasional petechial haemorrhages on the abdomen and confluent bronchopneumonia. Culture showed a heavy growth of *Staphylococcus pyogenes*. Both adrenal glands showed massive intracortical haemorrhage, and histological studies showed fibrin thrombi in the adrenal cortical and renal capillaries.

Case 3—An 89-year-old man was admitted to hospital with dysphagia. Investigations showed a benign oesophageal stricture, a hiatus hernia, and a pharyngeal pouch. The patient developed bronchopneumonia, and a sputum culture grew *Haemophilus influenzae*. Treatment was started with ampicillin 500 mg four

Sudden collapse in a patient with chest infection and skin petechiae may be a symptom of the Waterhouse-Friderichsen syndrome

times daily. Despite gradual clinical improvement the patient suddenly became hypotensive, with a systolic blood pressure of 70 mm Hg, and died two hours later. Small petechial haemorrhages were noted on the left arm. Blood culture was subsequently reported as sterile. Necropsy examination confirmed the presence of bronchopneumonia and the adrenal glands showed massive bilateral haemorrhage.

Comment

In the early 1900s the association of sudden death, skin purpura, and bilateral adrenal haemorrhage was noted in a wide range of clinical conditions.²⁻³ Over the years, however, the so-called Waterhouse-Friderichsen syndrome has been largely associated with meningococcal septicaemia. Interestingly, no cases of adrenal haemorrhage associated with fatal meningococcal infection were seen in this hospital during the 12 months that our cases were collected. Although the cause of the haemorrhage in many reported cases remains unclear, it is likely that in two of our cases it was a result of disseminated intravascular coagulation.

There has been considerable debate about the degree of functional adrenal insufficiency in patients with the Waterhouse-Friderichsen syndrome, and it may be that factors such as endotoxic shock and a generalised Schwartzmann reaction contribute to death. It is well documented, however, that dramatic clinical improvement and survival may follow treatment with cortisone.⁴

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References

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