Waterhouse-Friderichsen syndrome without purpura due to *Haemophilus influenzae* group B

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Summary: A second recorded case of Waterhouse-Friderichsen syndrome, without purpura, due to *Haemophilus influenzae* is described. It is suggested that the absence of purpura should not preclude the diagnosis of the Waterhouse-Friderichsen syndrome due to this organism.

Introduction

The Waterhouse-Friderichsen syndrome is a well recognized complication of meningococcal infection. It is less commonly seen in association with infections due to other organisms including *Haemophilus influenzae* (Beach *et al.*, 1979). Purpura is an important but not consistent feature of the syndrome. We describe a case of Waterhouse-Friderichsen syndrome without purpura in a child with *Haemophilus influenzae* infection, the second report of such a case (Ginandes and Howard, 1947).

Materials and methods

Case report

A five month old female Caucasian child presented with a 24 h history of offensive loose stools and an 18 h history of slow feeding, vomiting, pallor and grunting respirations. On admission she was severely ill, febrile (38.9°C), with tachycardia (200/min) and a respiratory rate of 14/min. She was centrally cyanosed, unresponsive to all stimuli, with absent tendon jerks. The liver was palpable 2 cm below the costal margin and the spleen tip was palpable. The anterior fontanelle was normal, the pupils were equally dilated and reacted slowly to light. The skin was mottled without a rash. She had been premature (birth weight 994 g) with

uneventful post-natal development and had no intervening illness.

A diagnosis of septic shock was made. Treatment was started with plasma infusion, intravenous penicillin and gentamicin, hydrocortisone, metaraminol, dopamine, sodium bicarbonate, oxygen and ventilatory support.

Laboratory investigations showed haemoglobin 10.5 g/dl, and white cell count $4 \times 10^9/l$ with 10% neutrophils, 87% lymphocytes and 3% monocytes. Blood urea was 10.1 mmol/l, serum sodium 139 mmol/l and potassium 7.0 mmol/l. The electrocardiograph showed tachycardia and hyperkalaemic changes. Chest X-ray showed no abnormality. In spite of treatment she died 2 h after admission.

Faecal culture subsequently grew no enteropathogens and blood culture was sterile after five days incubation.

An immediate post-mortem lumbar puncture was performed. The cerebrospinal fluid contained 13 white blood cells per cu.mm with 30% polymorphs and 70% lymphocytes. Gram stain showed no organisms. Culture of the fluid grew *Haemophilus influenzae* Group B sensitive to ampicillin and chloramphenicol.

Pathology

At autopsy there was no evidence of a skin rash. Both adrenals were diffusely haemorrhagic. Microscopic examination of haematoxylin and eosin stained sections of heart, thymus, brain and spinal cord were normal. The meninges showed no evidence of acute inflammation. There was extensive haemorrhage into the adrenal cortex bilaterally. There was pale pink fibrillar material which stained positively with stains for fibrin (phosphotungstic acid, haematoxylin and

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marius scarlet) within the capillaries of glomerular tufts, adrenal cortex, lung and liver sinusoids. The anatomic diagnosis was that of disseminated intravascular coagulation associated with generalized septicaemia and Waterhouse-Friderichsen syndrome.

Discussion

Six cases of Waterhouse-Friderichsen syndrome associated with *Haemophilus influenzae* infection have been reported (Beach *et al.*, 1979). Purpura was absent

in one of these cases. The features of this syndrome (Fox, 1981) are: sudden onset of illness with rapid progression; shock; cyanosis; petechial rash; death usually within 24 h, and haemorrhages into both adrenals.

It would seem from the report of Ginandes and Howard (1947) and the present case that a petechial rash should not be regarded as an essential feature of the syndrome when caused by *Haemophilus influenzae*. *H. influenzae*, one of the causes of septic shock in infancy, may give rise to a complicating Waterhouse-Friderichsen syndrome in the absence of purpura.

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