

Pathology Biology Section - 2011

G34 Epidemic Outbreak of Meningococcal Meningitis in a Nursery: Two Fatal Cases of Waterhouse-Friderichsen Syndrome

Alessandro Bonsignore, MD*, Francesca Fossati, MD, Federica Portunato, MD, Francesco Ventura, PhD, and Francesco De Stefano, MD, Department of Legal and Forensic Medicine, University of Genova, Via de Toni 12, Genova, 16132, ITALY

The goal of this presentation is to focus on two fatal cases of undiagnosed meningitis occurring simultaneously in two children from the same nursery. A forensic approach by means of autopsy, microscopic examination, and microbiological studies led to the conclusion that the cause of death in both infants was septic shock due to meningococcal meningitis in association with hemorrhagic adrenalitis.

This presentation will impact the forensic science community by demonstrating how important a thorough forensic investigation is to reach the correct postmortem diagnosis, as well as, by showing how rapidly children can develop a fatal meningococcal infection as well as explaining the importance of an early clinical diagnosis in order to avoid unexpected death and epidemic outbreaks.

Waterhouse-Friderichsen syndrome (WFS), first described in the early 1900s in England and Denmark, is the most severe form of meningococcal septicemia. The infection leads to massive hemorrhage in one or usually both adrenal glands. It is most commonly caused by *Neisseria meningitidis* (NM) but many other species of bacteria and also viruses are associated with WFS.

The onset of a meningococcal infection is non-specific with symptoms of fever, rigor, vomiting, and headache. Soon a rash appears; first macular, then rapidly becoming petechial and purpuric. In most cases the resulting hypotension rapidly leads to septic shock. In WFS, meningitis generally does not occur but if present, many clinical signs can be found such as hypoglycemia with hyponatriaemia and hyperkalemia, thrombocytopenia and typical markers of diffuse intravascular coagulation.

Only microbiological studies can lead to the final diagnosis through culturing of blood or cerebrospinal fluid (CSF).

Fulminate meningococcemia is a medical emergency and needs to be treated with adequate antibiotics as fast as possible, also in order to prevent an epidemic outbreak. The administration of corticosteroids can sometimes reverse the adrenal shock.

Case 1: A 21-month-old child, previously in good health, developed high fever (40°C) on a Sunday morning. Paracetamol was administered twice during the day but both times with a low response. Few hours after the onset of the fever, the child began to vomit. In the evening the parents noticed a red\black "purpura" on the abdomen and the back. However, at the time, the child seemed to feel better, ate with a good appetite, and was afterward sleeping normally. Early the next morning the father found him lifeless in his bed.

Case 2: A healthy 19-month-old child, later discovered to be taken care of in the same nursery as the previous child, had a very similar clinical history. On the same Sunday afternoon, he developed high fever (39°C) and was treated with paracetamol but with a weak response. The next morning, after a normal night's sleep, he suddenly started to vomit and became cyanotic. The parents immediately called an ambulance but the baby died on the way to the hospital.

Complete postmortem autopsy of both children were performed 24 hours after death. Gross examinations revealed that they were age- accordingly developed. They were covered with purple petechial spots all over the body but no other remarkable external findings were observed.

Autopsies showed cerebral oedema and venous congestion, diffuse whitish and milky subpial exudation, adrenal glands with massive hemorrhagic infiltration of the parenchyma, and polyvisceral stasis. No other significant abnormalities were found.

The macroscopic appearance led to the suspicion of meningeal infection and hence, CSF, buccal, pharyngeal, and nasal swabs, as well as blood samples were taken for microbiological studies. These showed NM DNA positive for NM serotype B. Furthermore, all samples were culture positive after 72 hours of incubation. No other pathogenic agents were present.

The microscopic histological study, performed by using formalin-fixed paraffin embedded tissue sectioned at 4 μ m and stained with haematoxylin-eosin, revealed subpial and cortical oedema, mild inflammatory infiltration along penetrating, deep brain vessels. The adrenal glands showed massive hemorrhagic infiltration, the lungs focal oedema, and there was polyvisceral stasis.

In conclusion, two infants died one shortly after the other 72 hours after they had last been in the same nursery and 24 hours after the onset of their symptoms. In both cases, a multidisciplinary approach revealed the cause of death to be septic shock due to acute meningococcal infection with hemorrhagic adrenalitis (Waterhouse- Friderichsen Syndrome).

Waterhouse-Friderichsen Syndrome, Epidemic Outbreak, Forensic Diagnosis

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* Presenting Author