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Case Report

Fatal pneumococcal Waterhouse-Friderichsen syndrome in a vaccinated adult with congenital asplenia

Abstract

Overwhelming postsplenectomy infection (OPSI) is a low-incidence entity with a high mortality rate despite aggressive therapy. Although initial symptoms may be mild and nonspecific, it can progress rapidly to Waterhouse-Friderichsen syndrome with full blown septic shock and disseminated intravascular coagulation (DIC). Overwhelming postsplenectomy infection is known to occur at any time after splenectomy, even in patients who have received pneumococcal immunization and/or chemoprophylaxis. Although the term *OPSI* gives the impression of a "postsurgical" complication, it has been seen in association with conditions predisposing to functional hyposplenism and in children with congenital asplenia. To our knowledge, there has been no previously reported case of OPSI in a pneumococcal vaccinated adult with congenital asplenia. We report a 67-year-old woman with congenital asplenia and current pneumococcal immunization who presented to our emergency department with low-grade fever and nonspecific symptoms that evolved to DIC, refractory hypotension, shock, and death in less than 24 hours. The diagnosis of Waterhouse-Friderichsen syndrome was made at autopsy. The causative organism was *Streptococcus pneumoniae*.

A 67-year-old woman with congenital asplenia presented to our emergency department complaining of chills, fever, severe low back pain, and vomiting of 24 hours of duration. She had a recent history of a treated urinary tract infection and a remote history of sepsis and pneumonia, for which she was hospitalized and treated in other institutions. The patient also had a diagnosis of lung fibrosis of uncertain etiology and was a chronic alcohol user. The patient had received pneumococcal vaccination 1 year prior.

On physical examination, she was found to be alert and in no acute distress. Her temperature was 38°C orally; blood pressure, 100/42 mm Hg; heart rate, 113/min; respiratory rate, 26/min; and oxygen saturation, 95% on room air. She had mild tenderness to palpation in the right lower abdominal quadrant and positive bowel sounds. Her initial blood workup showed an elevated white blood cell count of 15.5 $\times 10^3/mm^3$ with 94% neutrophils. A chest x-ray revealed right lung haziness. A computed tomographic scan of the abdomen and pelvis demonstrated no acute intraabdominal process. Blood and urine samples were obtained for cultures.

Within a few hours of presentation, the patient developed a diffuse purpuric rash with severe hypotension. Cefepime and vancomycin were started. Additional laboratory tests revealed disseminated intravascular coagulation (DIC) and metabolic acidosis. Cortisol levels where within normal limits, although lower than expected considering the patient's condition.

Despite administration of vasopressor agents and intravenous fluids, she developed shock and died in less than 24 hours after presentation. At autopsy a diffuse purpuric rash was noted (Fig. 1). There was massive bilateral adrenocortical hemorrhagic necrosis (Fig. 2). A recent subdural hemorrhage (Fig. 3) and scattered focal hemorrhages in the heart and gastrointestinal tract were present. The right lung was extensively fibrotic (Fig. 4). Interstitial fibrosis and bronchiectasis were seen bilaterally. There were foci of organizing pneumonia in the right upper lobe and a focus of acute bronchitis in the right middle lobe. The liver showed severe steatosis and findings consistent with chronic alcoholic liver disease. Arterionephrosclerosis was seen in both kidneys. The bone marrow was hypercellular with a few paratrabecular aggregates of monotonous appearing lymphocytes. Asplenia was the only congenital abnormality.

Premortem blood cultures reported after death grew *Streptococcus pneumoniae* sensitive to penicillin and cefotaxime. The urine culture had negative results.

Patients with anatomical or functional asplenia have significantly increased long-term risk for fulminant, potentially life-threatening infections. Although the term *overwhelming postsplenectomy infection* (OPSI) gives the impression of a "postsurgical" complication, it may occur in association with conditions predisposing to functional hyposplenism and in children with congenital asplenia. To our knowledge, no previous case of OPSI in an adult with congenital asplenia has been published.

The incidence of serious infections after splenectomy remains low, with the incidence of fulminant OPSI ranging from 0.1% to 8.5% [1]. The initial presentation may be mild and nonspecific but may rapidly progress to septic shock with DIC and Waterhouse-Friderichsen syndrome [2,3].

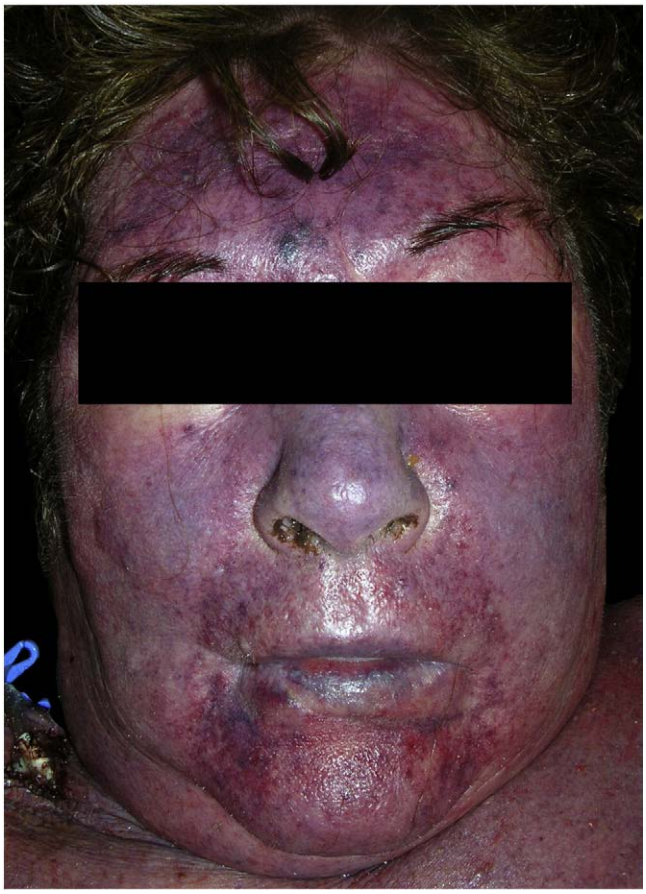

Fig. 1 Diffuse purpuric rash.

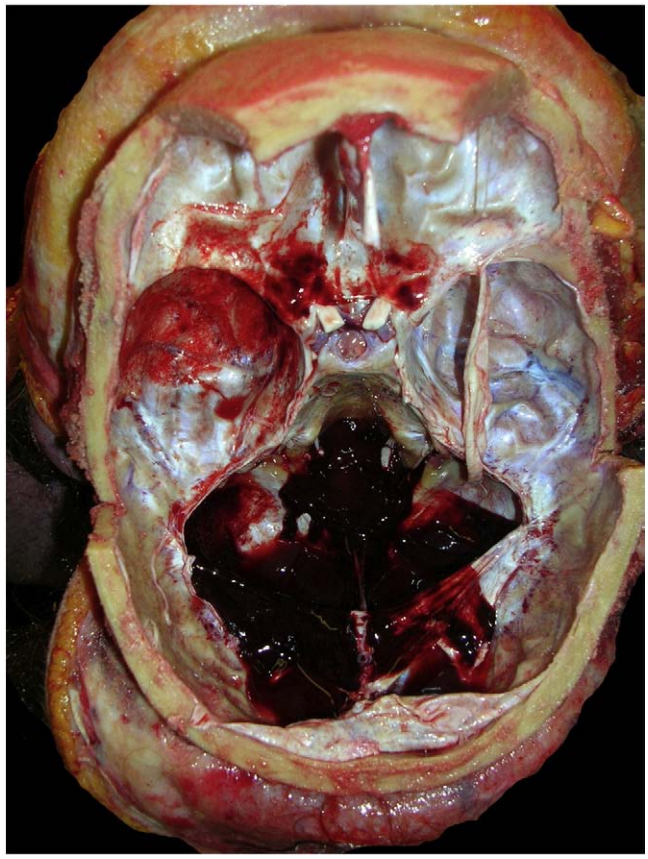

Fig. 3 Subdural hemorrhage.

The mortality rate for OPSI has been estimated as approximately 50% to 70%, despite aggressive therapy [4]. Of those patients who die, greater than 50% die within the first 48 hours of hospital admission [3].

Although our patient provided a history of congenital asplenia, the absence of alarming signs at presentation, in addition to an up-to-date pneumococcal vaccination and noncritical initial physical examination and laboratory studies, gave the false impression of a relatively stable patient who could be worked up for a precise diagnosis and subsequent treatment. Prior pneumococcal vaccination should not be reassuring as seen in our patient whose in vivo blood cultures grew *Streptococcus pneumoniae*.

The protective role of pneumococcal 23-valent vaccine for adults aged greater than 65 years remains controversial

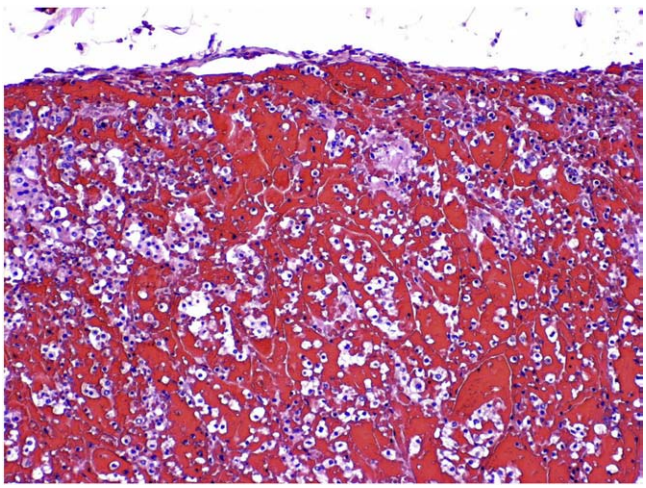

Fig. 2 Bilateral adrenocortical hemorrhagic necrosis.

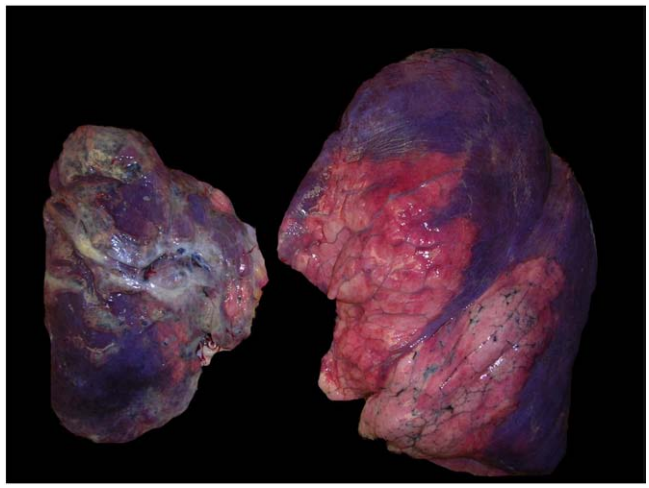

Fig. 4 Extensively fibrotic right lung.

because some older patients do not develop an immune response to the most prevalent serotypes causing invasive disease [5]. In addition, antibody concentrations and responses may be lower in elderly patients with comorbid illnesses such as alcoholic cirrhosis, chronic obstructive pulmonary disease, and lymphoproliferative disorders [6].

The 67-year-old patient we report had comorbid illnesses including chronic alcoholic liver disease and severe lung fibrosis.

Because OPSI is a virulent entity with a 50% to 80% case fatality ensuing in less than 48 hours, diagnostic workup should never delay the initiation of empiric antibiotic therapy [1]. As seen in our case, when the initial clinical presentation is mild or nonspecific, physicians may have a false sense of security and attempt to reach a precise diagnosis instead of intervening early with empiric antibiotics. Consequently, this may result in death, even when the delay is only of a few hours.

Overwhelming postsplenectomy infection is a low-incidence entity with a high mortality rate despite aggressive therapy. Although the initial symptoms may be mild and nonspecific, it can rapidly progress to septic shock, DIC, Waterhouse-Friderichsen syndrome, and death. Overwhelming postsplenectomy infection can occur at any time after splenectomy and as demonstrated by our patient should also be considered in adults with congenital asplenia. A high index of clinical suspicion is necessary for its diagnosis in any febrile asplenic patient, even among those who are immunized with pneumococcal vaccine and/or are receiving chemoprophylaxis. Diagnostic workup should never delay the initiation of empiric antibiotic therapy.

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