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A Civilian Case of Fatal Meningococcemia Due to Neisseria meningitidis, *Group Y*

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Bernad, Peter, Synder, David A., and Levey, Michael: A civilian case of fatal meningococcemia due to *Neisseria meningitidis*, group Y. Am J Clin Pathol 68: 296–298, 1977. The present report is that of a civilian episode of *fatal*, fulminant group Y meningococcemia in a previously healthy adolescent, who denied prior vaccination against group C meningococcus. The patient suffered abrupt onset of purpura, hypotension and cardiopulmonary arrest. A detailed clinical and pathologic report is included. (Key words: Group Y meningococcal disease; *Neisseria meningitidis*.)

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MENINGOCOCCAL INFECTIONS occur with alarming fulminance. The full clinical spectrum of this disease has been well described.[7,11,13,14] Epidemics have occurred in military training centers despite the use of effective prophylactic measures. To the authors' knowledge, the present report is the first case of a civilian with fatal group Y meningococcal disease.

Report of a Case

A 15-year-old black girl was admitted to LAC/USC Medical Center on March 14, 1976, 12 hours after awakening with malaise, chills, fever, myalgias, sore throat, and polyarthralgias, particularly of the right wrist. The patient also noticed a generalized petechial rash, which increased during the course of the day. She denied any past history of a similar event, weight change, visual symptoms, or cardiopulmonary symptoms. She had had anorexia and nonspecific diarrhea for 24 hours prior to admission, but denied nausea or vomiting. She denied any urinary tract symptom. She had had headache, primarily frontal, for 24 hours prior to admission. She denied neck stiffness or any history of seizures or weakness. She complained of marked joint pain and swelling. Her last normal menstrual period had been a month prior to admission. She was a non-smoker, non-alcoholic, and she denied any type of drug abuse. She had had a negative sickle preparation in the past. She denied exposure to tuberculosis, hepatitis, typhoid or any other communicable disease. Family history was entirely negative for collagen vascular diseases, bleeding diatheses, sickle-cell disease, arthritis, or immune deficiency states. No member of the family was ill, and there had been no exposure to a pet or recent travel. The patient was taking no medication and denied any allergy. She denied any prior vaccination against meningococcus.

On admission, the patient appeared alert, cooperative, slightly restless, oriented, and lucid. She was well developed and well nourished, looked her stated age, and was in mild distress. The blood pressure was 114/70 mm Hg, temperature 38.4 C, pulse 160/min, respiratory rate 36/min. Many small petechial hemorrhages were found over the skin and conjunctivae, covering the entire trunk, neck, and extremities, but not involving the palms or soles. Even during the physical examination new petechiae appeared before the eyes of the examiner. The conjunctival hemorrhages progressed to marked hemorrhagic, necrotic chemosis. There were numerous small, firm posterior cervical and inferior mandibular nodes. The pupils were miotic and reacted slowly to light and accommodation, making visualization of the fundi impossible. The tympanic membranes were clear, with no evidence of inflammation or trauma. The tonsillar pillars were markedly enlarged, with hemorrhagic, exudative necrosis. The gingivae were normal. The oral mucosa was pink and moist. The neck was supple. The chest was clear to percussion and auscultation, but the patient was markedly tachypneic. Examination of the heart re-

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vealed marked tachycardia and a regular rhythm, without gallops or murmur. Examination of the abdomen revealed hypoactive bowel sounds with diffuse tenderness. Results of pelvic examination were within normal limits. The patient had generalized polyarthritis with warm tender extremities and decreased range of motion. The extremities showed minimal cyanosis. Pulses were strongly palpable in all four extremities. Mental status was entirely within normal limits, and no abnormal reflex was elicited. Gait was normal.

Initial Laboratory Data

Hematocrit was 40.7 per cent, hemoglobin 13.7 mg/dl, mean corpuscular volume 84 cu μm, leukocyte count 4,300 with a differential of 43 polymorphonuclears, 4 band cells, 33 lymphocytes, 19 monocytes, 0 eosinophils, 0 basophils, and 1 metamyelocyte. The platelet count was 49,000/cu mm. On the peripheral blood smear many paired cocci, both intra- and extracellular, were seen. The patient unsuccessfully attempted to urinate. Arterial blood gases while breathing 28% oxygen were: P_{O_2} 178 mm Hg, P_{CO_2} 13 mm Hg, pH 7.25, base excess −20 m Eq/l, HCO_3 6 mm/l.

Hospital Course

In the process of the initial physical examination, the patient became increasingly cyanotic and dyspneic, as well as hypotensive. Within 15 minutes, pulses and blood pressure were unobtaintainable, and the patient was thought to be in shock with respiratory and cardiac arrest. The trachea was immediately intubated and cutdown was performed to obtain vascular access. Constant EKG monitoring (initially) revealed a normal sinus rhythm. Despite emergency treatment for shock the patient was unresponsive to all forms of therapy, including large intravenous doses of ampicillin and steroids. The EKG was flat after 90 minutes of vigorous resuscitative measures, and the patient was pronounced dead.

Pathologic Findings

At autopsy, there were generalized petechiae in the skin. Confluent focal hemorrhages were present on the undersurface of the scalp. The middle ears each contained minimal amounts of yellow, aqueous fluid. The meninges appeared normal grossly. On gross examination the brain appeared normal. Generalized petechial hemorrhages were present in the peritoneal surfaces. The epicardium was smooth and glistening but contained numerous petechiae. The chambers of the heart were essentially non-dilated; the atrial appendages were clear. The myocardium was firm, but on splitting there was indistinct pallor in the septum, along with glistening foci. The endocardium of the left ventricle was thin and translucent. Other cardiac structures and vessels were within normal limits. The pharynx and hypopharynx were severely hemorrhagic. The tonsils were beefy red and enlarged, with punctate, necrotic-appearing areas. Examination of the pleural surfaces revealed dark red areas with numerous petechiae. The esophageal mucosa was strutted with many petechiae. The serosal surfaces of the small intestine and appendix were similarly involved with petechiae. The pancreas was reddened but had normal lobular architecture. The liver showed areas of pale mottling; the lobular pattern was accentuated, with dark red central lobular zones. The spleen was normal. The kidneys were normal but both ureters contained numerous petechiae on the mucosal surfaces. Both adrenals were normal in size, shape, and color, with no obvious hemorrhage.

Further Laboratory Data

Neisseria meningitidis, group Y, was grown from three blood cultures, one throat culture, and one wrist fluid culture obtained prior to death. Gram stain of a centrifuged sample of postmortem cerebrospinal fluid revealed numerous polymorphonuclears with rare intracellular and extracellular gram-negative diplococci, strongly characteristic of Neisseria. Postmortem cerebrospinal fluid and spleen, but not adrenals or endocervix, grew *Neisseria meningitidis*.

Several petechiae developed on the dorsum of one foot of an intern involved with the case 18 hours later, and a course of intravenous penicillin was immediately initiated.

Discussion

The epidemiology of meningococcal disease is rapidly changing.[7,13,19] Before 1963, most epidemics were caused by group A meningococcus. From 1963–1968, however, most outbreaks of this disease were due to group B meningococcus. By 1971 most outbreaks of meningococcal disease were due to group C meningococcus. Groups B and C are now associated with sulfonamide resistance, making prophylaxis with this drug ineffective.[7]

In 1961, Slaterus initially identified group Y meningococcus,[15] but it was not until 1968 that serogroup Boshard (identical to group Y) was identified in U. S. Army isolates.[9] Three per cent of all meningococcal isolates from blood and cerebrospinal fluid submitted to the Walter Reed Army Institute of Research during 1964 to 1970 were found to be group Y.[4,9] A Fort Dix only one case of group Y meningococcal disease was

found in military personnel in the pre-vaccine years 1968–1969.[6] Even though group Y meningococcus previously was the cause of disease only rarely, it has been found to be commonly present in asymptomatic carriers.[1,3,8,10,18] Smith recently reported that group Y meningococcus now comprises 9.7–91% of all strains in group C-vaccinated carriers.[17]

Starting in 1971, all incoming Army recruits were vaccinated with the highly effective group C polysaccharide antigen.[2,5,12] Of 200 fatal cases reported by the Armed Forces Institute of Pathology in 1968, none were due to group Y meningococcus.[13] In 1974, however, Smilack pointed out the emergence of significant group Y meningococcal disease in military recruits, who previously had been given the group C vaccine.[16] In Smilack's report, group Y meningococcal disease appeared to be highly virulent, with significant mortality. In 1975, Yee and associates reported one civilian case of meningitis, pneumonitis, and arthritis caused by *Neisseria meningitidis*, group Y, successfully treated with ampicillin.[19] However, the significance of group Y meningococcal disease in civilians has not been emphasized.

The present case reports the first civilial episode of fatal, fulminant, group Y meningococcal disease, which occurred in a previously healthy adolescent who denied any prior vaccination against group C meningococcus. The patient had the abrupt onset of hypotension, shock, and cardiac arrest. Postmortem examination was significant in that the adrenal glands appeared normal but the heart and other organs showed diffuse evidence of vasculitis. Although the postmortem cerebrospinal fluid had many polymorphonuclear cells and rare gram-negative diplococci and the culture was positive for group Y meningococcus, the patient showed no sign of cerebrospinal fluid involvement clinically. In Smilack's series,[16] five of 12 patients showed no sign of cerebrospinal fluid involvement. Three of these five patients who died had positive cerebrospinal fluid and blood cultures. Furthermore, this case is the first in which group Y meningococcus is documented as a cause of polyarthritis.

The authors wish to alert the civilian medical community of the emergence of *virulent* group Y meningococcus as, potentially, the predominant serotype in meningococcal disease. We feel that emergency room personnel, in particular, should be aware of this virulent disease, since immediate diagnosis and therapeutic measures must be undertaken. The urgent need for prompt treatment is further emphasized by the fact that meningococcemia is a disease affecting primarily children and young adults.

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