INHALATION ANTHRAX IN A HOME CRAFTSMAN*

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Abstract

Inhalation anthrax with complicating subarachnoid hemorrhage due to simultaneous infection with two capsular biotypes of Bacillus anthracis of different virulence for the mouse is reported. The patient, a home craftsman, acquired his infection from imported animal-origin yarn.

In January 1976 the first reported case of inhalation anthrax in the United States in 10 years occurred in California. The patient was a self-employed artistic weaver who worked in his home and obtained his yarns from commercial sources. The source of his infection was contaminated yarn imported from Pakistan, which contained various types of animal-origin fibers. The clinical and pathologic findings in this unique case of inhalation infection by two separable capsular types of Bacillus anthracis is presented.

CLINICAL SUMMARY

A 32 year old Caucasian male was in apparently good health until five days prior to hospitalization. At that time he complained of fever and a sore throat. At the time of admission his complaints also included left sided chest pain, headache, nausea, and anorexia. Physical examination at the time of admission disclosed an acutely ill patient with sinus tachycardia and decreased breath sounds in the left lower lung field. Neurologic examination demonstrated an inability to carry out simple commands, left upper and lower extremity spasticity, disconjugate gaze, and intact tendon reflexes. A chest radiogram displayed a moderate sized left pleural effusion and findings suggestive of enlargement of the left pulmonary hilus. Serum chemistries were normal with the exception of an elevated bilirubin (2.6 mg. per dl.). A lumbar puncture performed within one hour of admission showed normal opening and closing pressures, crystal clear fluid, a protein level of 220 mg. per dl., and a glucose level of 65 mg. per dl. Gram staining of this spinal fluid demonstrated many large gram positive rods in chains. A second lumbar puncture was performed three hours later, and this demonstrated a xanthochromic fluid containing 7300 white blood cells, 83 per cent of which were polymorphonuclear. Many bamboo shaped, gram positive bacillary forms in chains were identified on Gram staining of this material. A concomitant investigation of the left pleural effusion was conducted and this too demonstrated gram positive rods.

A therapeutic regimen consisting of the intravenous administration of penicillin, 5 million units every four hours, and 500 mg. of streptomycin intramuscularly every 12 hours was instituted. Six hours after admission both the patient's pupils were dilated and failed to respond. A diagnosis of a hemorrhagic central nervous system process was made. Shortly thereafter the patient began to exhibit intermittent decerebrate rigidity and Cheyne-Stokes respiration. Despite extensive supportive measures, he expired 28 hours after hospital admission.

AUTOPSY SUMMARY

At necropsy, some 36 hours after death, the body displayed no cutaneous lesions other than those attributable to intravenous therapy and a small subungual hematoma of the left thumb. The subcutaneous tissues were firm and edematous. With the exception of congestive splenomegaly, the principal sites of pathologic alteration were in the thorax and central nervous system. The thoracic lesions included hemorrhagic mediastinitis, which diffusely infiltrated the fibroadipose tissues anteriorly and posteriorly. Lymphoid structures in this area contained intraparenchymal hemorrhage, which completely obscured their normal architecture. Microscopically these tissues exhibited zones of necrosis and acute inflammation with some small vessels containing fibrin-like, cosinophilic hyaline material. Rare gram positive bacillary forms were seen in these sites. The lungs together weighed 1300 gm. There was marked collapse of the left lung. When cut, both lungs failed to demonstrate intraparenchymal focal lesions that could be attributed to B. anthracis, and no organisms were demonstrated on Gram stain. Microscopic examination of the pulmona-

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BACTERIOLOGIC SUMMARY

Mediastinal and splenic tissues were fixed in 3 per cent glutaraldehyde, but owing to the sparsity of bacilli in the paraffin sections, these were not studied by electron microscopy. The brain was fixed in 10 per cent formalin. Lep­ tomeninges from the temperoparietal region, where bacilli were numerous, were postfixed in glutaraldehyde, followed by 1 per cent osmium tetroxide, and stained en bloc in uranyl acetate before dehydration and embedding in Epon.1

Thin sections were stained with uranyl acetate and lead citrate. Vegetative procaryotic cells were found within the subarachnoid space. Intact dividing forms in the intercellular fluid produced chains (Fig. 1). These had cell walls and protoplasts characteristic of the vegetative form of B. anthracis.2 Capsules were not visualized and there was no spore formation. Phagocyted bacilli were found within membrane bounded spaces of the leucocytes in various stages of degradation (Fig. 3). Lysis of the pro­ toplasts was followed eventually by disintegration of the cell walls.

Extensive bacteriologic investigation was conducted at the Center for Disease Control, Atlanta, Georgia. Material from the anterior mediastinum and from the cerebral cortex in the region adjacent to that from which the material used for electron microscopy was ob­ tained yielded a growth of well encapsulated and poorly encapsulated, gram positive, sporulat­ ing bacillary forms. Immunofluorescence micro­ scopy with specific anti-B. anthracis anti­ serum was positive. Subsequent biochemical analyses were consistent with B. anthracis and the isolated organisms were gamma phage posi­ tive. The same two biotypes of B. anthracis were also identified in the yarn obtained from the patient's residence.

Virulence testing in mice demonstrated that the poorly encapsulated form recovered from both the patient's mediastinal tissue and the yarn was less virulent for mice than typical encapsulated forms from yarn. In this testing, three poorly encapsulated isolates (two from yarn and one from the patient) and a well en­ capsulated yarn isolate were streaked on blood agar plates and incubated for 18 hours at 37° C. The growth was washed from the plates and suspended in a sterile physiologic saline solution. An aliquot of each suspension was inoc­ ulated intraperitoneally into five mice, at a dosage of 7.5 to 9.5 x 10⁴ B. anthracis colony forming units per mouse. All five mice inoculated with the encapsulated isolate died within 72 hours, which is normal in mouse virulence testing. In each of the three groups inoculated with the poorly encapsulated forms, only one of five mice died within 13 days after the challenge. These deaths occurred one, three, and six days after the challenge.

DISCUSSION

Inhalation anthrax, wool sorter's disease, has been an exceedingly rare entity in this country, usually confined to individuals with occupational exposure. Since 1900, 15 cases, 15 of which were fatal, have been reported in the United States.3 Undoubtedly the rarity of this disease accounts for the failure of numerous careful studies, supporting the Eppinger concept of spores transported via the lymphatics to hilar structures where germination occurs,4 to displace the erroneous Fraenkel pathogenetic concept of a primary bronchial pustular lesion followed by anthrax pneumonia, even in modern textbooks of pathology.5 Moreover, al­ though anthrax provided Koch with the model from which he derived his postulates, and also allowed Pasteur to demonstrate for the first time the possibility of immunotherapy, the pathogenetic mechanism of inhalation anthrax and the nature of the anthrax toxin are still not well understood.6

The morphologic constellation of lesions present in this patient, hemorrhagic mediastini­ tis without evidence of pneumonia, is consistent both with previous reports of human inhalation anthrax and with the morphogenesis of the les­ sions observed in animal models.5,2 The hemor­ rhagic meningitis noted in this patient has been observed in other cases of inhalation and cuta­ neous anthrax.8,10 This devastating complica­ tion appears to be common in the inhalation derived infection.11 However, only a small
number of cases of inhalation anthrax have been seen to date. The unique feature of this case is that for the first time there has been recovery of two capsular biotypes present simultaneously within the same individual. Both the well encapsulated and the poorly encapsulated biotype were also present in the yarn that was the source of this human infection. Mouse pathogenicity studies of the two capsular biotypes have demonstrated a marked difference in the pathogenicity of these variants.

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