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Pathological Case of the Month

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A 3-WEEK-OLD Hispanic boy was seen in the emergency department with a 2-day history of cough and rhinorrhea. This breast-fed infant was the 3.1-kg product of a 36-week gestation to a 20-year-old, Spanish-speaking primigravida woman. There had been exposure to a 16-year-old relative with an upper respiratory tract infection who was newly arrived from Mexico. In the emergency department the infant had a temperature of 38.2°C. Physical examination results were within normal limits and he was discharged.

On the third day, the infant was seen in the clinic with cough and rhinorrhea. Examination revealed bilateral rhonchi, occasional substernal retractions with grunting, and a 2/6 systolic ejection murmur. The infant was admitted and a chest x-ray film was interpreted as being within normal limits (**Figure 1**, A). The white blood cell count (WBC) was 24.7×10^9/L with 0.42 neutrophils and 0.45 lymphocytes. The infant appeared to be doing well and was discharged for 2 days.

The day after discharge (day 6 of illness), he was readmitted with lethargy, decreased oral intake, and diminished urine output. Tachycardia (180-200 beats/min), tachypnea, and temperature to 38.7°C were noted. Chest film showed a right upper lobe infiltrate (Figure 1, B). The WBC was 55.1×10^9/L with 0.48 neutrophils and 0.41 lymphocytes. Ampicillin, gentamicin, and erythromycin therapy was started. Paroxysms of coughing continued and oxygen by nasal cannula was administered. On day 7 of illness, the WBC was 65.6×10^9/L. Deterioration of his respiratory status necessitated endotracheal intubation. Chest film demonstrated an opaque right lung (Figure 1, C). An acute decrease in blood pressure to 27/13 mm Hg was unresponsive to boluses of albumin and epinephrine. Ventricular fibrillation developed and the child could not be resuscitated.

Autopsy findings included consolidation of the right lung and around the interlobar fissure on the left side (**Figure 2**). Combined heart-lung weight was 135 g. Clear serous pleural effusions were present (15 mL right lung, 5 mL left). The spleen weight was normal at 9.8 g. A sterile culture from the right lung showed no growth of bacterial or viral organisms.

A diffuse lobar pneumonia with alveoli filled by macrophages, proteinaceous material, and necrotic cell debris is seen in **Figure 3**. Multiple aggregates of gram-negative coccobacilli associated with the cilia of the respiratory epithelium were seen within bronchiole lumens.

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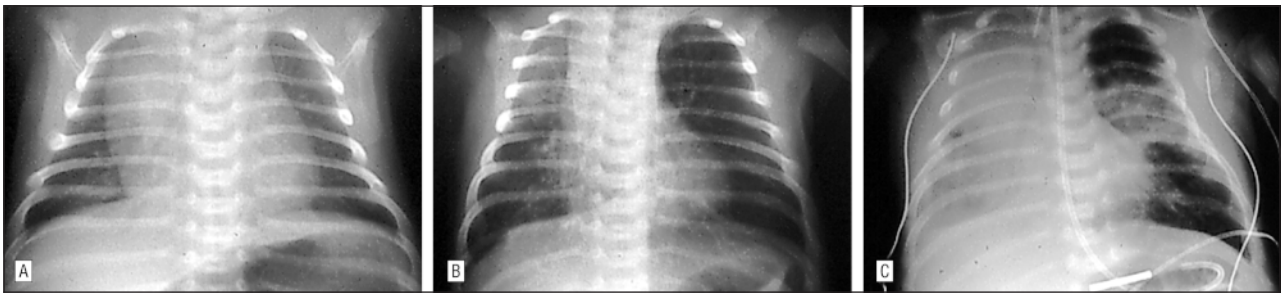

Figure 1.

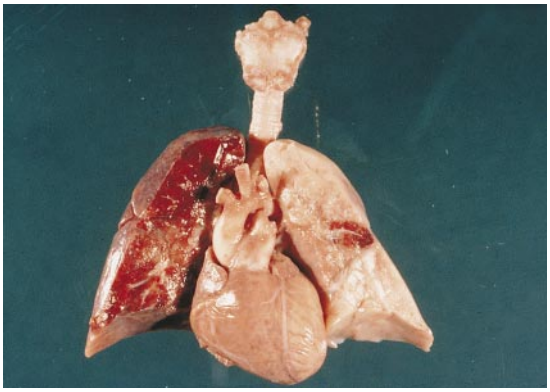

Figure 2.

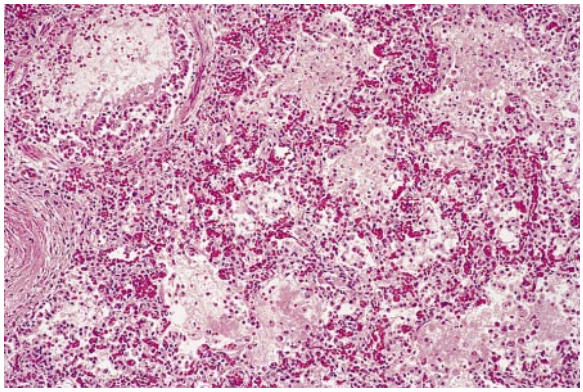

Figure 3.

Diagnosis and Discussion

Pertussis Pneumonia

Figure 1. *A, The lungs appear unremarkable on day 3 of illness. B, An infiltrate in the right upper lobe was present on day 6 of illness. C, Complete "whiteout" of the right lung field with incipient infiltration on the left along the oblique fissure was present on day 7.*

Figure 2. *At autopsy the right lung shows complete consolidation with focal involvement along the left oblique fissure, exactly mirroring the chest x-ray film.*

Figure 3. *A bronchiole (left upper corner) contains desquamated respiratory epithelium and exudate. There is no surrounding lymphocytic infiltrate. The interstitium is congested while the alveoli are filled with macrophages, proteinaceous debris, and necrotic cells (hematoxylin-eosin, original magnification ×40).*

Figure 4. *Immunohistochemical staining for pertactin demonstrates the organisms localized to the ciliated border on the surface of the bronchiole mucosal epithelium. No organisms were detected in the alveoli (alkaline phosphatase fast red with Harris hematoxylin counterstain, original magnification ×40).*

A polymerase chain reaction amplification assay[1] confirmed *Bordetella pertussis* in both premortem and postmortem specimens. Detection of *B pertussis* using standard microbiologic culture or direct fluorescent antibody detection may be unreliable. Polymerase chain reaction–based assays, while not widely available, have demonstrated increased sensitivity relative to traditional methods[2] and have the advantage of remaining positive even after therapy has started.[3] In addition, immunohistochemical staining with a monoclonal antibody for pertactin antigen[4] demonstrated that the organisms attached to the cilia were *B pertussis* (**Figure 4**). Special stains did not show other organisms; no viral inclusions were noted. These findings suggest that *B pertussis* was the sole infecting agent.

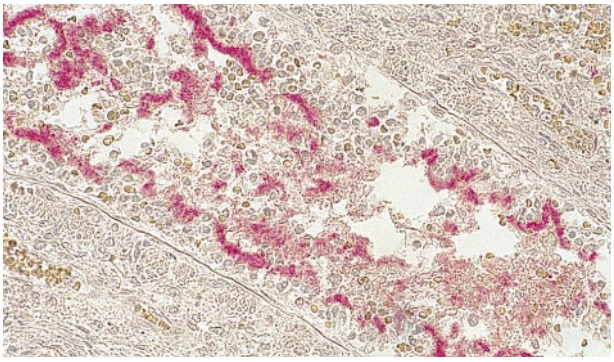

Figure 4.

A remarkable feature of this case is the rapid progression of *B pertussis* pneumonia. Premortem clues to the diagnosis include the absolute lymphocytosis noted on day 3 of illness and the paroxysms of cough interspersed with periods of apparent well-being. In retrospect, earlier administration of erythromycin might have altered the outcome. Suspicion of pertussis on clinical grounds should dictate initiation of treatment until diagnostic testing for the organism is completed.

Pertussis mortality, typically due to pneumonia, is associated with median maternal age of 20 years, gestational age of 36 weeks or less, and age younger than 1 year.[5,6] There may be an increased incidence of mortality in Hispanics, presumably as a result of difficulties with communication or genetic or socioeconomic factors. While the mechanism of death is uncertain, perfusion through the consolidated lung may lead to pulmonary hypertension with subsequent right-sided heart failure ending in cardiac arrhythmia.[7]

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