PNEUMOCYSTIS CARINII INFECTION OF THE SMALL INTESTINE

Kraig Kinchen, MD, MSc, Tina Harris Kinchen, MD, and Thomas Inglesby, Jr, MD
Baltimore, Maryland

Extrapulmonary *Pneumocystis carinii* infections are rare in comparison to other opportunistic infections in patients with acquired immunodeficiency syndrome (AIDS). In recent years, however, the number of reported cases of extrapulmonary pneumocystosis has increased. It is therefore important for physicians to recognize the various presentations of extrapulmonary *P carinii* infection. This article reports a case in which the initial clinically detected AIDS-related infection was extrapulmonary *P carinii* infection of the small intestine diagnosed after perforation of the jejunum. (*J Natl Med Assoc.* 1998;90:625-627.)

Key words: pneumocystosis ◆ small intestine ◆ acquired immunodeficiency syndrome

Pneumocystis carinii remains one of the most common pathogens in patients with acquired immunodeficiency syndrome (AIDS). *Pneumocystis carinii* pneumonia is the AIDS-defining condition in 28% of people with AIDS, and it represents the major identifiable cause of mortality.[1] Extrapulmonary *P carinii* infections, however, have been comparatively rare. This article reports a case of extrapulmonary pneumocystosis diagnosed after perforation of the small intestine in a patient with no evidence of pulmonary infection.

CASE REPORT

A 31-year-old previously healthy man presented to a medical clinic complaining of sharp, intermittent left upper-quadrant abdominal pain of approximately 10 days' duration. One day later, the clinic informed the patient that his hematocrit was low and referred him to our hospital's emergency department for further evaluation.

On admission, the patient described his pain as lasting for several seconds and occurring three to four times each hour. He also described recent weight loss, subjective fevers, and a "greasy film" on his stool for approximately the past week. He denied nausea, vomiting, melena, hematochezia, and dyspnea.

The physical examination was notable for a temperature of 101.4 degrees F, thrush, and very mild left-sided abdominal tenderness on palpation. No guarding or rebound tenderness was appreciated. There was no organomegaly. A rectal examination was unremarkable, and his stool was guaiac-negative.

Laboratory data revealed the following: hematocrit, 15.3%; blood urea nitrogen, 61; creatinine, 6; and lactate dehydrogenase, 319 units/L. His white blood cell count was 7000 with 52% polymorphonucleocytes, 33% lymphocytes, 7% monocytes, and 6% band forms. A urinalysis revealed oxalate crystals and granular casts, but no red or white blood cell casts. A chest radiograph was normal. Renal ultrasound showed increased echogenicity of both kidneys, but no hydronephrosis.

An abdominal computed tomography (CT) scan with oral contrast showed moderate, likely inflammatory, focal thickening of the distal jejunum and possible thickening in the region of the sigmoid colon. On the following day, a small bowel series also showed focal bowel wall thickening of the distal jejunum.

On the third hospital day, laboratory testing

From the Johns Hopkins Hospital, Baltimore, Maryland. Requests for reprints should be addressed to Thomas Inglesby, Jr, Johns Hopkins Hospital, Division of Infectious Diseases, Ross Bldg, Rm 1159, 720 Rutland Ave, Baltimore, MD 21205.

revealed that the patient was seropositive for human immunodeficiency virus (HIV). His CD4 count was 28 cells/mm^3. The patient continued to have intermittent low-grade fever, but blood cultures and a repeat chest radiograph were unremarkable. Stool remained negative for occult blood, and stool cultures were negative for bacterial pathogens and acid-fast bacilli.

By the fifth day of hospitalization, his abdominal discomfort had resolved. He underwent an esophagogastroduodenoscopy, which showed nonspecific inflammation of the duodenal bulb. A sigmoidoscopy revealed mild erythema as well as a small polyp in the sigmoid colon. He was discharged to home on the fifth hospital day, and close outpatient follow-up arranged. A renal biopsy was scheduled to evaluate further his renal failure, which was thought to be due to HIV nephropathy. Blood cultures for mycobacteria were pending at the time of discharge.

One day after discharge, the patient returned to the emergency department with sudden onset of severe abdominal pain. An abdominal radiograph showed pneumoperitoneum, and an exploratory laparotomy was performed emergently. The surgeons discovered a perforated mass in the mid jejunum and a near perforation in the proximal jejunum. An en bloc resection with primary anastomosis was performed. The methenamine silver stain was positive for *P carinii*, and the patient was started on a 21-day course of intravenous trimethoprim-sulfamethoxazole.

His postoperative course was complicated by profound changes in mental status, recurring fevers, worsening renal failure, multiple abscesses in the abdomen and pelvis, enterococcal sinusitis, anorexia, and pleural effusions. On hospital day 22, he required intubation after developing respiratory distress with bilateral alveolar infiltrates, attributed to nosocomial pneumonia. His respiratory status improved on broad-spectrum antibiotics. He was extubated on day 29. He was discharged to a rehabilitation facility on day 62 of his hospitalization. The patient was seen 2 months after discharge and was doing well at home.

DISCUSSION

Extrapulmonary pneumocystosis has been an uncommonly reported occurrence in patients with AIDS. In one New York hospital, only one case of extrapulmonary pneumocystosis was diagnosed for every 200 cases of *P carinii* pneumonia.[2] However, the true incidence of extrapulmonary infection due to *P carinii* is probably slightly higher. Some extrapulmonary infections have only minor clinical manifestations and may be discovered only at autopsy.[2] Using autopsy data, investigators at Memorial Hospital in New York reported a 2.5% incidence of extrapulmonary pneumocystosis in AIDS patients.[3] Raviglione[2] reviewed the 34 reported cases of extrapulmonary pneumocystosis in AIDS patients up to 1990 and found that the extrapulmonary infection was determined to have been clinically significant in only 65% of the cases. In some cases, undetected extrapulmonary infection may be treated successfully while a patient is being treated for concurrent *P carinii* pneumonia.[2]

The mechanisms by which *P carinii* infects various extrapulmonary sites have not been determined definitively. Possibilities include primary infection of the extrapulmonary site, reactivation of a latent infection, and hematogenous or lymphatic spread from a primary pulmonary infection.[3] Supporting hematogenous spread is the observation that during episodes of *P carinii* pneumonia, almost half of patients have been found by polymerase chain reaction to have *P carinii* nucleic acids in the blood.[4] In 30 of 37 cases of extrapulmonary pneumocystosis reviewed by Cohen and Stoeckle,[3] patients either had concurrent *P carinii* pneumonia or a history of *P carinii* pneumonia in the months to years prior to diagnosis.

Extrapulmonary *P carinii* infections may involve diverse anatomic sites. The most common sites reported in the literature include the lymph nodes, spleen, liver, and bone marrow. Other reported sites include the eyes, thyroid gland, ears, pancreas, kidneys, and adrenal glands.[2,3,5-10] Examples of the wide range of clinical presentations include hepatitis, abdominal discomfort, hearing deficits, peritonitis, and thyroid masses.[7,11] Some individuals may present with only fever and night sweats.[4,12]

A small number of cases involving the gastrointestinal tract have been reported. One such case was a patient with AIDS who presented with fever, chills, and diarrhea, and was found on colonoscopy to have *P carinii* infection involving his colon.[13] Rachal et al[14] described a patient with *P carinii* pneumonia who developed upper abdominal pain due to gastric *P carinii* infection.[14]

Like the case described here, several other cases described in the literature involved the small intestine specifically. In one case, a man with *P carinii*

pneumonia presented with dysphagia and weight loss and was found to have *P carinii* infection involving the duodenum and esophagus.[5] Bierhoff et al[15] described a patient with AIDS and known *P carinii* pneumonia who presented with an acute abdomen. The patient had a perforation of the small intestine secondary to *P carinii* infection, which also involved the spleen, lymph nodes, and thyroid gland.[15] Carter et al[16] described a patient who presented with left lower-quadrant abdominal pain and was found to have *P carinii* infection just proximal to the ileocecal valve.

Over the past decade, the number of reported cases of extrapulmonary pneumocystosis has increased.[10] Several factors may be responsible for this increase. First, the increased longevity of AIDS patients may have increased the number of cases seen.[17] Second, it has been speculated, but by no means proven, that strains of *P carinii* with different virulence factors may be responsible.[17] Third, some authors have argued that the use of aerosolized pentamidine for *P carinii* pneumonia prophylaxis increases the likelihood of developing extrapulmonary pneumocystosis.[2] This belief is based on the observation that there is little systemic absorption of aerosolized pentamidine,[18] and it is therefore unlikely to prevent extrapulmonary infection. In comparison, there are few cases in which extrapulmonary pneumocystosis was diagnosed in patients on systemic prophylaxis with trimethoprim-sulfamethoxazole.[4]

The relationship between extrapulmonary pneumocystosis and aerosolized pentamidine is somewhat complex. In nearly 60% of the cases reported prior to 1990, the patient was being treated with aerosolized pentamidine. In a review of 37 cases of extrapulmonary pneumocystosis, patients using aerosolized pentamidine tended to have disseminated pneumocystosis appearing long after a patient's initial AIDS-defining illness. In contrast, those not using aerosolized pentamidine tended to have a localized extrapulmonary pneumocystis infection, often as their initial AIDS-defining infection.[8]

The case described here represents one of a small number of reported cases of *P carinii* infection of the small intestine. It is unusual in that the patient had no known history of *P carinii* pneumonia and had not used aerosolized pentamidine.

CONCLUSION

In the past, clinically significant extrapulmonary *P carinii* infections in patients with AIDS have been relatively rare, but the number of reported cases has increased. In this light, it is important that physicians recognize the various presentations of extrapulmonary pneumocystosis.

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