Bacillary angiomatosis is a recently described vascular proliferative lesion that occurs most commonly in individuals infected with human immunodeficiency virus. Cutaneous lesions are the most frequently described manifestations of bacillary angiomatosis. However, as culture techniques and disease recognition have improved, additional manifestations have been identified in human immunodeficiency virus–infected individuals, including bacillary peliosis hepatis and isolated bacteremia. Two species of the genus Bartonella (formerly Rochalimaea), Bartonella henselae or Bartonella quintana, have been cultured from the cutaneous lesions of bacillary angiomatosis. A new manifestation of Bartonella infection is reported: an intra-abdominal mass presenting with massive gastrointestinal hemorrhage in a patient with human immunodeficiency virus infection. B. quintana was cultured from a percutaneous needle-biopsy specimen obtained from the highly vascularized intra-abdominal mass. The bacillary angiomatosis lesion resolved after 3 months of tetracycline treatment. Recognition of Bartonella infection is extremely important because it is readily treatable with antibiotic therapy.

Case Report

A 24-year-old white woman with acquired immunodeficiency syndrome (based on the presence of HIV antibodies and a CD4+ cell count of 10/mm³) presented to the emergency room with upper gastrointestinal hemorrhage. She reported a 1-month history of mild epigastric and right upper quadrant abdominal discomfort. The patient noted sudden onset of hematemesis and melena. The patient had two syncopal episodes before arriving at the emergency room. She had been taking no medications and had no history of prior gastrointestinal hemorrhage. The HIV risk factors included intravenous drug use and sexual contact with high-risk (intravenous drug use) men. The patient denied alcohol abuse, intravenous drug use, or other substance abuse in the previous 2 years. She resided with a group of people in England until 6 months before this presentation, when she had moved back to the Russian River area of Northern California to live with her mother. While in England, she was exposed to several cats belonging to her roommates but denied receiving any cat scratches.

There was no history of any opportunistic infection or malignancy. The past medical history was notable for presence of hepatitis C antibodies, with intermittent elevation of hepatic transaminase enzymes. Pertinent findings on admission included orthostatic changes (pulse increase from 125 per minute supine to 175 per minute standing) and a temperature of 38.2°C. There were no cutaneous or oral lesions, and the abdominal examination was benign without organomegaly, guarding, or palpable masses. The admission complete blood count showed a hemoglobin level of 7.4 g/dL, hematocrit level of 22%, and a normal leukocyte count, platelet count, mean corpuscular volume, prothrombin time, and partial thromboplastin time. Blood chemistry tests were remarkable only because of an albumin level of 3 g/dL.

Upper gastrointestinal endoscopy showed no source of bleeding, and no esophageal varices were present. A barium study of the upper gastrointestinal tract, including small bowel follow-through, showed abnormal proximal jejunal mucosa with evidence of an extraluminal mass displacing jejunal loops. Sonographic evaluation showed a left upper quadrant 8.5 × 4 × 6-cm intraperitoneal mass. Endoscopy was performed again, without visualization of specific pathology except for evidence of recent bleeding. Histopathologic examination of a
biopsy specimen of the mass, obtained by pushing through the jejunal wall, showed swollen villi with proliferating vascular channels and inflammatory cells. There was no evidence of Kaposi’s sarcoma, cytomegalovirus, mycobacteria, or lymphoma. Computed tomography of the abdomen and pelvis showed a lobulated heterogeneous 8 × 4 × 5-cm vascular appearing, contrast-enhancing mass in the left hemiabdomen centered within either the mesenteric root or the proximal jejunal bowel loop (Figure 1A). The patient underwent ultrasound-guided needle biopsy of the intraperitoneal mass. The histopathology performed at another hospital was suggestive of BA, and repeat ultrasound-guided biopsy specimens were obtained for culture and further pathological diagnosis.

The patient was administered 500 mg erythromycin four times a day but, soon thereafter, developed severe nausea, vomiting, diarrhea, and an elevated temperature of 39.8°C after initiation of therapy. The antibiotic was changed after 7 days to 500 mg clarithromycin twice a day with persistence of gastrointestinal symptoms and several days later was changed to 500 mg tetracycline four times a day, which was well tolerated. The patient’s fever resolved 2 weeks after initiation of antibiotic therapy. Her appetite and other systemic symptoms improved, and a follow-up CT scan of the abdomen and pelvis after 3 months of antibiotics showed resolution of the intraabdominal mass (Figure 1B). The patient continued tetracycline therapy for 3 months and then subsequently declined further tetracycline treatment. She died 6 months after the abdominal BA was diagnosed without further gastrointestinal hemorrhage or any signs of recurrent BA.

Methods and Results

Microscopic examination of an H&E-stained specimen obtained by percutaneous needle biopsy showed regions of fibrosis in addition to regions showing tissue changes characteristic of BA: proliferation of small vessels with interspersed neutrophilic debris. A Warthin–Starry stain showed clusters of degenerating bacteria in the foci of leukocytoclasis. The biopsy specimen was cultured for Bartonella species by homogenizing the tissue in inoculation media and spreading the homogenate onto chocolate agar. The plates were incubated for 14 days in 5% CO₂ at 36°C, and bacterial growth was noted 11 days after inoculation. The bacterial isolate was identified as B. quintana by sequencing a fragment of the 16S rRNA gene as previously described and confirmed by polymerase chain reaction–restriction fragment length polymorphism analysis of a citrate synthase gene fragment.

Discussion

Four of the five species of Bartonella, B. henselae, B. quintana, B. elizabethae, and B. bacilliformis, are known to cause infection in humans. In both immunocompromised and immunocompetent individuals, infection with these species may be manifest as relapsing bacteremia (B. quintana and B. henselae) or endocarditis (B. elizabethae and B. quintana). B. bacilliformis infection is biphasic, with an initial bacteremic phase accompanied by anemia (Oroya fever) often followed by a long-term phase characterized by verrucous cutaneous lesions. However, infection with B. bacilliformis occurs only at specific altitudes in the Andes of western South America. Focal infection with B. henselae or B. quintana results in different histopathologic manifestations, depending on
the immune status of the individual. In the presence of an intact immune system, infection with *B. henselae*, the agent of cat-scratch disease, results in necrotizing lymphadenitis. In contrast, focal *B. henselae* or *B. quintana* infection in patients with late stage HIV infection is characterized by striking vascular proliferative changes in the regions infiltrated with the *Bartonella* bacilli, hence the name BA.

Angiomatous lesions caused by *Bartonella* infection are identified most frequently in skin but occasionally have been reported in brain parenchyma, liver, spleen, bone, lymph nodes, and lung (reviewed by Koehler and Tappero). Several different gastrointestinal manifestations of BA have been described. Cockerell et al. describes 2 patients with oral, anal, or peritoneal BA lesions, and two other groups also have identified patients with oral BA. Endoscopically visualized BA lesions were described by Tuur et al.; these lesions were raised, nodular, ulcerated mucosal abnormalities of the stomach and large and small intestines. Extensive retroperitoneal and mesenteric lymphadenopathy that showed marked enhancement after administration of intravenous contrast has also been reported. In another patient, enlargement of intra-abdominal lymph nodes presumed to be due to BA caused extrinsic compression of the common bile duct. Another gastrointestinal manifestation of *Bartonella* infection, bacillary peliosis hepatis, was first described by Perkocha et al. in 1990. Although these lesions show neovascularization similar to BA lesions, the blood-filled cystic spaces of bacillary peliosis hepatis are often large and not lined by the protruberant endothelial cells that are characteristic of BA lesions. Similar peliotic and sometimes necrotic lesions are observed in the spleen of some patients. Many additional cases of bacillary peliosis hepatis have been reported in the past 4 years (reviewed by Koehler and Tappero). Concomitant laboratory abnormalities may include thrombocytopenia, pancytopenia, and elevated alkaline phosphatase; abdominal computed tomography usually shows multiple, hypodense lesions scattered throughout the liver parenchyma.

Deep soft tissue or osseous BA lesions are especially vascular appearing by many imaging techniques, and this characteristic often results in misdiagnosis as a malignant vascular neoplasm. In this patient, a large heterogeneous, highly vascular left hemiabdominal mass appeared to arise from either the mesenteric root or a proximal jejunal bowel loop, as visualized by computed tomography (Figure 1A). The patient became symptomatic when gastrointestinal hemorrhage began abruptly, presumably when the infectious mass eroded through the intestinal wall. Erosion of bone also has been shown adjacent to a soft tissue mass caused by BA. In this case, as previously, the highly vascular, erosive nature of the BA mass initially led the radiologists and clinicians to suspect a diagnosis of lymphoma.

Clinicians should include *Bartonella* infection in the differential diagnosis of an intra-abdominal mass in the immunocompromised patient in addition to the previously described abdominal manifestations of *Bartonella* infection, which include the following: intraluminal intestinal BA, contrast-enhancing abdominal lymphadenopathy, bacillary peliosis hepatis, and necrotizing splenitis. The relative risk of percutaneous biopsy of intra-abdominal, highly vascularized masses or liver with peliotic changes is unknown, and hemorrhage resulting from biopsy of intra-abdominal BA or peliosis hepatis is theoretically possible. However, the abdominal computed tomography findings of the intra-abdominal mass in this patient as well as of bacillary peliosis hepatis are not sufficiently specific for *Bartonella* infection. Thus, tissue should be obtained for evaluation by culture and H&E, Warthin–Starry, fungal, and acid fast bacilli staining. Tissue biopsy specimens usually yield information that enables rapid institution of therapy for the underlying process, whether infectious (e.g., *Bartonella*, *Mycobacteria*) or malignant (e.g., Kaposi’s sarcoma, lymphoma). Open biopsy may be appropriate for some patients, and those patients who undergo biopsies percutaneously should be followed closely for hemorrhagic complications.

Antibiotic treatment has not been studied systematically, but clinical experience has shown that erythromycin, doxycycline, or tetracycline therapy is usually effective. Patients should be advised that institution of antibiotic therapy may temporarily exacerbate symptoms because of a Jarisch–Herxheimer–like reaction, as occurred in this and previous patients. Duration of treatment is also unknown, but patients with gastrointestinal involvement are presumed to have systemic disease and should probably be treated for a minimum of 4 months, with close follow-up after cessation of antibiotics.

At present, recommendations regarding prevention of infection with *B. quintana* are limited to avoidance of contact with lice. In contrast, *B. henselae* infection is a zoonosis acquired from pet cats; many cats (41% in San Francisco, CA) remain persistently bacteremic with *B. henselae* and presumably transmit the organism by bites or scratches. In contrast, the only known vector of *B. quintana* is the body louse, which transmitted the organism from a human reservoir to other humans during the World War I trench fever epidemics. Although "urban trench fever” has been reported recently, the contemporary reservoir and vector of infection with *B. quintana* remain unknown.
References


