Massive retroperitoneal aortoiliac aneurysm rupture revealing chronic Q fever

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Chronic *Coxiella burnetii* vascular infection is rare and usually develops on a pre-existing vascular lesion, such as an aneurysm or vascular prosthesis. We report a case of proven *C. burnetii* aortic infection revealed by a massive retroperitoneal aortoiliac aneurysm rupture in a patient at apparent low risk for chronic Q fever. Emergency treatment consisted of resection of the infected aneurysm and replacement with an in situ graft angioplasty. Doxycycline and hydroxychloroquine therapy was started postoperatively. After 6 months of follow-up, the patient had no signs of infection, and *C. burnetii* serologic antibody titers had significantly decreased. (J Vasc Surg Cases 2016;2:1-3.)

Q fever is a ubiquitous zoonosis caused by *Coxiella burnetii*, a strictly intracellular and highly infectious bacillus. Prevalence of the infection is currently rising, partly because investigative methods are improving and diagnostic criteria are better defined.[1] Q fever is characterized by its clinical polymorphism and may be subclinical, acute, or chronic. Endocarditis, the main manifestation of chronic Q fever, may also appear as unexplained isolated fever. Q fever may involve arteries, joints, liver, and lung. *C. burnetii* vascular infection develops almost exclusively on a pre-existing vascular lesion, such as an aneurysm or vascular prosthesis. In this article, we report a case of proven *C. burnetii* aortic infection revealed by a massive retroperitoneal aortoiliac aneurysm rupture in a patient without known underlying cardiovascular abnormality or immune deficiency.

Consent was obtained from the patient to publish these images and clinical history.

CASE REPORT

A 62-year-old African man was admitted to our hospital because of proximal weakness of the left leg and abdominal pain. He was a heavy tobacco smoker and suffered untreated high blood pressure. The patient lived in an urban area of Ivory Coast and reported only occasional contacts with farm animals. He complained of fatigue and weight loss during a few weeks. On admission, the patient was afebrile and appeared well. Physical examination elicited a diastolic heart murmur, a beating abdominal mass, and a prominent left quadriceps and iliopsoas motor deficit. Hemoglobin level was 8.7 g/dL, with a normal leukocyte count and no left shift on hemogram. Serum C-reactive protein level was 83 mg/L. Blood creatinine and liver test results were normal. Human immunodeficiency virus serology was negative. Computed tomography (CT) body scan revealed a left retroperitoneal rupture of a large aortoiliac aneurysm (Fig 1, *A*).

Open abdominal aneurysm repair through median laparotomy was performed emergently. Because of the thickness of the retroperitoneum and apparent periaortic inflammation, a vascular infection was suspected. The infrarenal aorta was clamped, and an aortobi-iliac bypass procedure using a silver-coated Dacron bifurcated graft was started. Right distal anastomosis was performed on the common iliac artery. Left distal anastomosis was performed on the hypogastric and the external iliac arteries. Extensive débridement of the aortic aneurysm and retroperitoneum was performed, with all apparent septic tissues resected before graft implantation. An omentum flap was wrapped around the graft. The huge retroperitoneal hematoma was decompressed and drained, and a complete neurolysis of the left crural nerve was performed. Extensive irrigation of the operative field with povidone-iodine (Betadine) ended the procedure.

The duration of the operation was 4 hours. Three units of packed red blood cells were transfused during surgery. Samples of the aneurysmatic aorta were sent for pathologic and microbiologic examination.

Because of the systemic symptoms, the increased C-reactive protein level, and the inflammatory lesions revealed by surgery, screening for aortic infection was performed and disclosed a strongly positive serology for *C. burnetii*. Anti-phase I antibody immunoglobulin G and A titers were 3200 and 800, respectively; anti-phase II antibody immunoglobulin G and A titers were both 200.

The diagnosis of chronic Q fever was made, and a combination of doxycycline and hydroxychloroquine was initiated 10 days after surgery.

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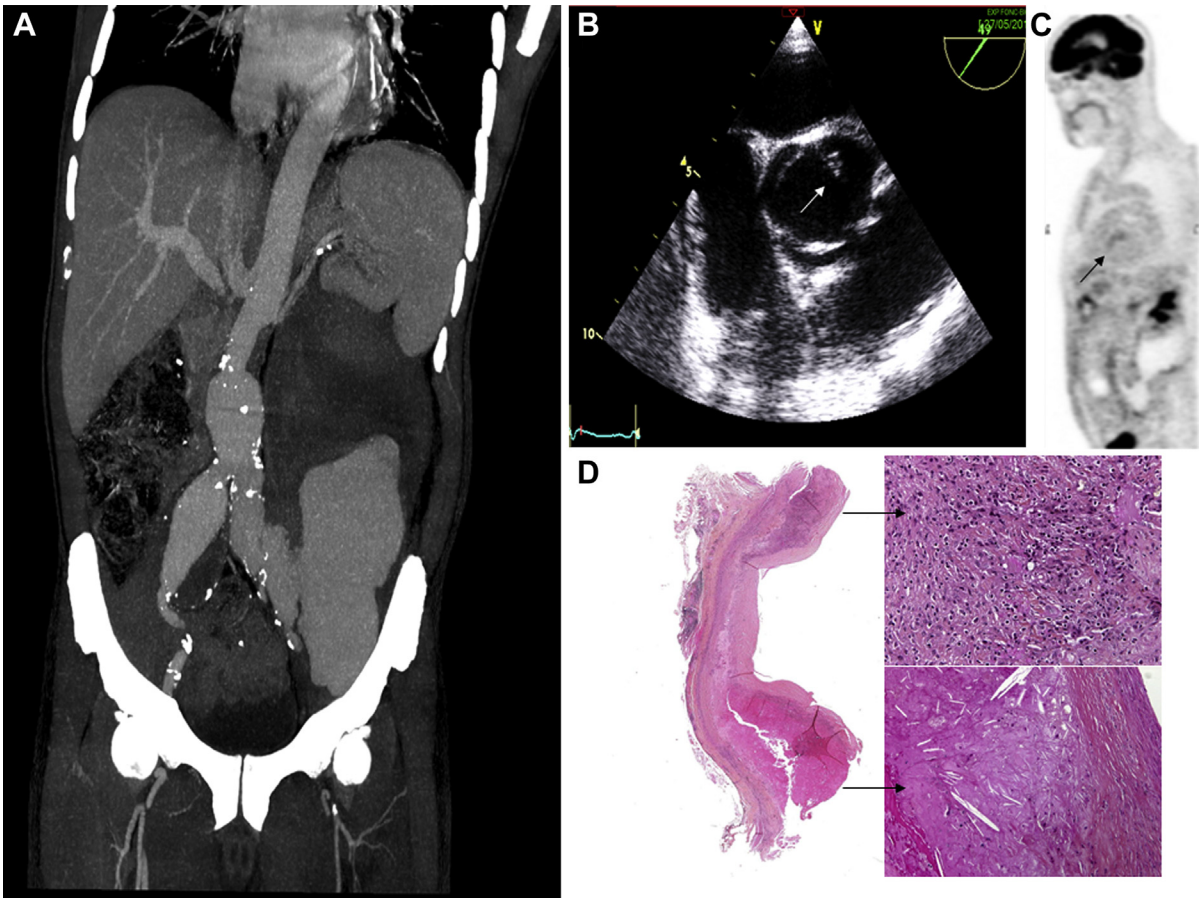

Fig 1. A, Multiplanar computed tomography (CT) coronal reconstruction showed a left retroperitoneal rupture of a large aneurysm involving the infrarenal aorta and both common iliac arteries. Spread calcifications of arterial wall suggestive of atherosclerosis were seen. **B,** Transesophageal echocardiography showed a vegetative mass on the sigmoid cusp of the aortic valve (*arrow*). **C,** ^{18}F-Fluorodeoxyglucose positron emission tomography combined with CT showed an abnormal focal uptake (maximum standardized uptake value = 2.9) in the aortic valve plane (*arrow*). **D,** Microscopic analysis of abdominal and iliac aneurysm (*left,* original magnification ×5) showed an intima-media inflammatory mononuclear infiltrate (*top right,* ×200) well separated from a lipid-rich necrotic core and fibrosis (*bottom right,* ×100).

Transesophageal echocardiography showed aortic valve vegetation with mild regurgitation (Fig 1, *B*). ^{18}F-Fluorodeoxyglucose positron emission tomography combined with CT showed an increased uptake of the anterobasal region of the heart ventricular septum (Fig 1, *C*). Findings on brain magnetic resonance imaging were normal.

Pathologic analysis of abdominal and iliac aneurysm specimens showed inflammatory cellular infiltrate with giant cells in the media associated with a lipid-rich necrotic core and fibrosis (Fig 1, *D*). No microorganisms could be identified by Gram staining. Molecular detection by polymerase chain reaction analysis of *C. burnetii* DNA was positive in the arterial wall.

The patient was discharged 25 days after hospital admission. After 6 weeks of treatment, *C. burnetii* serologic antibody titers had significantly decreased. Six months after aortic surgery, the patient was well except for persistent left lower limb weakness. Control CT scan showed no evidence of infection of the prosthetic graft (Fig 2).

DISCUSSION

Published case reports of *C. burnetii*-infected aortic aneurysms are scarce.[2-4] Lower limb motor deficiency resulting from lumbar plexus neuropathy caused by a retroperitoneal aortoiliac aneurysm rupture has never been reported in such a setting.

In our case, an infection was suspected at an early stage because of the presence of systemic symptoms (fatigue, weight loss), increased inflammatory parameters, and surgical findings (periaortic inflammation, retroperitoneum thickening). However, our patient had no obvious environmental exposure to *C. burnetii* and, without known underlying cardiovascular abnormality or immune deficiency, was at apparent low risk for chronic Q fever. Thus, the diagnosis was made by extensive screening for infection, including *C. burnetii* serologic testing. *C. burnetii* infection of the aorta was eventually confirmed by DNA

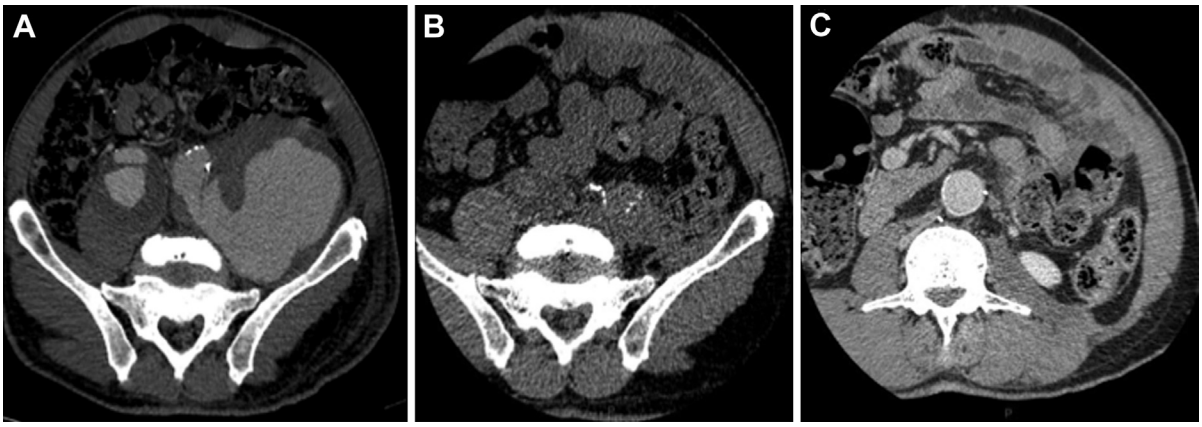

Fig 2. Axial computed tomography (CT) scan showing the large retroperitoneal hematoma at admission **(A)** and its complete resorption 6 months after surgery **(B)**. No evidence for infection of the prosthetic graft was seen **(C)**.

amplification of the bacteria from surgical samples. Given the nonspecific clinical features associated with Q fever, the significant mortality of *C. burnetii* vascular infection, and the importance of targeted and prolonged antibiotic therapy, the diagnosis of *C. burnetii* infection is crucial to a successful outcome. This observation highlights the fact that despite apparent low epidemiologic risk for Q fever, *C. burnetii* infection should be systematically considered in patients in whom infected aortic aneurysm is suspected.

C. burnetii is highly contagious and mostly spreads through inhaled aerosolized pseudospores, which are stable in various environmental conditions. The surgery was performed emergently around 10 days before the diagnosis of Q fever was made. No special precaution was thus taken to prevent potential nosocomial transmission. However, person-to-person transmission of Q fever appeared to be exceedingly rare.

Vascular rupture is a major complication of *C. burnetii* aortic infection and has clearly been associated with a fatal outcome.[3] Surgery performed after or shortly before diagnosis has been shown to improve the outcome significantly.[3] The association of vascular surgery and medical treatment is the "gold standard" therapy. The current recommendation for the treatment of Q fever vascular infection is a combination of doxycycline and hydroxychloroquine for 18 to 36 months.[5,6] Regular controls with CT scans and control of infection parameters are necessary because recurrent infection might develop.[7] In our case, the medical strategy was discussed with the National Reference Center for Rickettsia, Coxiella and Bartonella (Marseille, France). Doxycycline and hydroxychloroquine were given for 24 months. Aortic CT scan performed 6 months after the surgery showed no evidence of reinfection; *C. burnetii* serologic antibody titers had significantly decreased.

CONCLUSIONS

Chronic Q fever should be considered in patients thought to have infected aortic aneurysm rupture. A systematic screening for *C. burnetii* infection is warranted in such a situation.

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