CASE REPORTS

COXIELLA BURNETII ENDOCARDITIS IN A PATIENT WITH MYCOTIC CEREBRAL ANEURYSMS

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Abstract

Q fever is an ubiquitous zoonosis caused by Coxiella burnetii, an intracellular bacterium that can produce acute or chronic infections in humans. These forms are characterized by different evolution, serological profile and treatment that must be very long to achieve a cure in chronic forms. However, the serological profile for diagnosis and the real value of serology for predicting outcome are controversial, and management dilemmas for many patients with Q fever infection are continuously emerging. In this case report, we present a 20-year-old man from Nicaragua who worked as a farmer with a culture-negative infective endocarditis who presented with a mycotic aneurysm. The present report reviews the clinical presentation and diagnosis of Q fever IE.

Keywords: *Coxiella burnetii, Endocarditis, Q Fever, Mycotic aneurism*

INTRODUCTION

Endocarditis is the most common presentation of chronic Q fever, a zoonosis caused by the obligate intracellular bacteria *C. burnetii*. Commonly believed to be a rare disorder, it has been estimated to account for up to 5% of all IE cases worldwide[1,2]. It occurs almost exclusively in patients who have pre-existing valvular disease or who are immunocompromised.The clinical presentation of chronic Q fever is often nonspecific. Without prompt recognition and appropriate antimicrobial therapy, the course of Q fever IE is severe and potentially fatal. The following case demonstrates many of the clinical features of chronic Q fever and highlights the difficulty of obtaining the diagnosis[3].

CLINICAL CASE

A 20-year-old central american man had moved from Nicaragua to Spain for professional reasons. After two months of working on a farm he underwent emergency surgery because of a spontaneous intraparenchymal hemorrhage in the territory of the right middle cerebral artery (Fig.1). A craniotomy was performed for surgical drainage. At that time, the diagnosis of ruptured mycotic aneurysm was established. Three months later the patient went to the emergency room again due to general illness, fever and dyspnea. Given the patient´s history a study with echocardiography was completed to rule out possible endocarditis. Transthoracic echocardiogram showed an extensive calcified vegetation of approximately 5 cm anchored to the right cusp of the aortic valve, which had a bicuspid appearance and severe aortic regurgitation with normal systolic function (Fig. 2). Blood cultures and PCR in blood samples were consistently negative. Serology revealed that the antiphase I Ig G had a low titer, and the phase II titer was 1:184, consistent with a diagnosis of acute Q fever.

After two weeks of antibiotic treatment with combined doxycycline and hydroxychloroquine, surgical treatment of the endocarditis through full sternotomy and extracorporeal circulation was performed. After aortotomy and cardioplegia,

a calcified vegetation adherent to the right aortic valve leaflet was found. The aortic valve was bicuspid and no abscess cavities were identified (Fig.3) A 23 mm biological aortic prosthesis (Inspiris Resilia, Edwards Lifesciences) was implanted. A biological prosthesis was considered the best option due to the presence of another small brain aneurysm and the neurosurgeons´ recommendation to avoid anticoagulation because of the high risk of rupture. Microscopy and cultures of operative specimens revealed no microorganisms. Molecular biology study revealed negativity for the DNA of the bacterium.

The patient had a favorable postoperative course and he was discharged from hospital after six days from surgery with combined doxycycline and hydroxychloroquine.

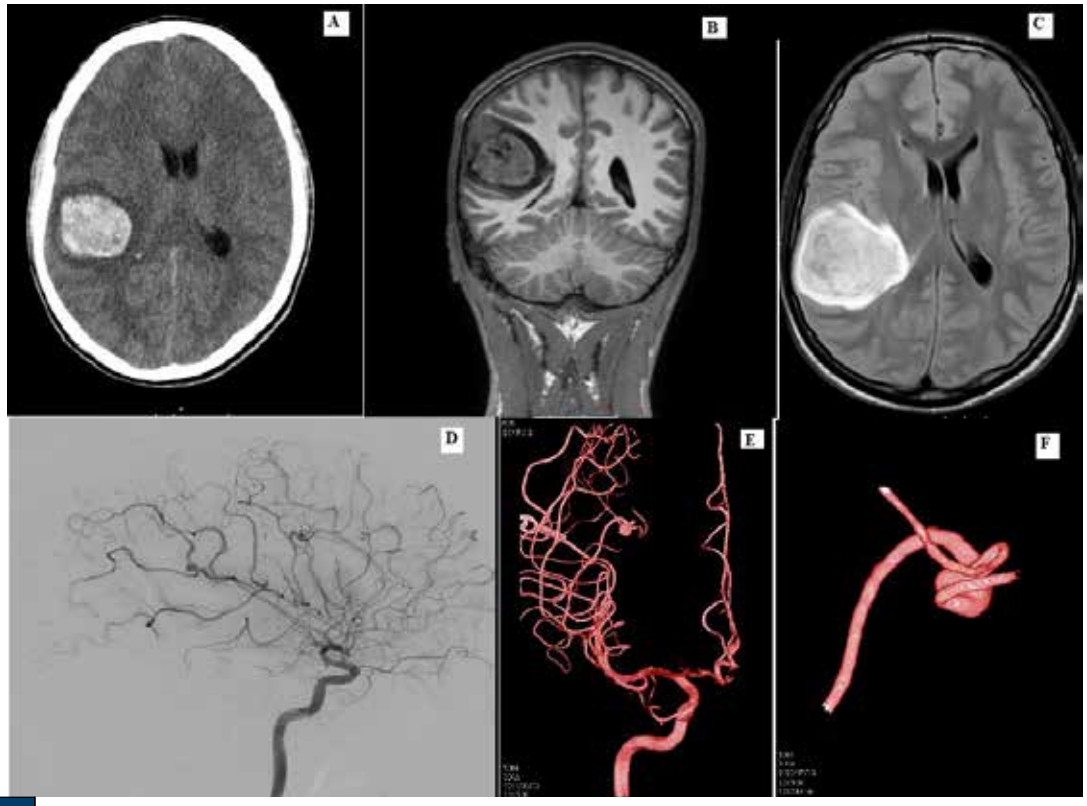

Figure 1 *Cranial CT (A) and contrasted MRI (B,C) showing a big intraparenchymal hematoma in the territory of the right middle cerebral artery. Cerebral angiography showing persistence of at least two small aneurysms in the right MCA (D,E,F).*

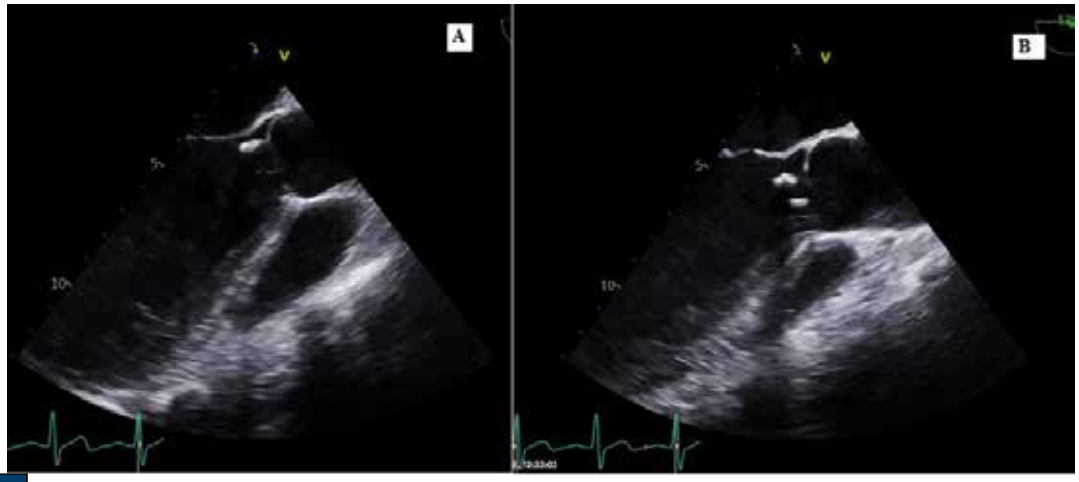

Figure 2 *Bidimensional echocardiography where a compatible image is observed with vegetation anchored to the aortic valve and entering the left ventricular outflow tract (A,B).*

He completed 18 months of antibiotic treatment. Follow-up at 3, 6, 12 and 18 months found the patient with no evidence of systemic infection, serology was negative for antiphase I and II Ig G, and normal functioning aortic valve prosthesis. He also had maintained vigilance with infectious diseases and neurosurgery specialists.

The limitation of this case is that we couldn´t send the valve tissue to additional specialized testing at a dedicated laboratory to isolate *C. burnetii* from the valve using special culture techniques due to administrative reasons.

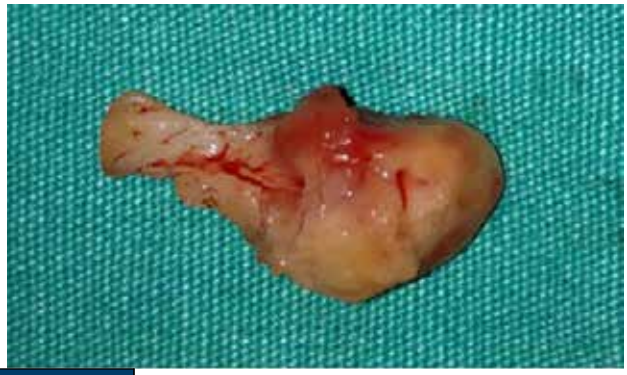

Figure 3 *Vegetation anchored to the right cusp of the aortic valve.*

DISCUSSION

While acute Q fever is common, only a very small proportion of individuals develop chronic infection, which usually manifests as endocarditis[1].

The clinical presentation of chronic Q fever is insidious and lacks many of the typical features of subacute, bacterial endocarditis. As a result, there is often a significant delay in diagnosis[2]. The majority of cases present with congestive heart failure due to valvular dysfunction. Unlike typical cases of endocarditis, fever is absent in a significant proportion and is frequently intermittent or low grade. Although embolic phenomena have been reported in up to one-third of cases, these are usually limited to advanced disease. Immune complex glomerulonephritis is frequent and usually manifests as microscopic hematuria. A purpuric rash can also be found in a minority of patients[3]. There are many peripheral manifestations of Q fever IE such as hepatomegaly and splenomegaly, systemic inflammatory syndrome and hematological abnormalities[4].

Echocardiography, usually the mainstay of diagnostic imaging in IE is of limited value in chronic Q fever. In fact, transthoracic echocardiography reveals abnormalities in only 12% of cases. This is due, in large part, to the small size and nodular shape of the typical vegetations. In the present case the echocardiographic appearances of aortic valve vegetation confirmed the clinical diagnosis. While transesophageal echocardiography is far superior in demonstrating lesions, it too has significant limitations[5,6].

The diagnosis of *C. burnetii* IE is based on the identification of *C. burnetii* in affected valve tissue obtained at operation or necropsy, or on the demonstration of a high antiphase I Ig G antibody titer > 1:800 of this organism according to new Duke criteria[7]. In the present case, serology titers do not meet the threshold for major criteria and the high phase II titers suggesting acute rather than chronic infection. The definite endocarditis diagnosis of our case was based on Dukes criteria such as presence of vegetation on a bicuspid aortic valve with severe aortic regurgitation, fever, mycotic aneurysms and intracranial hemorrhage[8].

The present case emphasises that chronic Q fever infection must be considered in the differential diagnosis of IE, particularly if there is a history of contact with farm animals and if blood cultures are negative.

The clinical course of Q fever endocarditis is often severe. The mortality among cases reported was 40%[9].

The optimal treatment of Q fever IE has not been completely defined, in part due to the difficulty of its culture. Prolonged doxycycline combined with fluoroquinolones treatment may be the preferred treatment for chronic Q fever. Patients should be followed closely with clinical assessment, echocardiography and laboratory investigation, including phase I serology, at regular intervals[10].

CONCLUSIONS

Unlike Q fever, the clinical presentation of chronic *C. burnetii* IE may be rare. *C. burnetii* should be considered when culture-negative IE are encountered. Molecular diagnostic methods are important tools for the diagnosis of *C. burnetii* EI.

There are no conflicts of interest.

Written informed consent was obtained from the patient for publication of this case report.

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