Concurrent aortic valve replacement and splenectomy for Q-fever endocarditis with massive splenomegaly and pancytopenia

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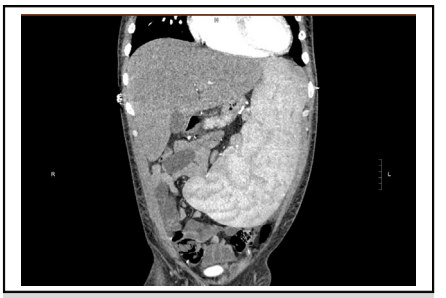


A patient with massive splenomegaly associated with Q-fever endocarditis.

CENTRAL MESSAGE

We present a unique case of a patient with Q-fever endocarditis presenting with massive splenomegaly and pancytopenia who underwent concurrent aortic valve replacement and splenectomy.

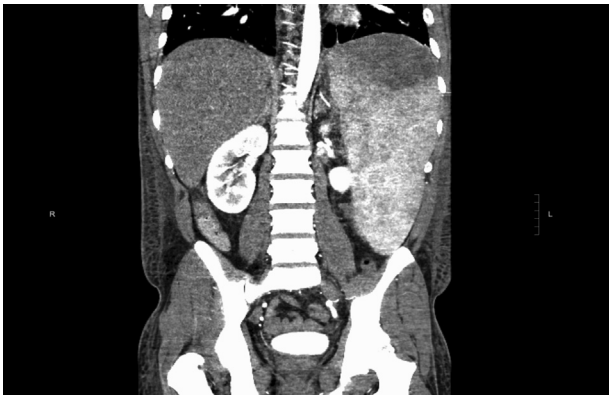
Video clip is available online.

Q-fever is a rare zoonotic cause of culture-negative endocarditis often associated with splenomegaly. We present a unique case of a 28-year-old male with Q-fever endocarditis who presented in acute decompensated heart failure with particularly massive splenomegaly and profound pancytopenia. Several aspects of his clinical presentation highlight the complexity of managing multiorgan disease in endocarditis and the importance of multidisciplinary planning for optimal outcomes.

CASE DESCRIPTION

A 28-year-old male with a history of bicuspid aortic valve (BAV) and moderate aortic regurgitation (AR) presented to the emergency room with several months of daily fevers and abdominal pain. He had sinus tachycardia to 140 bpm and was pancytopenic (white blood cell count, 1300/μL; platelet count, 83,000/μL; and hemoglobin, 9.1 g/dL). Computed tomography angiography (CTA) revealed massive splenomegaly (33 cm $\times$ 23 cm $\times$ 12 cm) with several splenic infarcts and a small focus of arterial blush (Video 1). The CTA also demonstrated a 2 cm $\times$ 1 cm aortic root abscess versus a pseudoaneurysm and associated nodular thickening of the BAV leaflets concerning for vegetation. Transthoracic echocardiography (TTE) demonstrated an aortic valve mass with severe AR and severe aortic stenosis (AS) with a mean gradient of 42 mm Hg, trace mitral regurgitation, mild tricuspid regurgitation, normal left ventricular (LV) size, and an LV ejection fraction of 37% (Video 2). Blood cultures were obtained, and broad-spectrum antibiotics were initiated. The patient was not an intravenous drug user, and the sole suspected infectious source was a routine dental procedure completed 4 months earlier.

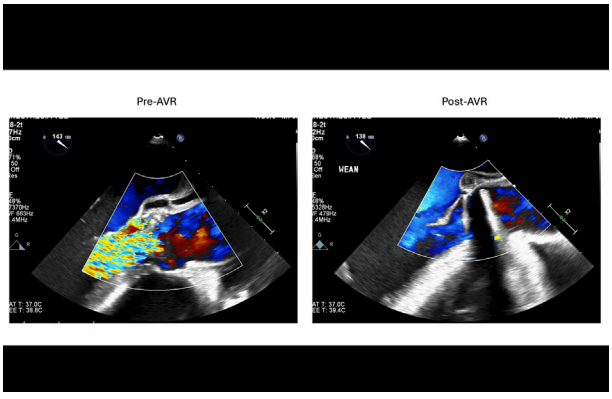
After initial resuscitation, the patient was taken to the operating room for concurrent splenectomy and aortic valve replacement with possible root replacement on day 2 of his hospitalization. His surgery was expedited because he was considered at relatively high risk of rapid deterioration. This unique operative plan was based on several factors. With severe AR, acute decompensated heart failure, and a known aortic valve vegetation, the patient clearly required urgent surgical intervention on the aortic valve and potentially the root as well. However, the safety of systemic heparinization was uncertain given the possible splenic arterial blush on CTA. Embolization of the arterial blush was considered but ultimately ruled out, as there was no hemodynamic compromise, a distal embolization was deemed of



VIDEO 1. Preoperative coronal computed tomography angiography demonstrating massive splenomegaly. Video available at: https://www.jtcvs.org/article/S2666-2507(25)00060-4/fulltext.

limited utility because of its location, and proximal embolization would cause significant necrosis and pain and increase the technical difficulty of the splenectomy. Additionally, the patient's massive splenomegaly likely was a significant contributor to his pancytopenia, and postoperative hemostasis could be challenging in the setting of ongoing platelet sequestration. Therefore, the splenectomy was planned prior to the cardiac portion owing to the combined risk of splenic hemorrhage with heparinization and postoperative platelet sequestration.

Several aortic valve replacement (AVR) options were considered. It was felt that both a homograft or biological valve would be suboptimal, given that the patient was 28 year old and would need further operations down the line. While a Ross might have been possible, we prioritized a shorter clamp time given the patient's degree of illness at presentation and the fact that he already would be undergoing a complex concomitant procedure with the splenectomy.



VIDEO 2. Intraoperative transesophageal echocardiography clips before and after aortic valve replacement. Video available at: https://www.jtcvs.org/article/S2666-2507(25)00060-4/fulltext.

In the operating room, the general surgery team began with a midline laparotomy, followed by dissection and division of the splenic vessels and removal of the spleen (Figure E1). As expected, the spleen was found to be extremely enlarged, occupying the entire left hemiabdomen. Once adequate hemostasis was achieved, the abdomen was packed and temporarily closed. Given the need for systemic heparinization, we felt it prudent to leave the abdomen open for a second look for hemostasis after the cardiac portion of the case was completed.

For the cardiac portion of the case, a median sternotomy was followed by standard central aortic and dual-stage right atrial cannulation. A root vent and retrograde cardioplegia catheter were placed. Cardiopulmonary bypass (CPB) was initiated, an LV vent was placed through the right superior pulmonary vein, the aortic cross-clamp was applied, and the heart was arrested with antegrade followed by retrograde Del Nido cardioplegia. There was a low threshold to open the aorta and administer ostial cardioplegia had there been any difficulty with the initial arrest, but this was not necessary. Following oblique aortotomy, the aortic valve was thoroughly inspected, which revealed a severely calcified Sievers 1 BAV with L/R fusion, with infectious destruction of all leaflets. A pseudoaneurysm with a 5-mm mouth was identified under the noncoronary cusp. The cavity appeared to be chronic with no active infection or purulence, as further evidenced by an endothelialized inner surface.

Consideration was given to exclusion of the cavity with a pericardial patch; however, it was determined that the pseudoaneurysm cavity could be easily excluded with the valve sutures alone. The aortic leaflets were then excised, and the aortic root was copiously debrided and irrigated with saline, followed by an 80 mg/L gentamicin solution.

Next, valve sutures were placed in inverting fashion around the aortic annulus. In the area of the pseudoaneurysm, 3 valve sutures were placed deep into the aortomitral continuity to fully exclude the cavity. The annulus was sized to a 25-mm On-X valve, which was seated and tied into position. The aortotomy was closed, the heart was deaired, and the cross-clamp was removed. After reperfusion and an additional round of deairing, the patient was weaned off CPB without incident. TEE demonstrated unchanged LV and right ventricular function, no paravalvular leak, trivial mitral regurgitation, and no ongoing flow into the excluded pseudoaneurysm. CPB and cross-clamp times were 106 minutes and 72 minutes, respectively.

Once the chest was closed and the cardiac portion of the case completed, the general surgery team returned to remove the packing from the abdomen, interrogate the splenic bed, and ensure hemostasis. This was completed without incident, and the abdomen was closed.

Postoperatively, the patient made an excellent recovery. He was extubated within 6 hours and was weaned from

inotropic support by postoperative day (POD) 1. He was transferred to the floor on POD 2, where his chest tubes and temporary pacing wire were removed. A TTE on POD 6 demonstrated a well-seated mechanical aortic valve with mean gradient 11 mm Hg, no AR, and LV ejection fraction 41%. Postoperatively, all his cell lines also recovered to normal levels; by POD 5, he had a platelet count of 414,000/μL, a white blood cell count of 7200/μL, and a hemoglobin concentration of 9 g/dL. At this time, there had still had been no growth on several sets of blood cultures.

On the recommendation of our infectious disease colleagues, a Q-fever antibody screen with titer reflex was sent, in addition to 16S bacterial ribosomal sequencing on the aortic valve specimen sent from the operating room. Both the antibody screen and 16S sequencing detected *Coxiella burnetii*, with significantly elevated antibody titer levels further confirming the diagnosis of Q-fever. On further discussion, the patient revealed that he had significant exposure to farm animals that had recently given birth at his parents' ranch, where he was presumably exposed to *C. burnetii*. His antibiotic regimen was changed accordingly to doxycycline and hydroxychloroquine, and he was discharged to home on POD 7.

DISCUSSION

C. burnetii is an obligate intracellular gram-negative bacteria and a rare but described source of endocarditis, with 1 review attributing an estimated 5% of worldwide endocarditis cases to this zoonotic bacteria.[1] The same review estimated that splenomegaly is present in one-half of patients with Q-fever endocarditis.[1] In cases of massive splenomegaly and endocarditis (of any microbial etiology), the traditional timing of operations is splenectomy followed by AVR, to prevent any reinfection of the newly implanted valve[2]; however, there are several reports of performing AVR followed by splenectomy with satisfactory results.[3,4] Concurrent splenectomy and AVR has been reported in a few clinical contexts—specifically, in cases of large splenic abscesses with hemodynamically significant endocarditis[5] and cases of severe immune thrombocytopenia associated with severe AS.[6] In our patient, severe pancytopenia coupled with concern for a splenic bleed and hemodynamically significant AR is what ultimately prompted us to perform the operations concurrently.

This case underscores how even the most complex of multiorgan endocarditis cases can be approached safely with proper preoperative planning and multidisciplinary care to achieve excellent clinical results. Further study is warranted on the short- and long-term outcomes of patients who undergo concurrent splenectomy and valvular cardiac surgery.

Conflict of Interest Statement

Dr Forrester reported serving as PI for investigator-initiated clinical trials funded by Varian and Pacira, as PI for industry-initiated clinical trials funded by Eclipse Regenysis, and as a consultant for Costa Surgical. All other authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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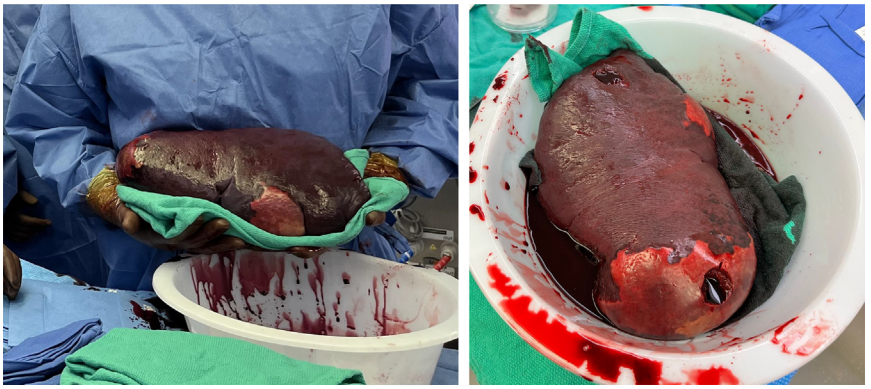

FIGURE E1. Photos of the enlarged spleen after removal.