The Heart Surgery Forum #2010-1114
14 (2), 2011 [Epub April 2011]
doi: 10.1532/HSF98.20101114

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Aortic Dissection Caused by Giant Cell Arteritis

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ABSTRACT

Aortic dissection is a very serious condition mainly caused by degenerative diseases of the connective tissue and hypertension. Ascending aortic dissection as a consequence of aortitis in association with giant cell arteritis is very rarely seen. In this article we report on the successful surgical repair of a Stanford type A aortic dissection caused by giant cell arteritis in a 74-year-old patient. We could visualize this dissection via echocardiography and computed tomography. Histopathology confirmed this rare complication of giant cell aortitis.

CASE REPORT

A 74-year-old Caucasian man was referred to the emergency department of our hospital soon after he collapsed at home. He noted progressive chest pain associated with dyspnea, fatigue, and arterial hypotension. The patient's history included a giant cell arteritis of the temporal artery diagnosed 1 year previously.

Upon admission to the hospital, the patient presented a blood pressure of 110/50 mmHg and a heart rate of 120 beats per minute (bpm). Transthoracic and transesophageal echocardiography depicted ascending aortic aneurysm with ambiguous dissection. The aortic annulus was dilated, and the aortic valve was tricuspidal and slightly insufficient. The intimal flap was floating approximately 20 mm above the aortic cusps within a large aneurysmatic ascending aorta (Figure 1A). Subsequently, a contrast-enhanced echocardiography (ECG)-gated 64-slice spiral computed tomography (CT) confirmed the circular aortic dissection and the aneurysm of the ascending aorta; the descending thoracic aorta was normal (Figure 1B and 1C).

The patient was taken to the operating theater urgently, where a median sternotomy was performed and cardiopulmonary bypass was installed through aortic arch–right atrium cannulation. Intraoperatively, there were approximately 300 mL of hemopericardium, and the ascending aorta appeared dilated and partially hemorrhagic. The aneurysm of the ascending aorta was present until the brachiocephalic trunk origin. The longitudinal aortotomy demonstrated that circular dissection started 20 mm above the aortic annulus, leaving the coronary arteries and ostia unaffected. The aortic annulus was slightly dehiscent. Afterward, the aortic annulus was resutured in the anatomic position with a Dacron 28 mm aortic prosthesis replacing the ascending aorta.

The resection specimen (70 × 60 × 4 mm) showed a longitudinal intimal tear 65 mm in length associated with dissection and fresh bleeding into adjacent layers of the aortic wall including the adventitia (Figure 2A). The surrounding intima displayed only a few atherosclerotic plaques. Histological examination revealed discontinuous granuloma-like inflammatory infiltrates composed of mononuclear cells and multinucleated giant cells associated with destruction of the elastic tissue of the tunica media (Figure 2B). The inflammatory process was transmural, with a segmental discontinuous distribution affecting mainly the media (Figure 2C and 2D).

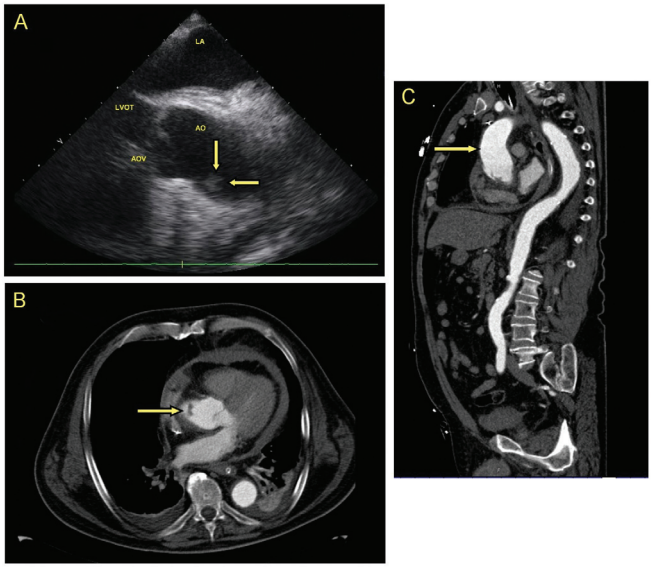

Figure 1. A, Transesophageal echocardiogram (ECG) of the type A aortic dissection. Longitudinal scan of the aortic root revealed a floating intimal flap within a large ascending aorta (arrows). AO indicates ascending aorta; AOV, aortic valve; LVOT, left ventricular outflow tract; LA, left atrium. B, Contrast-enhanced ECG-gated 64-slice spiral computed tomography (CT) demonstrated the circular aortic dissection and the aneurysm of the ascending aorta (arrows). C, The descending thoracic aorta shows no dissection and a normal diameter.

Received August 17, 2010; accepted September 24, 2010.

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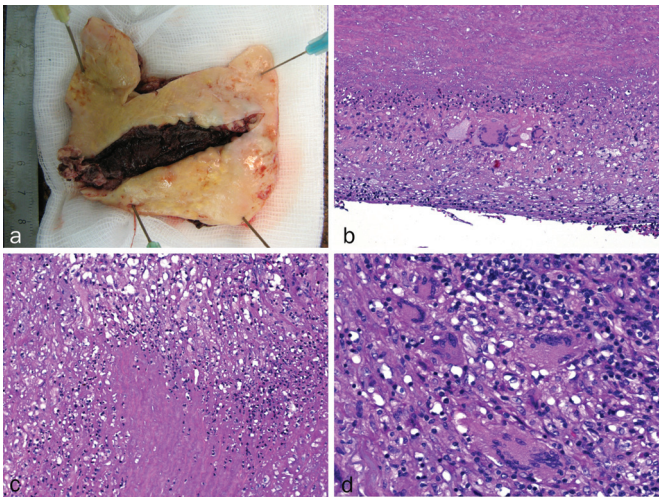

Figure 2. A, Gross specimen from the aorta showed extensive dissection with clotted blood in the aortic wall (note minimal atherosclerotic plaques). B, Prominent giant cell–rich inflammation obliterating the intima (below) and abutting the tunica media (above). C, Elastic tissue of the media (lower midfield) interrupted by inflammatory aggregates surrounded by palisading histiocytes (upper midfield). D, Higher magnification showed lmyphoplasmacytic infiltrates with prominent multinucleated Langhans-type giant cells.

DISCUSSION

Aortitis is a chronic, progressive disease typically associated with a variety of systemic disorders including giant cell arteritis, Takayasu arteritis, and juvenile rheumatoid arteritis [Ryder 2009]. Isolated cases of aortitis have been described in patients with systemic lupus erythematosus, scleroderma, rheumatic fever, and Kawasaki syndrome [Oberwalder 2003]. Aortitis can lead to aneurysm and dilatation because of medial destruction of the aortic wall with disruption and depletion of elastic fibers, but acute aortic dissection is extremely rare and often a late complication of an inadequately treated disease.

Giant cell arteritis (GCA) is a focal granulomatous inflammation of large and medium-sized arteries. The vessel most often affected is the temporal artery. GCA in association with ascending aortic dissection is very rarely seen. In a review of 24 patients with thoracic aortic dissection caused by giant cell aortitis, Liu and colleagues found that 11 of 24 patients had no previous clinical symptoms referable to the cranial arteries [Liu 1995].

Transesophageal ECG and CT are the standard diagnostic tools to clarify acute pathology of the ascending aorta. In patients with an inflammatory aneurysm, both methods are often unable to lead to the correct diagnosis. In such cases, uncharacteristic symptoms such as fever of unknown origin, progressive dyspnea, fatigue, and dizziness are present. Inflammatory aneurysms of the ascending aorta account for 1% to 5% of all operated aortic aneurysms [Bedeleanu 2000]. The healthy aortic wall does not contain any inflammatory cells, thus the presence of inflammatory cells in the aortic wall is indicative of aortitis. In the present case, the inflammation was characterized by disruption of the media and bleeding into adjacent layers of the aortic wall with patchy transmural chronic and focally acute inflammatory infiltrates.

Previous studies suggest that inflammatory thickening of the aortic wall is normal at the time of GCA diagnosis and that aortitis may be the first manifestation of GCA-associated aortic complications [Marie 2009]. Whether aortitis leads to vascular wall injury responsible for the late onset aneurysmal disease or even aortic dissection remains to be determined.

In conclusion, we recommend long-term monitoring for aortic aneurysms, especially in high-risk patients. Further studies are required to define the optimal frequency and imaging modality for patients with GCA.

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