Rupture of an Aneurysmal Aortic Diverticulum Associated With Coarctation and Bicuspid Aortic Valve

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A 28-year-old soccer player presented with collapse and left-sided chest pain during a prematch warm-up. Pain was also present in the left side of the neck. He had not suffered from chest pain before. A cardiac murmur had been described in childhood and attributed to pulmonary stenosis but never followed up.

On initial examination, he was hypotensive, with a systolic murmur in the aortic area and a pulsatile mass in the left supraclavicular fossa (Figure 1A). There was no blood pressure differential between arms, but the femoral pulses could not be felt. A chest radiograph (the first the patient had ever received) revealed widening of the left upper mediastinum (Figure 1B). Contrast-enhanced computed tomography showed an aneurysmal diverticulum of the aorta that extended into the neck and gave rise to a normal-sized left subclavian artery (Figure 1C). The wall was markedly thickened, with high attenuation before contrast, in keeping with an intramural hematoma. Coarctation of the aorta was identified distal to the aneurysm (Figure 1C). There was no rib notching.

Figure 1. A, Close-up photograph of left-sided neck swelling (arrowed). B, Chest radiograph. C, Postcontrast computed tomography image demonstrating the aneurysm’s thickened wall (white arrows) and coarctation (arrowhead). D, Magnified 3-dimensional reconstruction showing relationship between left subclavian artery and aneurysm. E, Computed tomography image of aberrant left subclavian vein (arrows) passing between the aortic arch and pulmonary arteries. F, Still of color flow across coarctation. Inset, Computed tomography rotated to match image orientation; direction of blood flow is arrowed.
In a 3-dimensional reconstruction (Figure 1D), the aneurysm was vertically orientated and joined at its midbody to a normal portion of the distal arch in a T-junction configuration. The dilatation cranially displaced the origin of the left subclavian artery into the neck. Although the origins of the brachiocephalic arteries were otherwise normal, the left brachiocephalic vein was unusual in its course, passing beneath the aortic arch (Figure 1E).

Transesophageal echocardiography confirmed the intramural hematoma and showed turbulent blood flow through the coarctation (Figure 1F; online-only Data Supplement, Movie 1). The aortic valve was mildly stenosed and bicuspid owing to fusion of the right and left coronary cusps (online-only Data Supplement, Movie 2).

A 2-stage operation was undertaken. Through a transverse incision in the left anterior triangle, a left common carotid artery to left subclavian artery bypass was performed and the apex of the aneurysm ligated. A left thoracotomy was then performed that drained 750 mL of blood and showed extensive mediastinal hematoma (Figure 2A). The left femoral vessels were cannulated for cardiopulmonary bypass and hypothermic circulatory arrest at 16°C. The aneurysmal diverticulum arose as an outpouching from the arch. The intimal tear from which the hematoma had arisen was clearly identifiable (Figure 2B). The diverticulum was overswn within the chest, and both aneurysm and coarctation were excised. A graft was placed between the arch and descending aorta (Figure 2C). Recovery was uneventful.

In the absence of an aberrant right subclavian artery or a right aortic arch and aberrant left subclavian artery, the diverticulum is not classically that of Kommerell. Its position does not fit with a diverticulum that arises from the aortoductal junction. The aortopathy associated with a bicuspid aortic valve and coarctation may have contributed to its aneurysmal transformation and rupture.

Disclosures

None.

References

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Circulation. 2011;123:102-103
doi: 10.1161/CIRCULATIONAHA.110.958355

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