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Interruption of the aorta with multilobulated arch aneurysms: A new clinicopathologic entity

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The aortic arch occupies an extremely strategic position with a capacity to influence blood supply to virtually all parts of the body. In addition, the aortic arch is surrounded by many vital structures. To date, several congenital and acquired abnormalities affecting the aortic arch have been described and well characterized from the clinical and pathologic points of view.¹

We here describe a “new” condition of complete or partial replacement of the aortic arch and some of its branches by multilobulated “aneurysms” that are interconnecting by a series of small openings and therefore are obstructive. In addition, we discuss its management and the possible developmental origin.

Clinical Summaries

PATIENT 1. A 26-year-old woman had a short history (approximately 6 months) of shortness of breath and diminished upper and lower limb pulses and a suprasternal pulsatile mass. Cardiovascular magnetic resonance (CMR) showed tapering of the aortic arch followed by interruption, bypassed by a $4 \times 5 \times 6$ -cm multilobulated aneurysm located superior to the expected curve of the arch (Figure 1, *a* and *b*). There was compression of the distal trachea. The tortuous distal aortic arch gave rise to anomalous, posteriorly located right and left subclavian arteries. No left common carotid artery was identified. The left brachiocephalic vein passed behind the ascending arch.

Repair was undertaken with deep hypothermic circulatory arrest at 15°C. Postoperatively, the patient had transient right leg weakness, which resolved completely. Postoperative CMR study (Figure 1, *c*) demonstrated a patent graft and bilateral vertebral arterial flow to subclavian arteries with cerebral arterial steal. At 8 months' follow-up, she remained well, participating in sports with no signs of subclavian steal. Her pulses were strong and equal in all extremities.

PATIENT 2. A 47-year-old woman was known to have had a heart murmur since childhood but was well until 36 years of age, when she was evaluated for left upper extremity weakness, which was treated conservatively. Ten years later she began having chest pain and dyspnea on exertion. CMR showed coarctation of a right-sided aortic arch followed by a tortuous, multilobulated aneurysmal sac located to the right of the ex-

Figures 1a and b. Contrast-enhanced magnetic resonance angiography showing anteroposterior and left lateral views of the aneurysmal aortic arch of patient 1. **c.** Contrast-enhanced magnetic resonance angiography showing a left lateral view of the aneurysmal aortic arch of patient 1 after graft repair.

Figures 1d and e. Contrast-enhanced magnetic resonance angiography showing left lateral and right lateral views of the aneurysmal aortic arch of patient 2.

pected curve of the right-sided arch. The maximal aneurysmal diameters were $6.5 \times 6 \times 6$ cm (Figure 1, d). In addition to left subclavian artery stenosis, the right subclavian and vertebral arteries originated from a 2-cm aneurysmal sac (Figure 1, e).

The aneurysms were excised with the use of hypothermic circulatory arrest at 15°C.

She remained clinically well with no neurologic or cardiopulmonary symptoms during the 2-year follow-up.

Histologic Findings

Histologic examination of the excised aneurysm (Figures 2 a, b) showed loss of medial smooth muscle cells, unusual widespread calcification, and cholesterol clefts together with focal accumulations of macrophages and foam cells. Gram staining and cultures of the aortic tissues were negative for microorganisms.

Discussion

This report describes for the first time a distinct clinicopathologic syndrome, which consists of tapering of the midportion of the aortic arch followed by replacement of the main channel by “cavernous” tissue consisting of thin-walled multilobulated aneurysms measuring between 1 cm and 5 cm and separated by thin-walled septa consisting of disorganized arterial wall with medial degenerative changes. The anomaly can involve the origin of one or more of the head vessels, which may be anomalous in origin. These features serve to distinguish this congenital anomaly from acquired false aneurysms associated with atherosclerotic pseudoaneurysm.

The anomaly appears to progress slowly with expansion of the thin-walled cavities, which are partially calcified. The affected patients can be asymptomatic for long periods of time and present with symptoms resulting from pressure on surrounding structures, hypoperfusion of affected regions, or inequality in pulses between the two upper limbs or between the upper and lower limbs. Although the blood pressure in the two patients described here was only mildly elevated, the obstruction coupled with renal ischemia could be expected to produce chronic systemic hypertension.

We believe that all patients with this anomaly should undergo surgical repair under deep hypothermia and circulatory arrest to enable complete excision of the thin-walled aneurysms, which could be adherent to surrounding structures.

Although the exact pathogenesis is not known, it is likely to be the result of failure of vascular remodeling during vasculogenesis.²

Figures 2a and b. Histologic features of the aneurysms wall of patient 2 showing the loss of smooth muscle cells and the extensive degeneration and calcification.

Developmental remodeling is an intricate process, through which the newly developed vasculature undergoes a series of changes in size, shape, and function in response to tightly regulated molecular cues as well as hemodynamic factors, with close interaction between endothelial cells and pericytes.^{3,4} Dysregulation of remodeling can result in different vascular anomalies, of which the syndrome described here could be one.

It is hoped that this report will help in the early diagnosis and management of the syndrome, as well as in stimulating research into vasculogenesis.

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