Remnant of repaired right aortic arch stump as a rare cause of recurrent strokes: The eyes see only what the mind knows!

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Anomalies of the aortic arch complex (vascular rings) occur in various forms, often causing tracheal and esophageal compression.¹⁻⁵ We describe a rare case of recurrent strokes resulting from thrombus in remnant of a repaired right aortic arch stump.

CLINICAL SUMMARY

A 50-year-old man with spina bifida was seen with a history of recurrent strokes during the previous 3 years. He had undergone ligation of patent ductus arteriosus and “aortopexy” through left thoracotomy 40 years previously. Seven years previously, he had had dysphagia caused by esophageal compression, which was attributed to vascular ring equivalent resulting from right-sided “cervical” aortic arch with an aberrant left subclavian artery coming off a remnant of aneurysmal mid-descending aorta. The right arch coursed leftward at the level of the carina, crossing the spine and compressing the esophagus before descending on the left side of spine with aneurysmal dilatation of the mid-descending aorta.

The esophagus was compressed by the confluence of the dilated distal right aortic arch/proximal descending thoracic aorta, the carina, and the azygos vein. At that time, the patient

underwent a left thoracotomy under cardiopulmonary bypass, and a 22-mm graft was attached to the side of the ascending aorta, brought down anterior to the right lung hilum, and anastomosed end-to-end to the descending aorta near the diaphragm. The right-sided aortic arch was then oversewn and tied off proximally to the right of the vertebral columns. The left subclavian artery was attached with an 8-mm tube graft to the descending aortic graft. The patient recovered well postoperatively and continued to do well until 3 years before presentation, when he had the first of 3 strokes. Brain imaging demonstrated different areas of infarction in the right cerebellum and occipital lobes after each stroke, suggesting proximal embolic episodes involving the dominant right vertebral artery. There was no thrombus in the left atrium or left atrial appendage according to echocardiography. The computed tomographic scan of the aorta revealed thrombus in the aortic arch remnant, which was thought to be the likely source of emboli (Figure 1, A and B). On the basis of the patient's recurrent strokes despite aggressive and escalating antiplatelet and anticoagulant therapies, surgical exclusion of the thrombosed arch remnant was planned. Surgery was performed through a median sternotomy with cardiopulmonary bypass and circulatory arrest. The aortic arch was disconnected from the right carotid and subclavian arteries, and a vascular graft was sewn around their common origin (Figure 2, A-D). The proximal anastomosis was done around the left carotid origin, ascending aorta, and previous tube graft to the ascending aorta, just above the sinotubular junction. The arch remnant was full of organized clots and was oversewn (Figure 2, B and C). The postoperative computed tomographic scan showed no flow in the ligated right arch (Figure 1, C), and the patient had an uneventful recovery.

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FIGURE 1. Computed tomographic scan showing the remnant of the right arch. A and B, Preoperative scan. Note large amount of thrombus in the arch remnant (arrow). C, Postoperative scan. Interval placement of graft from ascending aorta to right subclavian and carotid arteries. The arch remnant is now excluded from the circulation.

FIGURE 2. Intraoperative photographs. A, View of the distal ascending aorta and proximal arch (*Dasc/PrArch*). Note the separation between the origins of the right subclavian and carotid arteries (*left forceps*) and left common carotid artery (*LCCA*). The *arrow* points to the previous ascending aorta–descending graft anastomosis. B, Thrombus (*T*) in the aortic arch remnant, which was responsible for patient’s recurrent strokes. C, After excision of the intervening ascending aorta and proximal arch. The *arrow* points to the previous ascending aorta–descending graft anastomosis. D, Completed anastomosis. *Arrow* points to the reimplanted right subclavian artery (*RSCA*) and right common carotid artery (*RCCA*).

DISCUSSION

Clinical presentation and surgical approaches to aortic arch complex (vascular rings) anomalies depend on the specific lesion and presence of any associated congenital heart defects.^{1–5} In most patients, vascular rings manifest early in life, and respiratory symptoms are more common than evidence of esophageal compression. When they manifest in late childhood or adulthood, however, dysphagia is often the primary symptom.^{1–5} The major subtypes of the aortic arch complex anomaly requiring surgical correction have been well described.^{1–5} Patients with vascular rings require a high index of suspicion for correct diagnosis, and surgical therapy can be accomplished with very low morbidity, nearly zero mortality, and excellent long-term outcomes.^{1–5} In 2005, Backer and colleagues³ reported that among 209 patients undergoing surgery for complete vascular rings, they had seen no operative deaths since 1959.

Postoperative morbidity is chiefly related to airway issues (tracheomalacia and bronchomalacia).^{1–5} Recurrence of symptoms related to repaired vascular ring anomalies is rare. Ruzmetov and colleagues⁴ reported 96% survival at 35 years in their series of 183 patients undergoing surgical repair for vascular rings, with no patient showing any evidence of recurrent vascular ring anomalies at a median follow-up of 6 months.

Our patient was seen with recurrent strokes related to thrombus in the arch remnant. To the best of our knowledge, this complication after repair of vascular ring anomalies has not been previously described. What made the diagnosis a challenge was to correlate the embolic source of stroke to the arch remnant after more common etiologies causing stroke were ruled out. Careful assessment of imaging studies was the key to accurate diagnosis and appropriate surgical management through exclusion of the arch remnant from circulation.

This case demonstrates that an arch remnant after surgery for aortic arch anomalies could be a potential source of thromboembolism, and a high index of suspicion is necessary for accurate diagnosis and appropriate surgical management.

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