Are Congenital Arteriovenous Malformations of Proximal Upper Extremity, More on the Right: Case Report and Literature Review

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Int J Angiol 2016;25:e29–e31.

Abstract

Keywords

- AV malformation
- subclavian
- artery
- excision
- endovascular technique
- arteriovenous fistula

This is a case of a 9-year-old boy with a congenital arteriovenous malformation (AVM) of the right upper extremity arising from the right subclavian artery. He underwent open surgical excision of the vascular tumor. Upon reviewing the literature, a high incidence of right-sided congenital AVM of the proximal upper extremity was identified. It has been suggested that this high incidence may be related to the complexity of the embryologic development of the right subclavian artery.

Vascular anomalies, commonly vascular tumors or vascular malformations, are usually developmental in origin. Vascular malformations can be arterial, venous, lymphatic, or a combination of these.¹ We report the surgical management of a congenital right subclavian arteriovenous malformation (AVM) and review the literature on such lesions, which reveals an increased frequency of occurrence of these lesions on the right side.

Case Report

A 9-year-old, 29 kg male child was admitted with history of an enlarging swelling over his right supraclavicular region of 1-year duration. It was associated with an area of hyperpigmentation over the right axilla and anterior axillary fold which he had noticed over the last 10 months and pain in the arm for the last 6 months. A bruit could be auscultated over the right upper lung field.

Computed tomographic (CT) angiogram revealed the presence of an AVM with arterial feeders mainly from the right

subclavian artery and a few from the right vertebral artery (→**Fig. 1**). The venous drainage was through the right subclavian vein. The echocardiogram showed normal ventricular function with normal chambers and valves. Electrocardiogram showed normal sinus rhythm with right axis deviation. The blood investigations were found to be normal.

The patient was scheduled for excision of the vascular malformation. He was premedicated with ranitidine 75 mg and diazepam 2.5 mg. The monitoring included 5-lead electrocardiogram, a blood pressure cuff on the left arm and a pulse oximeter on the left hand. Anesthetic induction with thiopentone, fentanyl, and neuromuscular blockade with vecuronium bromide was uneventful. The trachea was intubated with a 6.0-mm ID cuffed endotracheal tube. The left radial artery was percutaneously cannulated with a 22 G catheter for arterial pressure monitoring. The malformation was accessed via a supraclavicular incision extending into the axilla. Dissection was difficult due to a large number of collaterals. The subclavian vein and artery were dissected underneath the clavicle. Feeders from the internal mammary,

Fig. 1 Computed tomographic angiogram showing the arteriovenous malformation involving the right subclavian artery.

thoracodorsal, and cervicodorsal arteries were ligated. Proximal control of the subclavian artery was gained at this stage. The sac of the malformation was dissected on the anterior chest wall. Upon opening the sac, there was significant blood loss, which had to be managed by rapid infusion of intravenous fluids and blood. The total estimated blood loss was around 1,000 mL and required transfusion of 2 units of packed red blood cells. The vascular communications to the sac were then closed directly. Following this, the sac was completely closed after disconnecting all the systemic connections.

Postoperative period was uneventful. Repeat CT angiogram showed no residual lesions. Post-op chest X-ray was normal. The patient was discharged home after 1 week. He is currently on follow-up with us every 3 months, shows no residual lesions, and is symptom free.

Histopathology of the lesion showed a conglomeration of medium-sized arteries, veins, and vessels suggestive of an AVM. The veins were in various stages of "arterialization" and demonstrated intimal thickening, increased smooth muscle within the media, and dilatation of the vasa vasorum. There was dilatation of the proximal arteries, with fibrosis, thinning of the media, and diminished elastic tissue.

Discussion

The normal right subclavian artery emerging from the brachiocephalic trunk has got three embryological components:¹ the most proximal segment of the artery is derived from the right fourth arch²; the middle segment is produced by the cranial portion of the right dorsal aorta; and³ the most distal segment is developed from the right seventh intersegmental artery (→ Fig. 2). The distal portion of the right dorsal aorta beyond the origin of the right seventh intersegmental artery disappears during normal embryological processes. The left subclavian artery on the contrary has a rather uncomplicated embryological origin from the left seventh intersegmental artery. Variations in the origin of the right

Fig. 2 Embryologic origin of the right subclavian artery.

subclavian artery are well described in the form of anomalous right subclavian artery (ARSA), arising from the descending thoracic aorta, and are thought to result from embryological abnormalities.

The development of the vascular system may be described in three stages. The first step is the condensation of undifferentiated cells into capillary blood spaces. The next stage involves the formation of a retiform plexus in which blood flows from an arterial to a venous side. The last stage involves the development of axial arteries in the limb buds. Developmental arrest can occur at any point during vessel formation, creating different types of vascular malformations.

The embryological development of the proximal aorta and the limb buds can often explain the course and multiplicity of the AVMs of the proximal upper extremity. It appears that the arterial and venous connections are consistent with abnormal fistulous communications between originally normal embryological arterial and venous channels. The therapeutic implications of these vascular anastomoses are obvious and they guide the appropriate management plan.

Although the right subclavian artery is embryologically derived from several components, the uncommon reporting of subclavian AVMs makes it difficult to comment on the preferential lateralization of these lesions to either the right or the left. We searched the literature for studies and cases which report the side of congenital subclavian AV fistulae²⁻⁷ and came across 11 such patients, 9 of whom had right-sided lesions. Including our case to the list makes it a total of 12 patients of whom 10 had congenital AVMs involving the right subclavian artery. Although this sample size is admittedly small to make definite observations, we believe that congenital subclavian AVMs have a greater propensity to occur on the right than on the left, reflecting greater complexity of the embryologic origin of the right subclavian artery. Stanley and Menzoian⁵ record similar observations in their case report of a right subclavian congenital AVM. Embryology may also be the explanation for the occurrence of other anomalies of the

right subclavian artery such as the ARSA or the retroesophageal right subclavian artery.

Therapeutic options for AVMs include endovascular, surgical, radiation therapy, or a combination of all of these. Angiography is usually utilized only after treatment planning is complete and is employed for superselective endovascular embolization before surgical intervention. Superselective endovascular embolization may have a role in palliation of symptoms, or it could be used as primary therapy for surgically inaccessible AVMs. The only therapy that carries any hope for long-term success is total resection of the tissue that is involved with the AV anomaly. Another technique to gain proximal control of the malformation might have been to use balloon tamponade during the dissection. However, the unavailability of the services of an interventional radiologist precluded use of such endovascular techniques.

Surgical excision of AVMs is rarely curative. These lesions have a high recurrence rate and many patients may require amputation of the extremity. Long-term follow-up of these cases is mandated.

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