

# An unusual arteriovenous malformation involving the cervical vessels treated with endovascular repair

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## ABSTRACT

We present an unusual and complex arteriovenous malformation involving the vertebral artery, subclavian artery, and internal jugular vein in a 31-year-old man with no history of trauma or catheterization. The repair was done using endovascular techniques to minimize complications from nerve or vascular injury. The massively dilated jugular vein has remained diminished in size and the patient has remained asymptomatic at 8 months. We discuss the occurrence of this rare malformation as well as treatment options along with their risks and benefits. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:136-8.)

**Keywords:** Arteriovenous malformation; Endovascular procedures; Internal jugular vein; Subclavian artery; Vertebral artery

Spontaneous or congenital arteriovenous malformations in the cervical region are rarely reported. The most comprehensive analysis of the incidence of these types of malformations was reported by Greenberg,<sup>1</sup> who found 19 cases in his review of the literature in 1970. They may be difficult to treat, with significant risks of bleeding, cranial nerve injury, and vessel injury. Often, they have required treatment with complex open operative techniques. We present an endovascular treatment of a young man with complex connections between his right vertebral artery, right subclavian artery, and right internal jugular vein. The markedly dilated right internal jugular vein has continued to be diminished in size at 8-month follow-up. The patient has not had any complications from the endovascular repair. Appropriate consent for publication was obtained from the patient.

## CASE REPORT

The patient is a 31-year-old man with no previous medical history. His wife, who was a nurse, first detected the presence of an arteriovenous malformation after hearing a bruit over the right side of his chest and neck. He denied a history of trauma, catheterization, congestive heart failure, or shortness of breath. He subsequently underwent computed tomography angiography, which demonstrated an arteriovenous fistula or malformation involving the right vertebral artery, subclavian artery,

and internal jugular vein (Fig 1). He was then referred to our institution for evaluation and management.

Arteriography was performed through a right brachial artery approach. Selective arteriography showed primary filling from the right vertebral artery with additional filling from the right subclavian artery (Fig 2, A). The vertebral artery was markedly dilated proximal to the fistula with a diameter of 8 mm, whereas distal to the fistula, its diameter was 3 mm. The vertebral connection with the arteriovenous malformation was first occluded with a 16-mm Amplatzer plug (Abbott-St. Jude Medical, Minneapolis, Minn) placed within the vertebral artery. It was thought that a covered stent constrained to 3 mm in diameter in the distal portion of the vertebral artery would not have remained patent. Subsequently, the connections with the subclavian artery were occluded using a 13-mm-diameter by 5-cm-long covered Viabahn stent (W. L. Gore & Associates, Flagstaff, Ariz). Completion angiography demonstrated no further arterial filling of the arteriovenous malformation (Fig 2, B). The patient was receiving therapeutic unfractionated heparin anticoagulation during the procedure and was treated with 6 weeks of low-dose aspirin and 75 mg of clopidogrel after the procedure. It was thought that embolic protection was not required because there was no thrombus present on computed tomography angiography, and the process was not atherosclerotic.

The patient has subsequently had 3-week and 6-month postoperative follow-up with computed tomography arteriography. He no longer has a bruit over the right side of his neck. Sequential computed tomography scans, which preoperatively showed prompt filling of the jugular vein during the arterial phase of the scan (Fig 3, left), demonstrated no filling from the fistulas after occlusion of the connections between the vertebral artery and jugular vein and between the subclavian artery and jugular vein (Fig 3, right).

## DISCUSSION

The primordial vascular network is formed from blood islands that later divide into the arteries and veins.<sup>1</sup> The proximal portion of the subclavian artery morphologically originates from the proximal vertebral artery, which

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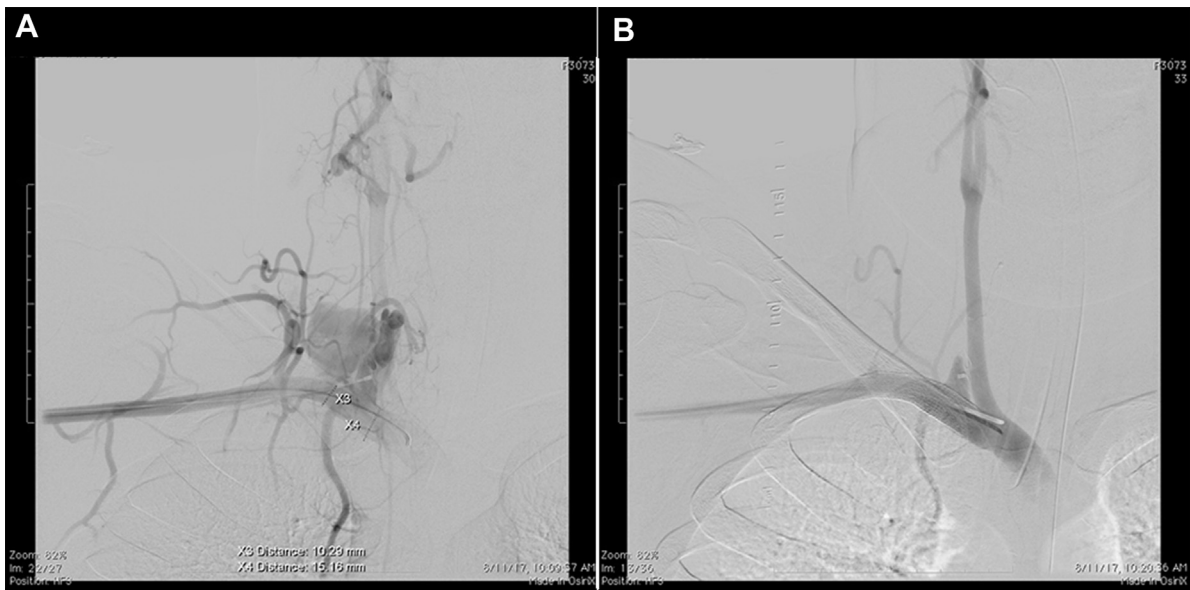
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**Fig 1.** The computed tomography angiography image shows the arteriovenous malformation between the vertebral artery and the jugular vein marked before treatment by the radiologist.



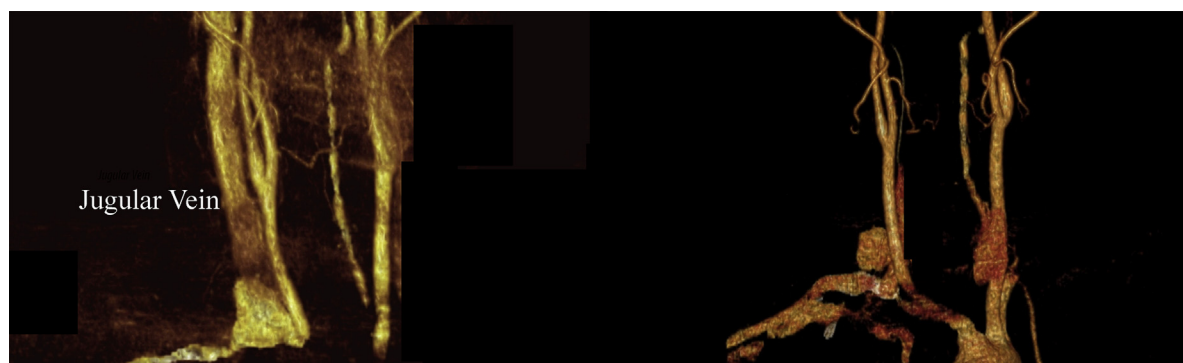
**Fig 2. A,** The arteriogram before treatment demonstrates the arteriovenous malformation with its connections to the right vertebral artery and right subclavian artery. **B,** The completion arteriogram demonstrates no residual filling of the venous structures after Amplatzer plug occlusion of its connection with the right vertebral artery and coverage of the subclavian artery connections with a Viabahn covered stent.

develops from the seventh dorsal intersegmental artery.<sup>2</sup> Thus, a congenital arteriovenous abnormality may be expected to involve both of these arteries simultaneously, as it did in this case study. The internal jugular veins are formed from the anterior cardinal veins in this region.<sup>3</sup> Most arteriovenous connections in the neck are caused by trauma, but there have been a limited number of reports of spontaneous or congenital interconnections in this region.<sup>4</sup>

Treatment of arteriovenous malformations of the neck may be complex because of the many nerves and vascular structures in this region.<sup>1</sup> Repairs may require either open or endovascular interventions and may need to be staged to reduce the risks of bleeding and

nerve injury. Ligation or occlusion of feeding arteries may not have long-term efficacy because of the complex nature of the interconnections between the arteries and veins.<sup>5</sup> However, the recurrence rate is high for these malformations even with surgical excision,<sup>6</sup> and redo intervention for recurrence is difficult. Some authors advocate a combination of endovascular and open procedures.<sup>3,7</sup>

In this particular case study, the arteriovenous malformation did not become prominent until adulthood. It involved the subclavian artery, vertebral artery, and internal jugular vein. The arterial abnormalities appeared to arise from structures formed by the seventh dorsal segmental artery and were able to be treated with



**Fig 3.** Three-dimensional computed tomography angiography reconstructions show prompt filling of the jugular vein before occlusion of the arteriovenous fistulas (*left*) and no filling from the fistulas after occlusion of the connections between the vertebral artery and jugular vein and between the subclavian artery and jugular vein (*right*).

combined coil embolization of the right vertebral artery as well as stent graft coverage of the involved right subclavian artery. The venous structures have had a sustained reduction in size without excision of the right jugular vein. Further treatment of any other arterial connections as well as the option of resection of a smaller vascular mass if evidence of malformation growth occurs during follow-up could still be pursued.

In this patient, we were able to embolize the right vertebral artery with minimal long-term adverse risks because the left vertebral artery was adequately sized and the patient had an intact basilar artery connecting both vertebral arteries as well as an intact circle of Willis. This would not be an option if the patient had a dominant right vertebral artery or abnormal collateralization of the basilar artery or posterior communicating arteries.

An alternative treatment would have included an open repair. However, it would have had increased risks of bleeding. The short fistulas between the vertebral artery and jugular vein and the subclavian artery and jugular vein would have been difficult to embolize directly without compromising the adjacent arteries.

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