MR Angiographic Demonstration of Bilateral Duplication of the Extracranial Vertebral Artery: Unusual Course and Review of the Literature

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Duplication and fenestration of the vertebral artery are considered developmental anomalies. In many case reports, the terms fenestration and duplication have been used incorrectly and interchangeably. Lasjaunias et al, however, maintain that there are 2 types of fenestration: an arterial split and true duplications, thereby using fenestration as a generic description of any situation in which there is double segment to the vertebral artery. In another report, Lasjaunias et al state that “the main difference between duplications and fenestrations is that in duplication one of the vessels leaves the vertebral canal to enter the spinal canal and has a subarachnoid course which starts and ends at the level of two consecutive foramina transversaria and follows a cervical root, whereas fenestration remains in the vertebral canal between two consecutive transverse processes.” This may not necessarily be true, though this new definition clearly separates the 2 entities. It is suggested that these definitions were based on angiographic data that may not be as accurate as such cross-sectional techniques as MR imaging for defining anatomic relationships. Duplication should be strictly applied to a vertebral artery that has 2 origins and a variable course and fusion level in the neck. With duplication, both segments are found outside of the spinal canal. In contrast, fenestration represents a vessel with a single origin and anywhere along its course the main trunk divides into 2 parallel segments that may lie within or outside of the vertebral canal, depending on the level of occurrence.

Duplications of the extracranial course of the vertebral artery are rarely reported in the literature and are seen as incidental findings in autopsy series, in angiographic studies, or, recently, in MR angiography (MRA) studies. We report what we believe to be the first case of bilateral duplications of the vertebral artery at the neck level as an incidental finding during the MRA examination of a patient with cognitive decline.

Case Report
An 83-year-old white man was referred to us for cranial MR imaging with MRA of the head and neck because of mild cognitive impairment. There was no other significant clinical abnormality or significant past medical history. His cranial MR images obtained on a 1.5T system showed no significant focal abnormality other than the incidental finding of a small right middle cranial fossa arachnoid cyst and volume loss. 3D time-of-flight (TOF) cranial MRA obtained at the same time was also normal. The 2D TOF neck MRA and 3D acquisition during intravenous gadolinium enhancement revealed bilateral proximal duplications of the vertebral arteries (Fig 1). A left proximal internal carotid artery severely ulcerated stenosis was also present.

On the right side, there were 2 separate origins of the vertebral artery from the right subclavian artery; one origin emanated proximally beyond the origin of the subclavian artery. This segment made a short loop and entered the carotid space, staying in a medial relationship to the right common carotid artery (Fig 2). The second origin emanated from the subclavian artery adjacent to the origin of the right internal mammary artery, coursing straight, and entered the foramen transversarium at C7 vertebral level. This posterior vessel was slightly larger than the anterior segment. Both vessels joined in the foramen transversarium at C4–C5 disk level to continue as the right vertebral artery.

On the left side, there were 2 separate origins of the vertebral artery from the left subclavian artery. One segment emanated adjacent to the origin of the internal mammary artery, looped slightly forward, and coursed straight up behind the left common carotid artery (Fig 2). The other segment emanated from the subclavian artery, just distal to the proximal and slightly larger in caliber, and entered the foramen transversarium at C7. Both vessels joined in the foramen transversarium at C5–C6 disk level to continue as the left vertebral artery.

Discussion
To the best of our knowledge, only 2 reports have described symmetrical extracranial fenestration of the vertebral artery: one at the extracranial and another at the intracranial level and describing double fenestration of the extracranial vertebral artery at the same level. The authors incorrectly described these fenestrations as duplication. Hasegawa et al reported that they collected a total of 74 cases of vertebral artery duplication from the literature and concluded that, in all cases, the
duplication was sited at either the atlantoaxial level or the intracranial segment of the artery. These cases were clearly not duplication according to the definition in our introductory passage, but rather fenestration, because the vessels did not have double origins but split somewhere along their lengths. These attest to the confusion in the literature with respect to the true definition of duplication. These changes have been reported more frequently by Japanese authors than by their American and European counterparts. Fenestration was located more commonly on the left side at the level of the atlantoaxial junction and only bilaterally in 4 cases. There are 21 cases of true vertebral artery duplication reported, most of them published in the Japanese literature. There appears to be a distinct difference between the histologic makeup of a fenestrated vertebral artery and the normal vessel. A histologic examination of a fenestrated vessel revealed variable composition of the wall. Findings include a muscular wall that is less developed with irregular pattern of the elastic fibers in one segment and a complete absence of elastic fibers in the other segment. A common adventitia surrounded both segments. There is no histologic correlate of true duplication, but one would expect that it should show normal configuration.

To the best of our knowledge, bilateral symmetrical extracranial duplication of the vertebral artery has not been previously reported. Our case is the first report describing a bilateral extracranial duplication of the vertebral artery with a double origin from each corresponding subclavian artery and a separate course in the neck—one duplicated segment coursing along the carotid sheath and the other segment through the foramen transversarium (Fig 2) and a different level of union of the duplicated vessels in the neck. Union of the duplicated segments has been noted to be at C4–C6 levels, as in our case. In our case, the proximal duplicate limb was the smaller of the 2 limbs bilaterally and both appeared to ascend within the carotid space or very close to the common carotid artery. The distal limb was the larger of the duplicated vessels bilaterally, and each directly entered the foramen transversarium at C7. These findings were incidentally demonstrated by MRA in a patient evaluated for cognitive deficits. MRA of the neck offers the opportunity to visualize multiple vessels at the same time, thus making it possible to discover anomalies that would not have been otherwise suspected on conventional angiography. The true incidence of duplication of the vertebral artery is unknown.

The embryogenesis of the vertebral artery begins at approximately 32 days and is completed by 40 days, between the 12.5- and 16-mm stages. The vertebral artery is formed from fusion of the longitudinal anastomoses that link cervical intersegmental arteries, which branch off the primitive paired dorsal aorta. The intersegmental arteries eventually regress, except for the seventh vessel, which forms the proximal portion of the subclavian artery, including the point of origin of the vertebral artery. As their connections to the primitive dorsal aorta disappear, the vertebral artery is formed and takes on the appearance of a beaded anastomotic chain with a tortuous course. The basilar artery is formed by the fusion of the 2 primitive vertebral arteries. Sim et al\(^1\) state that a portion of the primitive dorsal aorta may not regress along with 2 intersegmental arteries that connect to the vertebral artery. It is believed that this arrangement may give rise to vertebral artery duplication or double origin to that vessel. A failure in the controlled regression process of the intersegmental arteries themselves can result in vertebral artery fenestration. Bilateral occurrence of these failures gives rise to bilateral duplication or fenestration of the vertebral artery as the case may be. Our unique case of bilateral symmetrical duplication of the vertebral artery may be explained by persistence of a portion of the primitive dorsal aorta segment on both sides along with lack of regression of the fifth and sixth intersegmental arteries (Fig 3).

Some authors stated that vertebral artery duplications or fenestration were incidental findings with no significant pathologic and clinical consequences. The abnormal histologic makeup of a fenestrated vessel may predispose such vessel to significant pathologic consequences. A duplicated vertebral artery, however, may not be as vulnerable. A possible
A pathoetiologic association was suspected in a case of dissection in a duplicated right vertebral artery. A kinked origin of a duplicate vertebral artery was operatively corrected for suspicion of being responsible for dizziness in a 70-year-old woman. Her symptoms improved postoperatively. There has also been some association of symptoms and other pathologies with fenestrations of the vertebral artery, which were erroneously reported as duplications. In the present case, the duplication of both vertebral arteries is an incidental finding and there is no other associated vascular anomaly of the extracranial or intracranial circulation as demonstrated by MRA. It is conceivable that duplication could afford a protective mechanism for stenosis or injury to or occlusion of one of the duplicated limbs. Such anomalies may become important in the planning of interventional procedures.

We believe that the presence of these developmental anomalies in the blood supply to the brain can be noninvasively evaluated by MRA. Further studies can assess the real incidence of these conditions. The presence of duplication or fenestration of the vertebrobasilar or carotid system is important; it may or may not be associated with specific symptoms or other pathologies and may influence the choice or route of endovascular treatment of central nervous system disorders. This should be kept in mind in the evaluation of neck and intracranial disorders.

References