Right Vertebral Artery Originating from the Aorta Distal to Left Subclavian Artery and Ending in Posterior Inferior Cerebellar Artery in a Patient with Moya-Moya Disease

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Introduction

Moya-moya disease is a chronic cerebral vasculopathy characterized by slowly progressive carotid artery narrowing and occlusion at the base of the brain with the formation of an extensive network of anastomoses and the characteristic angiographic appearance of the "puff of smoke". It can have acquired as well as congenital causes and various intracranial and systemic vascular anomalies are known to be associated with it. However, to our knowledge, an aberrant origin of the right vertebral artery distal to the left subclavian artery, which in itself is a rare anomaly with only 17 cases being reported to date [1–13], has never been described in association with moya-moya disease.

Case Report

A 19-year-old girl presented with sudden onset of severe occipital region headache followed by altered sensorium and repeated episodes of vomiting.

A plain CT scan of brain showed intraventricular hemorrhage (more in the left lateral ventricle). A cerebral DSA (digital subtraction angiography) was done which showed a typical pattern of moya-moya disease (Figure 1) with occlusion of both internal carotid arteries in the supraclinoid segment along with the "puff of smoke" appearance produced by the proliferating lenticulostriate arteries, formation of leptomeningeal collaterals from the posterior circulation with minor contribution to the intracranial flow from the external carotid arteries as well. The right vertebral artery originated from the aorta distal to the left subclavian artery origin (Figure 2) and ended in the posterior inferior cerebellar artery (Figure 3). An aneurysm was present distally on the left lateral posterior choroidal artery (Figure 1).

The patient was managed medically and has done well to date.

Discussion

The progressive occlusion of intracranial vessels [14] with the consequent proliferation of collateral circulation typical of moya-moya, has been accepted by some to be acquired [15, 16]; yet others hypothesize a developmental etiology [17] considering the clustering of the disease according to race and families and its presence in syndromes like Noonan [18], Turner [19], Down [20], cardiofaciocutaneous [21], and neurofibromatosis [22]. This is especially true for the childhood-onset variety, which is common, and different from the adult form [23]. A familial moya-moya disease has been mapped to chromosome 3p24.2-p26 [24].

Extracellular matrix metabolism is altered in these patients leading to elastin accumulation in arterial smooth muscle cells [25]. Ikeda used histopathologic examination and morphometric analysis to demonstrate eccentric intimal thickening with medial thinning and

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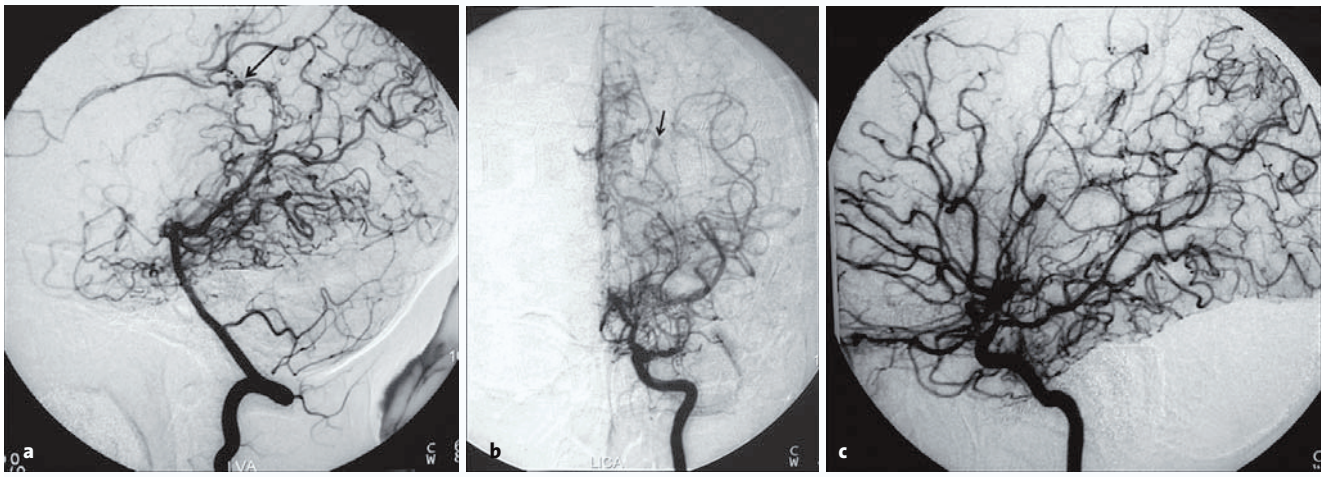


Figures 1a to 1c. Left vertebral artery injection, lateral view (a), left internal cerebral artery injection, Towne's view (b), and right internal cerebral artery injection, lateral view (c), showing the moya-moya phenomenon. The arrow points to an aneurysm on the left lateral posterior choroidal artery.

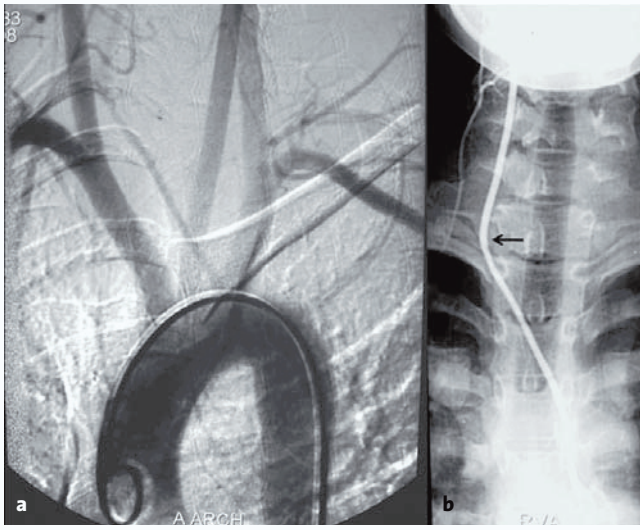
fibrosis in the systemic and pulmonary vessels [26]. Jansen et al. have demonstrated cases with noninflammatory fibrodysplasia in the extracerebral arteries [27]. Thus now this disease is considered more of a systemic vasculopathy with prominent cerebral manifestations.

Accordingly, multitudes of vascular anomalies – of both intra- and extracranial head and neck vasculature – are associated with it [28–38]. These include vessel agenesis [28], persistent primitive arteries [29], anomalous origins [30], fenestrations [31], vessel ectasia [32], aneurysms [33], and vascular malformations [34].

Equally important but less researched is the concurrent involvement of the extracranial vasculature. Various rare anomalies of the heart, aorta and large vessels [35, 36], as well as renal artery stenosis and aneurysms have also been described [27, 37].

Among all the branches of the subclavian artery, the vertebral artery is considered to have the most constant origin. The commonest variation in the origin of the vertebral artery is that of the left vertebral artery directly arising from the aortic arch, which is seen in 3% of the population [38]. Origin of right vertebral artery from aorta distal to that of the left subclavian artery is very rare. Only 17 cases have been reported to date [1–13], including cases wherein both vertebral arteries arose from the aorta.

Development of vessels is influenced by rheologic and tension mechanics. Tension is created on the developing vessels due to the continuous process of resorption and formation of the primitive vessels. Blood flows into the channels which offer least resistance. Thus the channels with least tension and offering least resistance to flow persist while others perish [39]. The vertebral arteries develop from longitudinal anastomosis of dorsal intersegmental arteries in the cervical region. Due to the caudal shifting of the aorta during embryogenesis, the proximal parts of the intersegmental arteries are exposed to longitudinal tension and bending with a resulting retarded blood flow. This may result in abnormal connections between the developing vertebral artery and the adjacent large arteries offering less resistance [38].



Figures 2a and 2b. Arch aortogram showing origin of the right vertebral artery distal to left subclavian artery (a), and right vertebral artery injection, unsubtracted image (b), showing the entry of the vertebral artery into the foramen transversarium of the 7th cervical vertebra (arrow).

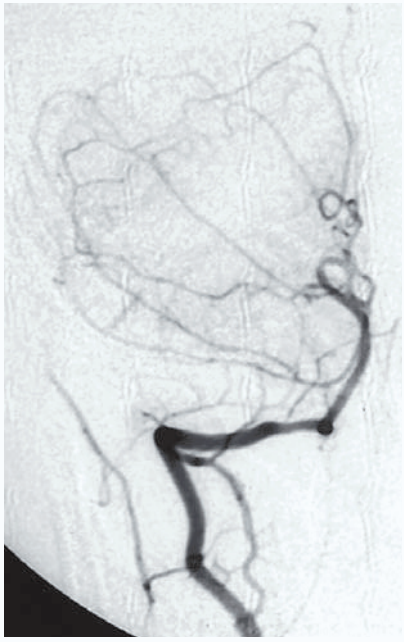


Figure 3. Right vertebral artery injection, Towne's view, showing the artery ending in the posterior inferior cerebellar artery.

In our case, the mechanics described above would translate anatomically as origin of the right vertebral artery from the 8th dorsal intersegmental artery instead of the 7th, along with physiological closure of the right dorsal aorta distal to the 7th intersegmental artery. That it arose from the 8th intersegmental artery can be deduced from the fact that it entered the foramen transversarium of the 7th cervical vertebra.

Two anomalies which are known to be associated with moya-moya disease are aortic coarctation and aberrant origin of the right subclavian artery, and they can be explained by mechanisms which are similar to the one described above. Aortic coarctation occurs when there is partial involution of the left dorsal aortic arch while the aberrant right subclavian artery, an otherwise relatively common anomaly seen in 0.1% of the population, occurs when the right 4th aortic arch and right dorsal aorta involute cranial to the 7th intersegmental artery [40].

Other anatomic alterations have been found associated with this anomalously originating right vertebral artery. Albayram et al. found prominent filling of the posterior circulation via the posterior communicating arteries [7]. A theoretical possibility of altered hemodynamics with resultant aneurysm formation is also plausible.

Overall, vertebral artery ending in the posterior inferior cerebellar artery is a common anomaly, however, not seen in any of the previously reported cases. Conversely, in the case by Satti et al. [11], this anomalous vertebral artery was the sole supply of the posterior fossa arteries, the left one not showing any contrast filling.

The occurrence of this anomaly in our patient with moya-moya disease might have been coincidental; however, an embryopathologic connection of this constellation cannot be ruled out entirely at this stage, the instant case being the first such example in addition to various other anomalies of the major vessels being reported in earlier literature.

Apart from these fascinating embryologic and pathologic relationships between the extra- and intracranial vascular anomalies, the clinical significance cannot be overemphasized, whether it is from the point of view of the cardiothoracic surgeon, the neurosurgeon, the radiologist, trauma specialists or even residents and nurses [41–43].

Conclusion

We report the first case of anomalous origin of right vertebral artery distal to the left subclavian artery origin in a patient with moya-moya disease. A plausible association between the two can be made in view of the other reported anomalies of this region with a similar anatomic, physiological and embryologic pathway. Vertebral arteries frequently terminate at the posterior inferior cerebellar artery, but this has never reported in the previous 17 cases. The present case is the first one where the anomalously originating right vertebral artery ended at the posterior inferior cerebellar artery.

Conflict of Interest Statement

We certify that there is no actual or potential conflict of interest in relation to this article.

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