

Near complete tracheal rings in an adult male cadaver associated with variation in V2 and V3 segments of vertebral artery

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SUMMARY

During routine dissection in an adult male cadaver, the presence of long segment near-complete tracheal rings associated with variation in the second (V2) of right vertebral artery (VA) and third (V3) segments of the left VA was encountered. This case report highlights the normal lifespan of an individual despite anatomical variations that are reported to be incompatible with life.

Congenital complete and near-complete tracheal rings are extremely rare tracheal deformities, and long-segment occurrences of these variations are usually incompatible with life. They are reported to be associated with cervical chondrogenic anomalies and craniosynostotic syndromes.

In the same cadaver the left VA entered the foramen transversarium (FT) of C6 vertebra and exited through the FT of the axis. It formed a loop below the arch of atlas and entered the vertebral canal between the atlas and axis, completely bypassing the FT of the atlas. A branch from the loop divided into two branches, one of which supplied the dorsal *ramus* of C1, the other ending in the surround-

ing neck musculature. The right VA was normal, except that it entered the FT of C5 vertebra bypassing C6 vertebra. It was noted that the calibre of the VA on the left side was considerably more than that on the right. Variations in the vertebrobasilar system have a potential clinical impact, since they are the feed arteries of the brain.

Key words: Near complete tracheal rings – Vertebral artery – Foramen transversarium – Atlas vertebra – Tortuosity

INTRODUCTION

Anatomical variations have been defined as the normal flexibility in the topography and morphology of body structures (Shoja et al., 2006). Advances in imaging and surgical techniques have provided an accurate knowledge of anatomical variations an absolute prerequisite. This case highlights the presence of long-segment near-complete tracheal rings in an adult male cadaver associated with variations in the second (V2) segment of the right VA and the third (V3) segment of the left VA,

called by some authors the C2 segmental type of VA (Tokuda et al., 1985; Sato et al., 1994).

Congenital complete and near-complete tracheal rings are extremely rare tracheal deformities. Long-segment occurrences of these anomalies are usually incompatible with life. They are reported to be seen in patients with cervical chondrogenic anomalies and craniosynostotic syndromes (Somers and Suskind, 2008; Faust et al., 1998).

The vertebral artery is the first branch of the subclavian artery. The course of the artery from its origin to the point where it joins the opposite VA to form the basilar artery is divided into 4 segments: V1, V2, V3, and V4. Segments V1 to V3 represent the extracranial course of the VA. The V1, or pre-transverse segment, usually extends from its origin from the subclavian artery to its entry into the foramen transversarium (FT) of the C6 vertebra. The V2 segment extends from the transverse process of the C6 vertebra to where the VA exits the atlas. The V3 segment extends from the point of exit from the atlas to its entry into the spinal canal, which then continues intracranially as the V4 segment. The two vertebral arteries then fuse to form the basilar artery (Standring, 2008; Adnan, 2011). The variations in the vertebrobasilar system have a potential clinical impact, since they are the feed arteries of the brain.

CASE REPORT

During the dissection of an adult Indian male cadaver, the cervical segment of the trachea was resected, and sent for endoscopic measurement of its inner diameter as part of a study conducted at St. Johns Medical College and Hospital, Bangalore, South India. It was noted that the inner diameter of the trachea was unusually small and circular, with near complete tracheal rings. This finding was present in all the tracheal rings and was confirmed by endoscopy (Fig. 1). The tracheal rings were dissected at the posterior tracheal pars membrane, which was less than one millimetre wide.

In the same cadaver, the VAs had an abnormal course. The right VA originated from the right subclavian artery, passed through the scalenovertebral trigone, and then entered the FT of the C5 vertebra, bypassing the FT of the

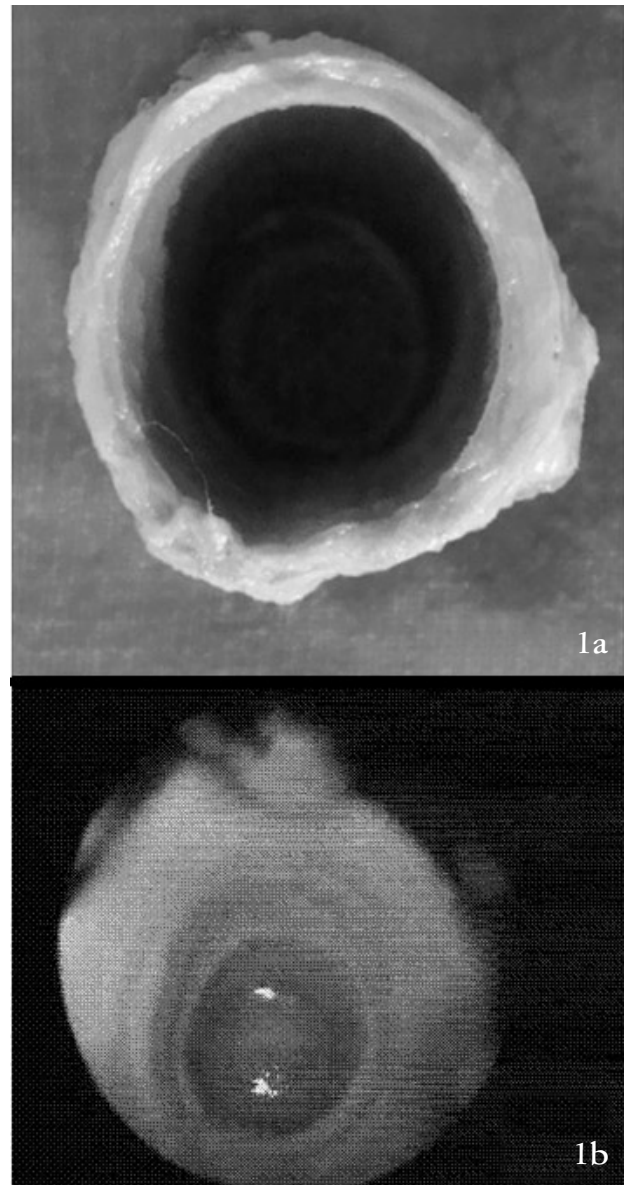


Fig. 1. Near-complete tracheal rings (Axial section of the trachea - superior view). 1a: Gross view. 1b: Endoscopic view.

C6 vertebra. The further course of the right VA was normal.

The left VA originated from the left subclavian artery, passed through the scalenovertebral trigone, then entered the FT of C6 vertebra, and emerged out of the FT of C2 (axis) vertebra. The artery then formed a tight lateral loop (Fig. 2). It then entered the vertebral canal by piercing the *dura mater* below the atlas vertebra, bypassing the FT of the atlas vertebra. The C2 spinal nerve emerged superomedially to the left VA, and was noticed to course just above, where it gave off the ventral and dorsal *rami*. The ventral *ramus* ran anteriorly in close approximation to the loop formed by the VA. A branch from the loop of the left VA divided into two branches one of which supplied the dorsal *ramus* of C1, the other end-

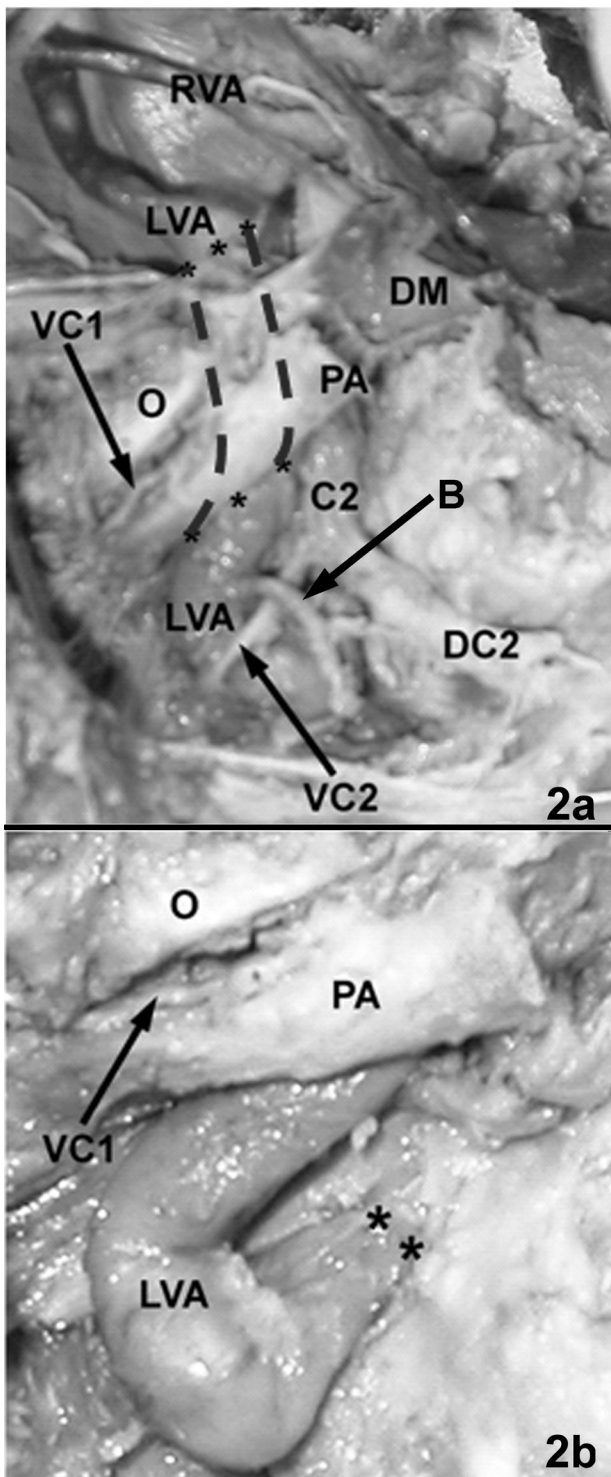


Fig. 2. Vertebral artery anomaly. 2a: Posterolateral view of the left vertebral artery (LVA), bypassing the foramen transversarium of the atlas vertebra. The red dotted lines indicate the course of the LVA anterior to the posterior arch of the atlas (PA). The asterisks indicate the level of entry in front of the PA and entry into the foramen magnum. B: branch from LVA; C2: trunk of 2nd cervical spinal nerve; DC2: dorsal *ramus* of C2; DM: spinal *dura mater*; O: occipital bone; RVA: right vertebral artery; VC1: ventral *ramus* of 1st cervical spinal nerve; VC2: ventral *ramus* of C2. 2b: Enlarged view of the left vertebral artery (LVA) after the removal of overlying structures. Asterisks indicate the exit of the LVA from the foramen transversarium of the axis vertebra. O: occipital bone; PA: posterior arch of atlas; VC1: ventral *ramus* of 1st cervical spinal nerve.

ing in the surrounding deep suboccipital neck musculature. The cadaver was further dissected to look for any associated variations. An enlarged heart was observed, which could be due to prolonged cardiac stress secondary to congenitally narrow tracheal lumen. Unfortunately the medical history of the subject was unavailable.

DISCUSSION

Patients with complete tracheal rings usually present symptoms in the first year of life. The most common clinical presentation is long-standing shortness of breath and chest tightness (Boiselle, 2007). Stridor, sternal, and subcostal retractions and cyanosis may be present at birth. A survey of the literature reveals that congenital complete and near-complete tracheal rings are extremely rare tracheal deformities (Somers and Suskind, 2008). Long-segment occurrences of these anomalies are usually incompatible with life (Faust et al., 1998), but cases have been reported presenting asthma-like symptoms in adulthood (Nagappan, 2002; Reed et al., 2011). They are also associated with cervical chondrogenic anomalies, craniosynostotic syndromes such as Crouzon's syndrome, foreshortened neck, pulmonary agenesis, abnormal vasculature as seen in this case study, and Down's syndrome (Bravo et al., 2006). Down's Syndrome has also been associated with VA variations (Yamazaki et al., 2008).

The ring-sling complex is an uncommon condition in which a pulmonary artery slings around complete tracheal rings, presenting with severe upper airway obstruction due to tracheal and bronchial compression. It usually appears as an isolated abnormality, but can be associated with other congenital cardiac defects, including Tetralogy of Fallot (Calcagni et al., 2008).

Complete or near complete tracheal rings arise from disproportionate growth of cartilage relative to the posterior tracheal membranous part (Chen and Holinger, 1994). The incidence of these tracheal rings in elder children or adults has not been determined (Volland et al., 1986).

Treatment depends on the severity of the symptoms. Short stenosis may be treated with resection and end-to-end anastomosis. For lengthier stenosis, pericardial patch repair and

slide tracheoplasty are used (Aneeshkumar et al., 2005).

High resolution CT or MRI can be useful for diagnosis of vascular variations like the pulmonary sling, and diagnosis of tracheo-bronchial tree anomalies (Hodina et al., 2001; Manson et al., 1994). Not all patients with complete tracheal rings require tracheoplasty. Some may have satisfactory airway growth, do not require airway reconstruction, and may be completely asymptomatic (Rutter et al., 2004; Shiga et al., 1999; Yokomura et al., 2005). The authors feel that the presence of complete or near-complete tracheal rings should lead to an active search for vascular anomalies, especially the ring sling association (Dunham, 1994; Faust et al., 1998).

The right VA was found to enter the FT of the C5 vertebra. The incidence of this was variable as found by different workers – varying between 4.5% and 7% (Adachi, 1928; Rieger and Huber, 1983; Bruneau, 2006; Bergman, 1988). The left vertebral artery was found to bypass the FT of the atlas, but passed through the FT of C6 through C2. This has been called C2 segmental type of the vertebral artery (Tokuda et al., 1985; Sato et al., 1994). The explanation for the variation described above can be found by delving into the embryological origins of the VA. During development, each somite receives its blood supply from the corresponding intersegmental artery. Each of these arteries gives off a spinal artery in the dorsal part. In the cervical region, the intersegmental arteries are interconnected by the postcostal anastomotic arteries. The V1 segment of the VA is formed from the dorsal division of the 7th intersegmental artery, V2 segment from the postcostal anastomoses, V3 segment from the spinal branch of the first cervical intersegmental artery, and V4 from the preneuronal division of the spinal branch,

which joins with the corresponding branch of opposite side to form the basilar artery. In the present case it appears as if the postcostal anastomosis between the first and second cervical intersegmental arteries, as well as the spinal branch of the 1st cervical intersegmental artery, had regressed. The spinal artery of the 2nd cervical intersegmental artery has contributed to the formation of variant V3 segment of the VA. Hence the artery enters the foramen magnum directly, bypassing the FT of the atlas vertebra (Fig. 3) (Dutta, 2005).

Studies have been reported in literature where this variant course of the VA has been discovered either during autopsy, routine angiography, or recently with the help of 3D-CT angiography. In cases of occipitalisation of

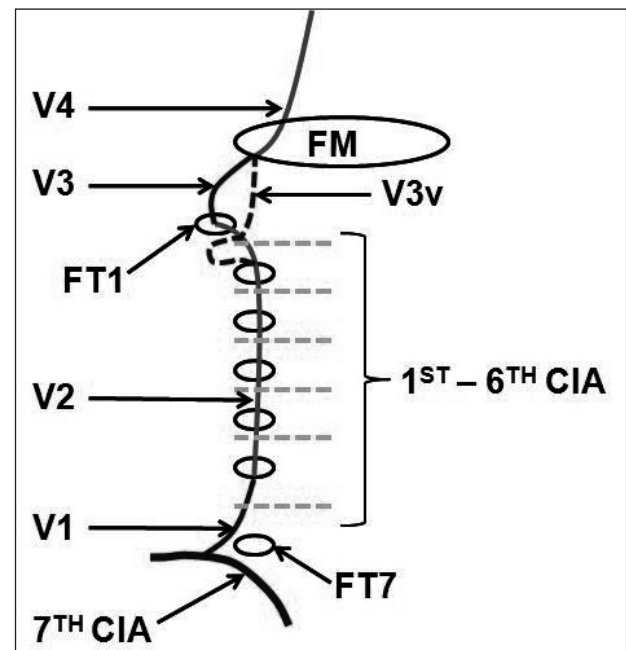


Fig. 3. Schematic diagram showing the development of different parts of the vertebral artery. 1st - 6th CIA: first to 6th cervical intersegmental arteries; 7th CIA: seventh cervical intersegmental artery; FM: foramen magnum; FT1 and FT7: foramen transversaria of 1st and 7th cervical vertebrae; V1-V4: four parts of the vertebral artery; V3v: variable course of vertebral artery in present case.

Table 1. Frequency of variation of the vertebral artery where it bypasses the foramen transversarium of the atlas in various studies.

Sl.no.	Author & Year	Country	Methodology	Frequency/ No. of cases studied (percentage)
1.	Tokuda et al. (1985)	Japan	Angiography	5/300 (1.67%)
2.	Sato et al. (1994)	Japan	CTA/MRI	10/1436 (0.69%)
3.	Hong et al. (2008)	Korea	3D-CTA	48/1013 (4.7%)
4.	Yamaguchi et al. (2008)	Japan	3D-CTA	(1.8%)
5.	Duan et al. (2009)	China	3D-CTA	4/68 (5.8%)
6.	Jian et al. (2003)	China	Anatomical	Case report
7.	Yi Zhi-Quiang et al. (2009)	China	CTA	Case report

the atlas vertebra, the artery has been reported to pass below the C1 vertebra in 33.3% of the cases (Wang et al., 2009). The frequency with which this variant has been reported varies from less than 1% up to 6%. Additionally, a few case reports have been published concerning this variant course. The results of these studies have been summarized in Table 1.

The VA is the major artery acting as the feed artery for the circle of Willis that supplies most of irrigation of the brain. A variation in the course may lead to altered hemodynamics that may lead to cerebral disorders by predisposing the patient to aneurysmal formations, greater risk of thrombosis, occlusions, arterial dissections, and potential atherosclerosis with the associated morbidity and mortality (Thevenet et al., 1984; Jackson et al., 2000; Muller et al., 2008).

It has been reported that an abnormal loop of the VA caused compression of the accessory cranial nerve and C2 root, leading to occipital neuralgia (Vincentelli et al., 1991; Sharma et al., 1993). The literature has reported that this kind of VA anomaly may be associated with Klippel-Feil syndrome (Tokuda et al., 1985; Sharma et al., 1993). The tortuosity of the variant artery may be responsible for nerve root compression and bony erosion leading to fractures (Fisher et al., 1965; Matula et al., 1997).

As conclusion, this case signifies that, despite possessing these anatomical variations that are reported to be incompatible with life (the presence of long segment near-complete tracheal rings), a normal lifespan is possible for certain individuals. However, when suspecting these kinds of associated abnormalities, an extensive evaluation of an individual to detect any vascular abnormalities is recommended, as it could contribute to the individual's prognosis with the existing variation.

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