

C3 segmental vertebral artery and its surgical implication in craniovertebral junction anomalies: Insights from two cases

ABSTRACT

A spectrum of vertebral artery (VA) anomalies have been described with or without an associated congenital craniovertebral junction (CVJ) anomalies. C3 segmental VA, where the VA enters the dura at the level of C2/3 intervertebral foramen is an extremely rare anomaly. We report two cases of congenital CVJ anomaly (irreducible in one with C2/3 fusion and reducible in the other; without any subaxial fusion but with articular agenesis at C2/3 joint on the anomalous artery side). Computed tomographic angiography revealed intraspinal intradural entry of VA through the C2/3 intervertebral foramen on the right side with the contralateral artery found crossing the atlanto-axial joint. Both the patients underwent posterior approach and C2 was spared from instrumentation in both cases. Postoperatively, the patient with irreducible dislocation recovered well while the patient with reducible dislocation expired, possibly secondary to the thrombosis of the dominant VA from C2/3 foraminal encroachment. C3 segmental VA may be advantageous in aggressively exposing the C1/2 joint but instrumentation of C2 or C3 needs caution in view of the possibility of VA injury. Our experience shows that VA may be endangered even while exposing and protecting the artery. For such cases, we recommend posterior decompression of the C2/3 neural foramen during instrumentation in the absence of associated C2/3 fusion, as an abnormal joint morphology of C2/3 indicates a C2/3 instability.

Keywords: C3 segmental artery, craniovertebral junction, injury, lateral spinal artery, posterior fusion

INTRODUCTION

Craniovertebral junction (CVJ) anomalies represent a complex set of disorders. More often than not, the bony anomalies are shadowed by anomalies involving the V3 segment of the vertebral artery (VA).^[1,2] These anomalies have important surgical implications, often putting the artery under substantial risks during routine surgical procedures.^[1] C3 segmental VA, where the artery enters the dura at the level of C2/3 intervertebral foramen is extremely rare, having been reported only in four occasions before [Figure 1].^[3-5] However, none of the previously reported patients had associated CVJ anomaly. Therefore, no information is available on the spectrum of associated CVJ anomalies with C3 segmental VA and their surgical implications. Here, we present the clinico-radio-surgical finding of two such cases operated recently and discuss the learning points.

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CASE REPORTS

Case 1

A 31-year-old female presented with complaints of gradually progressive spastic quadriplegia. On clinical examination, there was hypertonia affecting all four limbs with a muscle power of 3/5 (medical research council) in both upper limbs and 4/5 in both the lower limbs. Bilateral hand grip was weak and graded sensory deficit was present in all four limbs. Plantar reflexes were bilaterally extensors and the deep-tendon reflexes were exaggerated.

On magnetic resonance imaging (MRI) of the CVJ, there was severe cord compression without any associated syrinx. Axial MRI sections at the same level revealed an intradural, lateral location of the right VA [Figure 2a and b]. On computed tomographic (CT) study, there was C2/3 congenital fusion and rotatory atlanto-axial dislocation (AAD) with right-sided neck tilt [Figure 2c and d]. CT angiography (CTA) revealed bilaterally symmetrical VAs, with the anomalous course. The right VA was entering the spinal canal through the C2/3 intervertebral foramen and then continued cranially. On the left side though, the artery entered between occipitalized C1 vertebra and the C2 vertebra (C2 segmental VA). The basilar artery (BA), bilateral posterior inferior cerebellar artery, and posterior cerebral arteries were normal [Figure 2e and f].

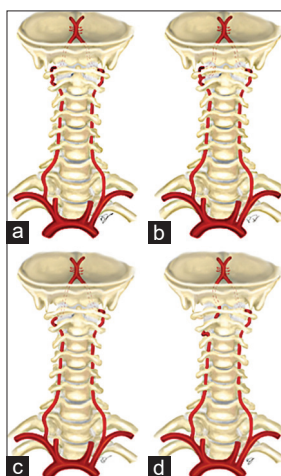


Figure 1: Artistic illustration of the normal course of the V3 segment of the vertebral artery, its common and rare variations. The commonest course is when the artery runs on the upper surface of the posterior arch of atlas (a) after having negotiated through the foramen transversarium of C1. The other common variations include a fenestrated vertebral artery where the vertebral artery has a duplication and one part has a normal course while the other part courses below the C1, crossing the C1/2 joint posteriorly (b) or when the artery is not fenestrated but courses over the C1/2 joint (c), below the posterior arch of C1 (also called a persistent intersegmental artery or C2 segmental artery). The rarest variation, which we present here, is when the vertebral artery takes a medial turn below the C2 pars interarticularis, enters the dura through the C2/3 neural foramen (d). This variation is also known as the persistent second intersegmental artery or C3 segmental artery

We took the patient for surgery through a posterior approach and did a foramen magnum decompression with occipito-cervical fusion. The bilateral C1/2 joints were asymmetrical and the posterior arch of C1 was occipitalized. We entered the left C1/2 joint by using a subperiosteal dissection on the upper surface of the C2 pars, taking due care not to endanger the crossing VA. On the left side, we could expose the C1/2 joint more easily as the VA had entered the spinal canal below C2, leaving the C1/2 area “risk free.” The right C1/2 joint was drilled across to allow placement of a bone filled titanium spacer (6.5 mm) in an attempt to reduce the dislocation as well as the neck tilt. The left-sided joint was packed only with bone, after denuding the joint surface. Cervical fixation included C3/C5 on the right side while O-C3/4 fixation was performed on the left.^[6] The patient recovered uneventfully after surgery, after a period of elective ventilation for the first 48 postoperative h. She was discharged on the 7th postoperative day. Postoperative CT scan of the CVJ showed bony decompression and occipito-cervical construct *in situ*. She was ambulating with one person’s support and doing well at 6-week follow-up.

Case 2

A 50-year-old man presented with neck pain, numbness of the left hand and clumsiness in the legs while walking for 1 year. On examination, he had hyperreflexia in all 4 limbs

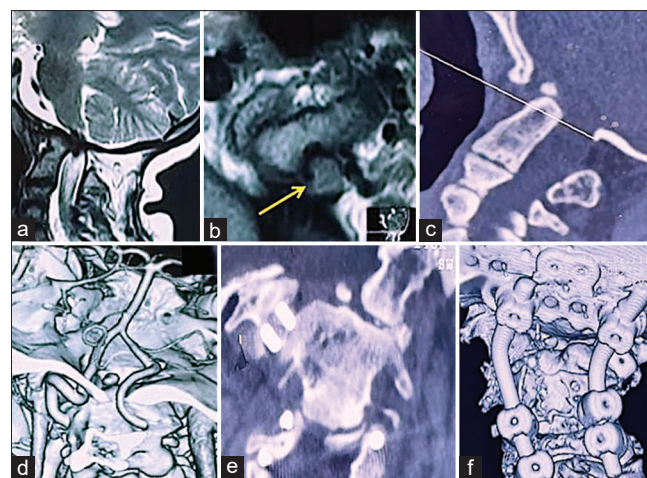


Figure 2: On magnetic resonance imaging, there is compression of the spinal cord at the level of C1 (a) with the flow void of intradural vertebral artery (yellow arrow) seen on the right side (b). On computed tomographic, there was increased atlanto-dental interval with narrowing of the foramen magnum (c). There was anterior dislocation of the right C1 lateral mass/occipital condyle on C2 (d). On computed tomographic angiography, bilateral vertebral artery were equal in calibre with abnormal course, computed tomographic angiography shows that the anomalous side vertebral artery is coursing below the isthmus (which represents the C2/3 foramen) and the contralateral side is normal route. Postoperative computed tomographic images showed the canal decompression, spacer placed at right C1/2 joint and the occipito cervical construct (e and f)

with bilaterally extensor plantar reflexes. There was subtle sensory impairment in the left hand, however, motor power was preserved in all four limbs.

On MRI, T2-weighted imaging abnormal cord signal intensity was observed at the level of C2 vertebra, suggesting an ongoing dynamic cord compression without any evidence of Chiari malformation. Axial sections at the level of C2 vertebra revealed an intradural location of the right VA with a hypoplastic left VA coursing normally through the bony foramen [Figure 3a-c]. CT imaging of the CVJ area revealed an abnormally increased anterior atlanto-dental interval suggesting an anterior AAD. There was the assimilation of the atlas at the level of the lateral mass, however, both the arches were free. The C2/3 joint on the right side was malformed with hypoplasia of the inferior articular process of C2 and an abnormally long C3 lateral mass forming the C2/3 joint. On CTA, a dominant right VA was seen entering the spinal canal below the pars of C2 vertebra before becoming vertical again. The C2 pars was thin and VA foramen on C2 as well as C1 were present, despite the aberrant course of the artery. Contralateral VA was hypoplastic with a normal course. The distal BA and posterior cerebral arteries could not be visualized clearly, unlike the first case.

We took the patient for posterior fixation to address the dynamic instability. Bilateral joint exploration and preparation of the joints proceeded normally. VA course was confirmed intraoperatively using a careful dissection. We performed C1 lateral mass-C2 pars interarticularis rod and screw fixation but resorted to C1–C3 (lateral mass) fixation on the right side, in view of the dominant VA coursing below a thinned out C2 pars. The thickness of the left C2 hemilamina did not permit us a translaminar screw placement. There was no gross injury to the VA [Figure 3d].

The patient was shifted to neurosurgical intensive care unit unreversed on a ventilator. At this point, he was neurologically intact. When the patient was re-assessed the next morning for possible extubation, a lack of response and sluggishly reacting pupils led to an urgent CT scan of the head and the CVJ. While the spinal CT showed a complete correction of the dislocation and well placed screws the cranial CT revealed hydrocephalus with diffuse cerebral edema including an effaced posterior fossa. An urgent external ventricular drainage ensued, however, without any improvement in the patient's status. The patient continued to deteriorate and eventually expired the next day. We speculated the cause of death to be thrombosis of the dominant VA secondary to the operative manipulation (discussed further).

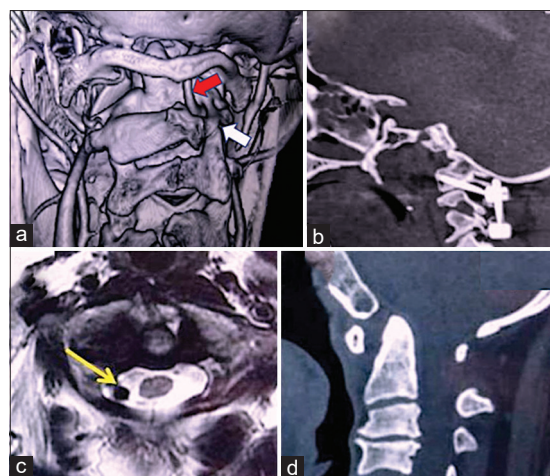


Figure 3: On computed tomographic angio, the left vertebral artery was hypoplastic and C2 segmental type while the dominant right sided vertebral artery was C3 type (a). On magnetic resonance imaging, there is cord intensity change at the level of C2 with intradural vertebral artery at the same level (b). There was associated hypoplasia of the right C2 hemilamina and poor visualization of the distal BA. Sagittal computed tomography of second patient show anterior atlanto-axial dislocation with mild basilar invagination (c). Postoperatively, there was total correction of the dislocation on postoperative computed tomographic with well-placed screws (d)

DISCUSSION

We described two cases of C3 segmental VA in the presence of CVJ anomaly with contrasting surgical outcomes. C3 segmental VA, where the extradural VA becomes intradural at the level of C2/3 vertebral foramen, is an extremely uncommon occurrence. To the best of our knowledge, this condition has been documented only four times before.^[3-5] Such an anomaly, although extremely rare, may have significant surgical considerations and hence neurosurgeons dealing with such patients need to be aware of such associations.

Embryology of C3 segmental vertebral artery

The V3 segment of the VA is intimately related with C1/2 area and the anatomy of this segment, with its variations, do have great implications during the surgical exposure and instrumentation for CVJ anomalies, particularly from a posterior approach.^[1,7,8] Normally, the V3 segment is divisible into 3 parts: the vertical part between C1 and C2 transverse processes, a horizontal part on the upper surface of C1 (the sulcus arteriosus) and an oblique part after leaving the sulcus arteriosus before entering the dura between the occiput and the C1.

In both our cases, the VA became intradural at C2/3 intervertebral level, a variation that has come to be known as the C3 segmental VA. This is an extremely rare situation, described first by Lasjunias.^[3] To the best of our knowledge, ours is the first report in patients with CVJ anomalies.

A more common anomaly related to our discussion is the C2 segmental VA, also called the persistent intersegmental artery. The prevalence of this anomaly is nearly 3.2%, the prevalence being higher in MR angiographic studies than the conventional angiographies. Here, the artery becomes intradural between C1 and C2.^[9]

Pertaining to our cases, the distal VA and its development assumes importance.^[10,11] The posterior circulation of the brain, i. e., BA develops as longitudinal neural artery and initially derives its supply from the anterior circulation through the posterior communicating artery, trigeminal artery, otic artery, and the hypoglossal artery. The Pro-atlantal artery represents an embryological artery that develops between the first intersegmental artery below and the hypoglossal artery above, at the level of the C1 nerve root. This artery persists in part in adult life as the distal part of the VA and origin of the anterior spinal artery (ASAs).

The adult VA develops from interlinking of the 6 intersegmental arteries.^[11] The last intersegmental artery generally persists as the subclavian artery while the remaining arteries disappear, allowing the continuation of subclavian artery with pro-atlantal artery derived distal VA and eventually the BA. When there is the persistence of the 1st intersegmental artery (passing between C1 and C2 foramen with C2 nerve root), the V3 segment of VA takes a medial course over the C1/2 joint (C2 segmental VA) or there is the duplication of the VA. When the second intersegmental artery persists, C3 segmental VA (between C2 and C3, traversing with the C3 nerve root) results, with accompanying disappearance of the distal VA in its expected course.

The final part of the puzzle pertains to the lateral spinal artery (LSA), described by Lasjunias.^[12] This artery is an embryonic structure that supplements the single ASA and two posterior spinal artery (PSA) on the spinal cord. The LSA is present only in the upper cervical spine which connects intradural VA or the PICA superiorly at the level of C1 and PSA below at the level of C4. This artery is located in front of the dorsal nerve root, as against the PSA which is positioned behind the root. The intersegmental arteries connect the metameric arteries extradurally through the intersegmental arteries. In both of our patients, LSA persisted owing to the persistence of the 2nd intersegmental artery, as evidenced by the lateral location of the artery with respect to the spinal cord.

Morbid anatomy of craniovertebral junction anomaly in association with C3 segmental artery

Our cases allow unique insight into the associated bony and soft-tissue anomalies, which have largely been speculated before. The first case represented a complex CVJ anomaly with

the presence of occipitalized atlas, C2/3 fusion, rotational deformity at C1/2 with right condyle-C1 lateral mass assembly dislocated anterior to C2. The C2 pars was rather thin. On angiography, contralateral VA was C2 segmental type and wider in caliber. The intracranial parts and BA were well visualized. There was no tonsillar ectopia. On the other hand, the bony complexity was of much lesser magnitude in the second case. Although C1 was partly occipitalized (at the level of lateral mass), there was no C2/3 fusion and there was no rotational component to the dislocation. Here also, the C2 pars was thinned out. In addition, the right hemilamina and the inferior articular process of C2 was hypoplastic and the superior articular process of C3 being unusually long. Angiographically, the C3 segmental VA was dominant, the contralateral artery being a C2 segmental type, but the caliber was much smaller. The VA foramina on the right side were well developed on C1 and C2 despite the variation in the course of V3, suggesting a delayed disappearance of the original VA. Distal BA was not well visualized on CTA which could be due to technical factors or it could indicate congenital narrowing. No soft-tissue CVJ anomaly was detected in this patient also.

Surgical implications and lessons learnt

C3 segmental VA may have several clinical and surgical implications. It can lead to spinal cord compression and myelopathy particularly if the condition is bilateral. Unlike at the level of C1, the subarachnoid space at the level of C2 is much narrower and C3 segmental VA can lead to pulsatile cord compression.^[13,14]

More importantly, this abnormal VA course assumes importance as far as the surgical exposure of this area and instrumentation are concerned. During the surgical exposure of the C1/2 joints, we found that the C3 segmental artery provided an advantageous situation as we did not have to worry about the VA while exposing the atlanto-axial joint. This was in contrast to the C2 segmental artery, which lies on the C1/2 joint and limits proper exposure of the joint and joint remodeling.^[1,7] It, however, posed challenges with respect to the choice of instrumentation. The fact that the artery was taking a course right beneath the C2 pars with thinning of the overlying pars, neither the pars interarticularis nor a transarticular screw placement was considered a safe option. C2 pedicle screw, taking a slightly higher and lateral entry point on the C2 pars could be one option but the screw heads (of C1 and C2 would have come too close to each other. The safer option, that we utilized, remains C3 lateral mass screw. When C2/3 are fused, the translaminar screw on the side of the abnormal artery is also a viable option.

We speculated postoperative VA thrombosis in our second patient. The C3 segmental VA being dominant, we thought

gradual thrombosis of this artery led to brainstem infarction. The questionable patency of the distal BA could have contributed as well but we cannot say for sure. What could have led to this? During surgery, there was no inadvertent damage during exposure and the C3 screw was a lateral mass screw, directed laterally away from the VA. Hence, we thought it could have resulted from the narrowing of the C2/3 foramen by the superior articular process of C3 during the tightening of the rod between C1/3. This appeared plausible as the joint was deformed and had radiological signs of instability. Such a situation should not have arisen when C2/3 remains fused and the C2/3 intervertebral foramen is completely formed, like our first patient. In the hindsight, we believe a prior decompression of the C2/3 foramen from behind on the side of the anomalous VA before placing the rod between C1 and C3 could have been a wiser decision. However, we agree that this is only our speculation but it remained a strong possibility as there was no other apparent cause of the mishap.

CONCLUSIONS

C3 segmental VA is an extremely rare anomaly in the development of VA and our paper is the first report of C3 segmental VA in CVJ anomalies. We saw that the anomalous artery affects the right side, associated with contralateral VA anomaly as well (C2 segmental VA) and a thin pars interarticularis of the ipsilateral C2 vertebra. This anomaly requires specific considerations for exposure and instrumentation in this area. Whether or not C2 and C3 are fused may have great surgical implications and it is desirable to decompress the C2/3 foramen before tightening the rod if C2/3 are not congenitally fused and C3 is chosen to be a part of the construct.

Consent

Informed consent for using clinical and radiological data from both the patients has been taken.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Sardhara J, Behari S, Mohan BM, Jaiswal AK, Sahu RN, Srivastava A, *et al.* Risk stratification of vertebral artery vulnerability during surgery for congenital atlanto-axial dislocation with or without an occipitalized atlas. *Neurol India* 2015;63:382-91.
2. Das KK, Deora H, Srivastava A, Khatri D, Shankar S, Sardhara J, *et al.* Elongated clivus with deficient anterior atlantal arch and congenital posterior atlantooccipital dislocation: Pathoembryology and management nuances of a rare form of proatlax segmentation anomaly. *World Neurosurg* 2019;126:286-90.
3. Lasjaunias P, Berenstein A, Ter Brugge KG. Surgical neuro-angiography. In: *Clinical Vascular Anatomy and Variations*. Vol. 1., 2nd ed.. Berlin: Springer; 2000. p. 224-60.
4. Uchino A, Saito N, Uemiya N, Sonoda K. Diagnosis of a C3 segmental type of vertebral artery by magnetic resonance angiography: report of two cases. *Surg Radiol Anat* 2016;38:873-6.
5. Moon JU, Kim MS. C3 segmental vertebral artery diagnosed by computed tomography angiography. *Surg Radiol Anat* 2019;41:1075-8.
6. Goel A, Laheri V. Plate and screw fixation for atlanto-axial subluxation. *Acta Neurochir (Wien)* 1994;129:47-53.
7. Srivastava A, Sardhara J, Behari S, Pavaman S, Joseph J, Das K, *et al.* Knock and drill technique: A simple tips for the instrumentation in complex craniovertebral junction anomalies without using fluoroscopy. *J Neurosci Rural Pract* 2017;8:14-9.
8. Sindgikar P, Das KK, Sardhara J, Bhaisorra KS, Srivastava AK, Mehrotra A, *et al.* Craniovertebral junction anomalies: When is resurgery required? *Neurol India* 2016;64:1220-32.
9. Uchino A, Saito N, Watadani T, Okada Y, Kozawa E, Nishi N, *et al.* Vertebral artery variations at the C1-2 level diagnosed by magnetic resonance angiography. *Neuroradiology* 2012;54:19-23.
10. Padget DH. The development of the cranial arteries in the human embryo. *Contrib Embryol* 1948;32:207-61.
11. Siclari F, Burger IM, Fasel JH, Gailloud P. Developmental anatomy of the distal vertebral artery in relationship to variants of the posterior and lateral spinal arterial systems. *AJNR Am J Neuroradiol* 2007;28:1185-90.
12. Lasjaunias P, Vallee B, Person H, Ter Brugge K, Chiu M. The lateral spinal artery of the upper cervical spinal cord. Anatomy, normal variations, and angiographic aspects. *J Neurosurg* 1985;63:235-41.
13. Satoh S, Yamamoto N, Kitagawa Y, Umemori T, Sasaki T, Iida T. Cervical cord compression by the anomalous vertebral artery presenting with neuralgic pain. Case report. *J Neurosurg* 1993;79:283-5.
14. Takahashi T, Tominaga T, Hassan T, Yoshimoto T. Cervical cord compression with myelopathy caused by bilateral persistence of the first intersegmental arteries: Case report. *Neurosurgery* 2003;53:234-7.