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Case Report

A case involving a giant aberrant craniocervical arteriovenous malformation

Agung Budi Sutiono, Jusuf Desman Banjarnahor

Neuroscience Centre Santosa Hospital Bandung Central, Department of Neurosurgery, Hasan Sadikin Hospital Padjadjaran University, Bandung, Indonesia.

E-mail: *Agung Budi Sutiono - agungbudis@gmail.com; Jusuf Desman Banjarnahor - jusupdesman@gmail.com

***Corresponding author:**
Agung Budi Sutiono,
Department of Neurosurgery,
Hasan Sadikin Hospital
Padjadjaran University,
Jl Pasteur 38, Bandung,
Indonesia.

agungbudis@gmail.com

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ABSTRACT

Background: Spinal cord arteriovenous malformations (AVMs) comprise about 3%–4% of primary intraspinal masses and are only rarely found external to the C2–C7 cervical vertebral foramen.

Case Description: A 21-year-old female presented with neck pain and a spastic quadriparesis of 1 year duration. The cervical magnetic resonance imaging and three-dimensional computed tomography angiograms documented an AVM/dural arteriovenous fistula on the right fed by multiple arteries located in the C5–C6 and C6–C7 foramen intervertebralis; utilizing a laminectomy, the large feeding arteries were double-clipped. This allowed for devascularization of the AVM and facilitated resection while preserving the aberrant vertebral artery. The patient was discharged within 1 week and, 2 months later, was able to ambulate to the outpatient clinic.

Conclusion: Double clipping of the two main right-sided arterial feeders at the C5–C6 and C6–C7 levels allowed for devascularization and resection of this large cervical AVM while carefully preserving the aberrant vertebral artery.

Keywords: Aberrant vertebral artery, Craniocervical junction, Dural arteriovenous fistulas, Spinal cord arteriovenous malformation

INTRODUCTION

Spinal arteriovenous malformations (AVMs) may be variously treated with microsurgery, endovascular embolization, and radiation therapy. However, surgery may be associated with considerable morbidity. Here, we present a type 1 AVM found in a 21-year-old female treated with both microsurgery and endovascular embolization.[1-4]

CASE REPORT

History and examination

A 21-year-old female presented with a progressive spastic cervical quadriparesis of 1 year duration. This was accompanied by significant bilateral upper and lower extremity spasticity and urinary retention.

Imaging studies

The craniocervical magnetic resonance imaging/magnetic resonance angiography/digital subtraction angiography revealed a large C1–C7 right-sided cervical spinal dural AVM; the main

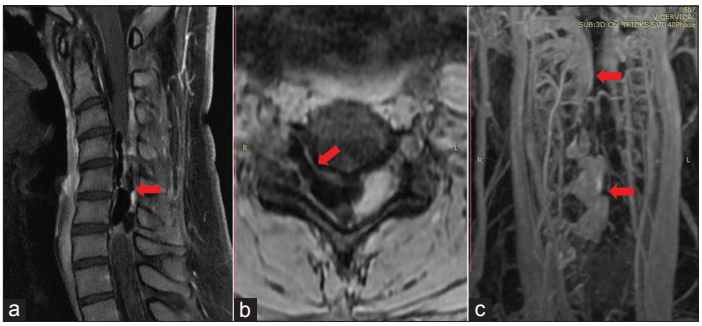

Figure 1: MRI Imaging. (a) Sagital section T1 weighted MRI imaging shows the hypointense multilobulated mass from cervical 4-6 suggested artery venous malformation (red arrow). (b) Axial section T1 weighted MRI shows the feeding artery course in the foramen intervertebralis to the spinal canal and compressed the spinal cord at level C6 (red arrow). (c) MR angiography shows huge AVM from Cranio-cervical junction to C7 (red arrow).

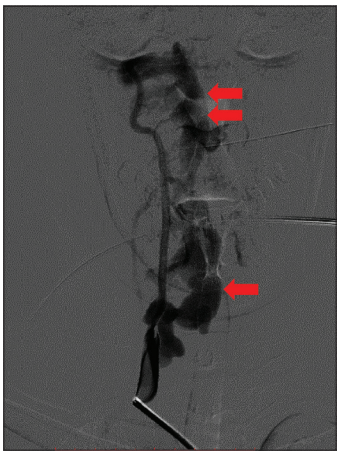

Figure 2: Digital subtraction angiography shows the AVM with huge feeding artery (single red arrow) at level C5-6, 6-7 and draining vein to the marginal plexus nearby foramen magnum (double red arrow).

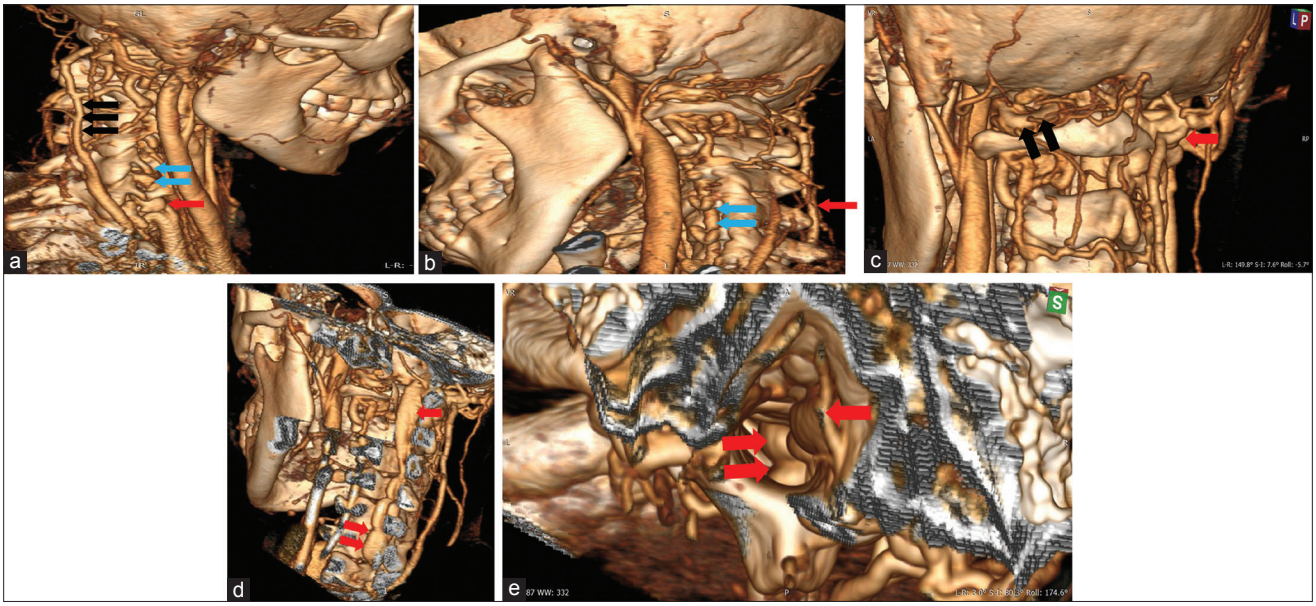

Figure 3: 3D CT angiography. (a) A huge feeder courses into the canalis spinalis through the foramen intervertebralis(single red arrow), transverse foramen are empty from vertebral artery (two blue arrows), the aberrant vertebral artery courses outside transverse foramen (three black arrows). (b) Right aberrant vertebral artery (single red arrow) and normal left vertebral artery courses inside the transverse foramen (double blue arrows). (c) Right vertebral artery loop at the C1 to occipital bone just above the C1 transverse (single red arrow), Left vertebral artery courses normally from inside transverse foramen C1 to occipital bone to enter the intracranial portion (double black arrows). (d) 3D CT after doing hemilaminectomy in computer simulation shows a long thin AVM courses at C2-3 (single red arrows) and huge widen AVM at level C4-6 (double red arrows). (e) 3D CT in axial section at level foramen magnum and C1 shows AVM at C2-3 (single red arrow), and huge AVM at C4-6 is larger than in the level C2-3 (double red arrow).

two feeders were coming from the C5–C6 and C6–C7 neural foramen [Figures 1 and 2]. Smaller feeders were also coming from the C2–C3 foramen, in conjunction with huge draining veins. Three-dimensional computed tomography angiography further elucidated the location of the AVM and its feeders [Figure 3].

Surgery and post-operative course

Using a midline C1–C7 cervical exposure, a laminectomy was performed at the C5 and C6 levels. On the right, huge feeding arteries were identified entering the respective vertebral foramina of C5–C6 and C6–C7; utilizing extensive foraminotomies, double

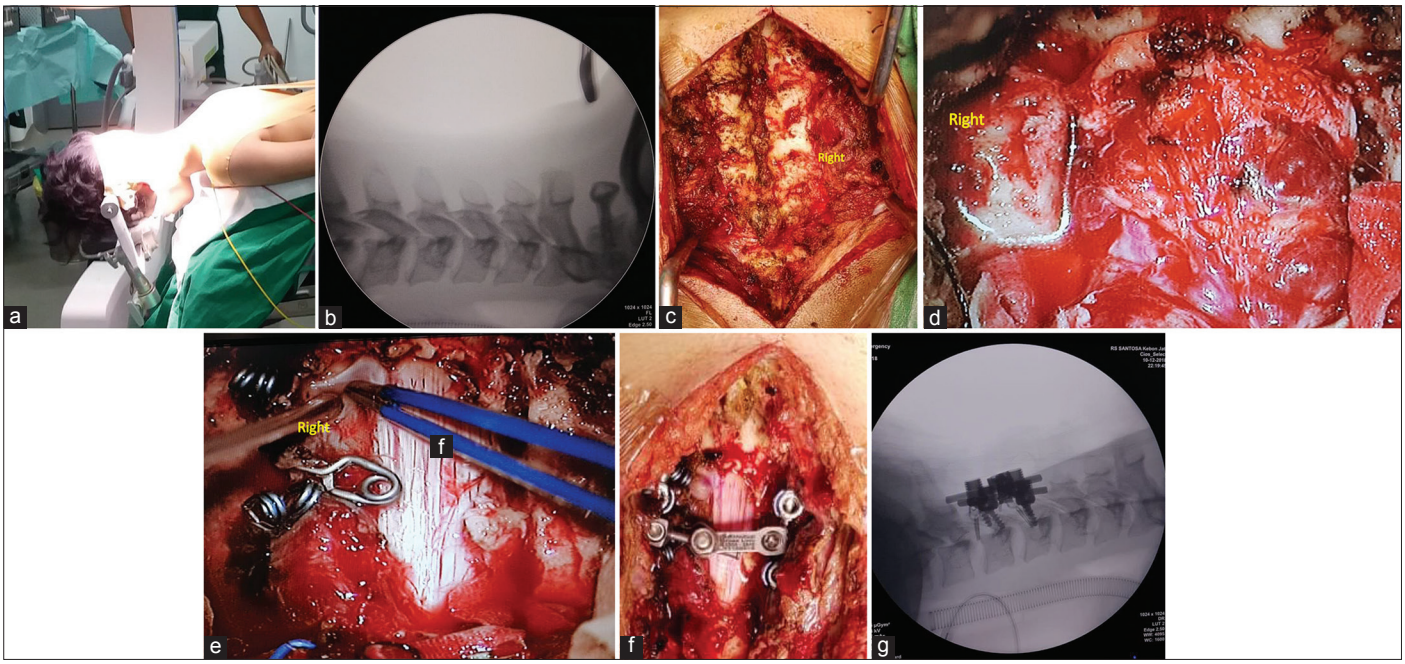

Figure 4: Patient position. (a) Prone position with head is holding by Mayfield head frame to fix the cervical spine in straight position. (b) Cervical x ray using C-arm to confirm cervical level. (c) Midline incision and dissecting the paraspinal muscles to expose the lamina from Occipital to C7. (d) Laminectomy was performed at C5-6 and exposed the AVM with very thin AVM wall. (e) Double permanent clips was done at huge main feeding artery at C4-5, 5-6 and resect the AVM to expose the dura and decompress cervical cord. (f) The lateral mass was placed at C5-6 bilaterally and tightened with cross link titanium plate. (g) The position of lateral mass at C5-6 and clip are confirmed using C-arm.

curved permanent clips were placed on the major feeders at each respective level. This allowed for decompression and resection of the AVM and was followed by lateral screws/rod fusion [Figure 4]. Her quadriparesis continued to improve over 6 post operative days, and she was discharged on day 7. 2 months later, she was able to ambulate (e.g., Karnofsky Performance Status score 100) [Figure 5].

DISCUSSION

Vascular malformations of the spinal cord and dura account for approximately 3%–4% of spinal cord lesions; they are the least common in the cervical region.[6] Our patient, with a 1-year onset of a spastic quadriparesis, had a spinal type I AVM with multiple feeders that originated predominantly from the right vertebral artery (C5–C6 and C6–C7 neural foramina).

Reports of Other Aberrant VA Most vertebral arteries enter the transverse foramina at the C6 level (93%, 855 cases of 920 cases).[7,5] Ulusoy *et al.* reported a 15-year-old female with a Klippel–Feil syndrome who showed an extraforaminal, cranially ascending aberrant right VA that originated from the ipsilateral carotid bulb. Shin *et al.* also reported a similar aberrant VA. In our case, the feeding arteries originated from the right C5–C6 nd C6–C7 transverse foramen; laminectomy and foraminal decompression allowed for double clipping of the feeders, facilitating decompression, and subsequent complete resection of the AVM while preserving the aberrant right vertebral artery.

Figure 5: Post-operative course. (a) 4 Days after surgery, she improved her upper extremities gradually. (b) 1 Month after surgery, she could stand up but still having difficulties in walking. (c) 2 Months after surgery, she could walk normally.

The patient then underwent C5–C6 lateral mass screw fixation/fusion [Figure 4]. Within 2 post operative months, her spastic quadriparesis markedly improved [Figure 5].

CONCLUSION

For patients with cervical AVM, preserving the aberrant VA while double clipping other AVM feeders is critical to maintain neurological function.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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