May 2012

Concurrent intracranial and spinal arteriovenous malformations: Report of two pediatric cases and literature review.

Hussain Shallwani
Aga Khan University

Muhammad Z Tahir
Aga Khan University

Muhammad Ehsan Bari
Aga Khan University

Tanveer-Ul-Haq
Aga Khan University

SURGICAL NEUROLOGY INTERNATIONAL
AN OPEN ACCESS, INTERNATIONAL JOURNAL OF NEUROSURGERY AND NEUROSCIENCES
Home | Browse articles | Instructions | Submit articles

Surg Neurol Int. 2012; 3: 51.
Published online 2012 May 14. doi: 10.4103/2152-7806.96074
PMCID: PMC3356988

Concurrent intracranial and spinal arteriovenous malformations: Report of two pediatric cases and literature review

Hussain Shallwani, Muhammad Z. Tahir,[1] Muhammad E. Bari,[2,*] and Tanveer-ul-Haq[3]

Student, Medical College, Aga Khan University Hospital, Karachi, Pakistan
[1]Department of Neurosurgery, Aga Khan University Hospital, Karachi, Pakistan
[2]Department of Surgery, Aga Khan University Hospital, Karachi, Pakistan
[3]Department of Radiology, Aga Khan University Hospital, Karachi, Pakistan
Hussain Shallwani: hshallwani1@gmail.com; Muhammad Z. Tahir: doctorneuron1@yahoo.com; Muhammad E. Bari: ehsan.bari@aku.edu; Tanveer-ul-Haq: tanveer.haq @aku.edu
*Corresponding author

Received 2012 Feb 1; Accepted 2012 Apr 12.

Abstract

Go to:

Background:

Concurrent intracranial and spinal arteriovenous malformations (AVMs) are very rare with only a few cases being reported in literature. Two of the rare concurrent intracranial and spinal AVM cases are presented.

Case Description:

Case 1 is a 12-year-old girl with headache and motor disturbances in the lower limbs. Her spinal and brain angiogram was done and she was diagnosed to have a spinal AVM at level T8–T9 and an intracranial AVM in the left mesial temporal lobe. Her spinal

AVM was embolized, while no treatment was given for her intracranial AVM. Case 2 is a 10-year-old girl who presented with headache and quadriparesis. Her brain and spinal angiogram revealed an intracranial AVM in the left parietal lobe and a spinal AVM at level C2, respectively. Craniotomy and excision was done for her intracranial AVM and embolization for the spinal AVM.

Conclusion:

It is proposed that multiple AVMs may be a result of yet unrevealed pathogenesis or strong embryogenetic anomaly, which may be different from that involved in single AVM. With lack of consensus over the best therapeutic strategy, multimodality treatment based on the individual's needs is suggested.

Keywords: Arteriovenous malformation, cerebral, concurrent, intracranial, multiple, spinal

INTRODUCTION

Go to:

Single intracranial arteriovenous malformation (AVM) along with a spinal AVM is very rare with less than 10 cases being reported in literature since 1969. Multiple intracranial AVMs with a spinal AVM are even more infrequent; only three other cases have been reported in the literature, one being an autopsy case. We report two such uncommon cases involving an intracranial AVM along with a spinal AVM.

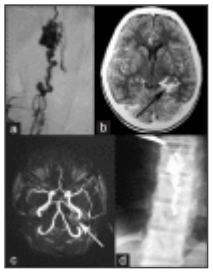
CASE REPORTS

Go to:

Case 1

The first case of intracranial AVM along with a spinal AVM involves a 12-year-old girl who presented to us with a 3-month history of headache and progressively worsening spastic paraparesis with reflex spasms of both lower limbs. There was no history of lower back pain and no bladder or bowel disturbances. Examination showed power of 3/5 in the right lower limb and 4/5 in the left lower limb, with normal bulk but increased muscle tone. She also had brisk reflexes in both lower limbs. Cognitive functions, speech, and upper limb reflexes were normal, along with intact sensations and cerebellar functions. She had been in a good state of health in the past and her family history was negative for any hereditary vascular disorders or AVMs. Magnetic Resonance Imaging (MRI) of dorsolumbar spine showed epidural flow voids. Spinal angiogram showed an AVM in mid-dorsal region at level T8–T9 with three feeders comprising left 8th and right 9th and 10th intercostal arteries [Figure 1a]. Brain imaging studies were done to investigate any intracranial pathology responsible for her persistent headache. Her brain MRI and Magnetic Resonance Angiography (MRA) revealed a small AVM in the left hippocampus supplied by left posterior cerebral artery and with deep venous drainage [Figure 1b and c]. There was no hemorrhage from the lesions, and ischemia due to spinal AVM seemed to be the probable cause of her paraparesis.

Figure 1



(a) Perimedullary arteriovenous malformation (AVM) located in mid dorsal region. (b) Po weighted image showing concurrent AVM (black arrow) in left hippocampus. (c) MRA sl (white arrow) supplied by left posterior cerebral artery. **...**

Spinal AVM embolization was carried out by polyvinyl acetate (PVA) and histoacryl particles under general anesthesia and complete embolization was achieved [Figure 1d]. No treatment was offered for her cerebral AVM and her headache was managed conservatively with analgesics. The hospital course was smooth and she was discharged after a total of 5 days of hospital stay.

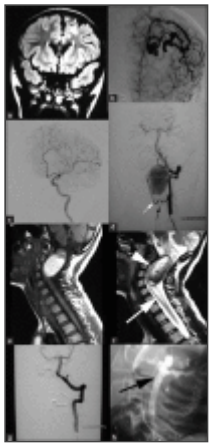
On follow-up after 1 year, the patient was doing well with no significant symptoms. She was able to work independently and did not require further treatment.

Case 2

The second case of single intracranial AVM with concurrent spinal AV fistula involves a 10-year-old girl who presented to us with headaches and progressively increasing weakness of all four limbs for 2½ years. On examination, she had decreased bulk of muscles and increased tone in both upper and lower limbs. She also had brisk reflexes, upgoing planters, and clonus. She was quadriparetic with power of 3/5 in the right upper and lower limbs and 4/5 in the left half of body. She was otherwise conscious and well oriented to time, place, and person, and her sensations were intact. Her past history was unremarkable and there was no significant family history of any hereditary vascular disorder or AVMs. Her brain MRI and cerebral angiography revealed left parasagittal AVM of medium size in the parietal region with superficial venous drainage [Figure 2a and b]. Craniotomy and excision was done in another institute. Postoperatively, she was doing well with no residual AVM on her cerebral angiography [Figure 2c].

Figure 2

(a) Left parietal arteriovenous malformation (AVM). (b) Left parasagittal AVM. (c) Post-procedure angiogram showing no residual AVM. (d) Perimedullary AV fistula with large aneurysm in cervical region (black arrow showing early filling vein, white arrow **...**



After her cranial surgery, her symptoms recurred and she showed progressive weakness and signs of myelopathy in all four of her limbs. Angiography was repeated, which revealed an AV fistula in the cervical region [Figure 2d]. Multiple feeders were observed that included bilateral vertebral arteries, bilateral posterior inferior cerebellar artery (PICA), and left costocervical artery from subclavian artery. Some tortuosity was observed in the previously treated AVM in the left parietal region, but no arteriovenous (AV) shunting was seen. Few tortuous abnormal vascular channels were seen in venous phase. MRI also showed a cervical aneurysm [Figure 2e and f]. There was no hemorrhage from the lesion and her symptoms were probably because of ischemia secondary to steal phenomenon.

Angiographic coiling of aneurysm was done and feeders from vertebral arteries and PICA were embolized. Tiny feeder from left costocervical artery was embolized with histoacryl glue. No residual AV fistula was seen in post-procedural angiography [Figure 2g and h] and no postoperative complications occurred; patient was stable and got discharged.

On follow-up, the patient was doing well and the treatment of AV fistula seemed to be adequate. However, after 1 year and 4 months, the patient showed signs of mild weakness in left-sided upper and lower limbs. She was able to carry out most of the tasks independently; however, she limped while walking and had weakness of left hand and fingers. The patient refused further evaluation and management.

DISCUSSION

Go to:

Multiple AVMs have been seen to be associated with hereditary disorders such as Rendu–Osler–Weber syndrome or hereditary hemorrhagic telangiectasia. It is extremely uncommon to see concurrent intracranial and spinal AVM not associated with such syndromes.

Single intracranial AVM along with a spinal AVM is very rare, with total number of cases reported in the literature being less than 10 since 1969.[8,9,15,20,22] Multiple intracranial AVMs with a spinal AVM are even more infrequent; only three other cases have

been reported in the literature, one being an autopsy case.[7,13,14] The earliest cases comprise those reported by Di Chiro *et al*. in 1972[1] and 1973;[3] however, exact details of those cases were not available. Also, Krayenbuhl *et al*. reported such a case involving AVMs in cerebellum and spinal cord earlier in 1969.[11] The details of all the intracranial AVMs coexisting with spinal AVMs are listed in Table 1, along with our cases. The sizes of the intracranial AVMs were graded as small (<3 cm), medium (3–6 cm), or large (>6 cm), and their venous drainage as superficial or deep, in accordance with Spetzler and Martin's grading system.[18] The spinal AVMs were grouped into single coiled, glomus, and juvenile types as Di Chiro *et al*. classified them.[2,4]

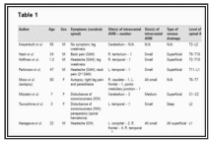


Table 1

Concurrent intracranial and spinal arteriovenous malformations – Comparison of cases in the literature

The age at presentation ranged from 1.3 to 55 years, with a mean age of 23.8 years. There were five males and six females. Six cases presented with subarachnoid hemorrhage (SAH), intraventricular hemorrhage (IVH), or intracerebral hemorrhage (ICH) as a result of hemorrhage from AVM. Out of the 19 intracranial AVMs whose size was known, there were 16 small and 3 medium intracranial AVMs. Moreover, out of the 17 lesions with known venous drainage, 15 intracranial AVMs had superficial and only 2 had deep venous drainage. The level of spinal AVM ranged from C1 to L2. There were four glomus, two dural, and one juvenile spinal AVMs.

AVMs are congenital lesions occurring between the fourth and eighth weeks of embryonic development when vessels differentiate into arteries, veins, and capillaries.[5] These lesions are formed by masses of abnormal arteries and veins lacking a true capillary bed in the nidus.[23] The nidus of the AVM comprises large vessels without the elastic layer in the walls. The arteries are deficient of muscularis layer and the draining veins are often dilated due to the high velocity of the flowing blood. Various studies have been done to identify the role of angiogenic factors in the pathogenesis of AVM.[17,21,24] Tamaki *et al*. proposed various developmental defects or multiple failures in the persistence of primitive capillary beds as the pathogenesis of multiple AVMs.[19] According to Hasegawa *et al*., extensive disturbances in early ephrin/ephrin receptor interactions in the embryo may cause multiple AVMs.[7]

Familial AVMs are extremely rare, manifesting at a younger age and occurring more frequently in females.[25] Yet, it has been proposed that genetic factors may be involved in the occurrence of familial AVM.[25] Previously, *ALK1* and *ENG* genes had been shown to be associated with sporadic brain AVM.[16] Moreover, single nucleotide polymorphism in interleukin (IL) genes has also been associated with increased risk of AVM among certain racial or ethnic groups.[10] Nevertheless, multiple AVMs may be result of yet unrevealed pathogenesis or strong embryogenetic anomaly, which may be different from that involved in single AVM.

AVM is known to produce harm by its rupture or rupture of an associated aneurysm, by causing seizures, or by causing ischemia of the adjacent brain matter by the steal phenomenon.[12] Hence, it is essential to screen and treat AVMs as the symptoms require. Hash *et al*. suggested that screening for multiple AVMs is warranted in cases when a single lesion does not explain the presenting

symptom or sign.[8] Parkinson *et al*. also advised spinal angiography for investigation of spontaneous SAH in patients with no demonstrable intracranial source.[15]

The angioarchitectural factors that increase the risk of hemorrhage from an AVM include the presence of flow-related aneurysm, presence of intranidal aneurysm, deep venous drainage, deep (periventricular) location, small nidus size (<3 cm), high feeding artery pressure, slow arterial filling, and venous stenosis.[6] With the main aim of complete angiographic obliteration of the AVM, treatment modalities like microsurgery, endovascular embolization, and stereotactic radiosurgery have an established role in the treatment of patients with AVM, and a staged approach has been proposed for patients with multiple AVMs.[6] We suggest that multimodality treatment tailored for individual cases should be practiced.

CONCLUSION

Go to:

Multiple AVMs can co-exist in brain and spinal cord, and can be difficult to detect and manage. We suggest that in cases where one lesion does not explain all the symptoms or if there is progressive worsening of symptoms, there should be a high index of suspicion for another lesion and further investigations are warranted. For patients with concomitant intracranial and spinal AVM, multimodality treatment tailored for individual cases currently seems to be the best approach.

Footnotes

Go to:

Available FREE in open access from: http://www.surgicalneurologyint.com/text.asp?2012/3/1/51/96074

REFERENCES

Go to:

1. Di Chiro G. Recent successes and failures in radiographic and radioisotopic angiography of the spinal cord. Br J Radiol. 1972;45:553–60. [PubMed]

2. Di Chiro G, Doppman J, Ommaya AK. Selective arteriography of arteriovenous aneurysms of spinal cord. Radiology. 1967;88:1065–77. [PubMed]

3. Di Chiro G, Wener L. Angiography of the spinal cord. A review of contemporary techniques and applications. J Neurosurg. 1973;39:1–29. [PubMed]

4. Doppman JL, DiChiro G, Ommaya AK. Selective Arteriography of the Spinal Cord. Warren H. Green: St. Louis; 1969.

5. Fisher WS., 3rd . Concomitant intracranial aneurysms and arteriovenous malformations. In: Wilkins RH, Rengachary SS, editors. Neurosurgery. 2nd ed. New York: McGraw-Hill; 1996. pp. 2429–31.

6. Fleetwood IG, Steinberg GK. Arteriovenous malformations. Lancet. 2002;359:863–73. [PubMed]

7. Hasegawa S, Hamada JI, Morioka M, Kai Y, Takaki S, Ushio Y. Multiple cerebral arteriovenous malformations (AVMs) associated with spinal AVM. Acta Neurochir (Wien) 1999;141:315–9. [PubMed]

8. Hash CJ, Grossman CB, Shenkin HA. Concurrent intracranial and spinal cord arteriovenous malformations. Case report. J Neurosurg. 1975;43:104–7. [PubMed]

9. Hoffman HJ, Mohr G, Kusunoki T. Multiple arteriovenous malformations of spinal cord and brain in a child. Case report. Childs Brain. 1976;2:317–24. [PubMed]

10. Kim H, Su H, Weinsheimer S, Pawlikowska L, Young WL. Brain arteriovenous malformation pathogenesis: A response-to-injury paradigm. Acta Neurochir Suppl. 2011;111:83–92. [PMC free article] [PubMed]

11. Krayenbühl H, Yaargil MG, McClintock HG. Treatment of spinal cord vascular malformations by surgical excision. J Neurosurg. 1969;30:427–35. [PubMed]

12. Laakso A, Dashti R, Juvela S, Niemelä M, Hernesniemi J. Natural history of arteriovenous malformations: Presentation, risk of hemorrhage and mortality. Acta Neurochir Suppl. 2010;107:65–9. [PubMed]

13. Mizutani T, Tanaka H, Aruga T. Multiple arteriovenous malformations located in the cerebellum, posterior fossa, spinal cord, dura, and scalp with associated port-wine stain and supratentorial venous anomaly. Neurosurgery. 1992;31:137–41. [PubMed]

14. Moss JG, Sellar RJ, Hadley DM. Intracerebral and spinal vascular malformation in a patient without hereditary haemorrhagic telangiectasia. Neuroradiology. 1989;31:280–1. [PubMed]

15. Parkinson D, West M. Spontaneous subarachnoid hemorrhage first from an intracranial and then from a spinal arteriovenous malformation. Case report. J Neurosurg. 1977;47:965–8. [PubMed]

16. Pawlikowska L, Tran MN, Achrol AS, Ha C, Burchard E, Choudhry S, et al. Polymorphisms in transforming growth factor-beta-related genes ALK1 and ENG are associated with sporadic brain arteriovenous malformations. Stroke. 2005;36:2278–80. [PubMed]

17. Rothbart D, Awad IA, Lee J, Kim J, Harbaugh R, Criscuolo GR. Expression of angiogenic factors and structural proteins in central nervous system vascular malformations. Neurosurgery. 1996;38:915–25. [PubMed]

18. Spetzler RF, Martin NA. A proposed grading system for arteriovenous malformations. J Neurosurg. 1986;65:476–83. [PubMed]

19. Tamaki N, Fujita K, Yamashita H. Multiple arteriovenous malformations involving the scalp, dura, retina, cerebrum, and posterior fossa. Case report. J Neurosurg. 1971;34:95–8. [PubMed]

20. Tsurushima H, Meguro K, Matsumura A, Narushima K, Nakada Y, Nose T. Multiple arteriovenous malformations of spinal cord and brain in a child. Pediatr Neurosurg. 1995;23:166–70. [PubMed]

21. Uranishi R, Baev NI, Ng PY, Kim JH, Awad IA. Expression of endothelial cell angiogenesis receptors in human cerebrovascular malformations. Neurosurgery. 2001;48:359–68. [PubMed]

22. Wang Y, Zhang H, Ling F. Coexistence of a single cerebral arteriovenous malformation and spinal arteriovenous malformation. Neurol India. 2009;57:785–8. [PubMed]

23. Weinsheimer S, Kim H, Pawlikowska L, Chen Y, Lawton MT, Sidney S, et al. EPHB4 gene polymorphisms and risk of intracranial hemorrhage in patients with brain arteriovenous malformations. Circ Cardiovasc Genet. 2009;2:476–82. [PMC free article] [PubMed]

24. Wong JH, Awad IA, Kim JH. Ultrastructural pathological features of cerebrovascular malformations: A preliminary report. Neurosurgery. 2000;46:1454–9. [PubMed]

25. Yokoyama K, Asano Y, Murakawa T, Takada M, Ando T, Sakai N, et al. Familial occurrence of arteriovenous malformation of the brain. J Neurosurg. 1991;74:585–9. [PubMed]