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Brief Report: Concurrent cervical giant perimedullary arteriovenous fistula, aneurysm on a feeding artery of fistula and unilateral congenital carotid aplasia

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Abstract

Giant perimedullary arteriovenous fistulae (GPAVFs) are extremely rare, particularly cervical GPAVFs whose incidence has not been tabulated. The occurrence of aneurysm on an artery feeding a GPAVF has previously not been described. Internal carotid artery aplasia is also very rare (0.01%). The concurrence of these disorders has previously not been recorded. We report a case of a 5-year-old female with increasing headaches, who was found to have intraventricular hemorrhage and above anomalies. Coil embolization of GPAVF and the adjacent aneurysm was attempted. Treatment was complicated by stroke and death. Embryological and anatomical factors underlying these anomalies as well as, management options are discussed.

Introduction

Cervical giant perimedullary arteriovenous fistulae (GPAVF) and internal carotid artery (ICA) aplasia are rare disorders in their own right. Because of the paucity of its occurrence, the incidence of cervical GPAVFs has not been tabulated.[1] The prevalence of ICA aplasia is estimated at 0.01%.[2] The occurrence of an aneurysm on an artery feeding the GPAVF has previously not been reported, nor has the concurrent occurrence of any two or all three of these rare entities. While GPAVF and ICA aplasia are congenital anomalies, the aneurysm most likely was acquired consequent to the hemodynamic stress placed upon the feeding artery owing to high flow to the GPAVF. We report the first case presenting with all three entities and discuss the anatomical and clinical implications of this unique presentation.

Methods

Clinical presentation

A female of 5 years of age presented with increasing headaches. At transfer to our institution, she was neurologically intact. Computed tomography (CT) scan demonstrated intraventricular hemorrhage and ventricular enlargement (Figure 1A). She was admitted to pediatric intensive care unit. Magnetic resonance imaging (MRI) of the brain and spine demonstrated a large cervical lesion suggestive of spinal arteriovenous malformation (Figure 1B). Angiography performed by one of the senior authors (SHK), revealed this to be a 4 cm 1 cm GPAVF with a 5 mm aneurysm on a feeding artery, just proximal to a large venous varix (Figure 2). The fistula was supplied by cervical branches coming off both vertebral arteries (VA), the radicular branch from the left VA to the anterior spinal artery (ASA), and the left thyrocervical trunk. Angiography also revealed absence of the right internal carotid artery with a “fetal” type circulation to the right cerebrum via anterior communicating and right posterior communicating arteries (Figure 3). No embryonal type vessels (rete mirabile) were detected. Additionally, the bone window on CT scan revealed a well-developed left carotid canal and a significantly hypoplastic one on the right (Figure 3B). The treating physician (CT) opted for coil embolization of the GPAVF and the associated aneurysm. During intervention, a part of the coil was inadvertently deposited in the adjacent ASA. Approximately 3h later, there was a profound decline in the patient’s level of consciousness. She became unresponsive, was noted to be hypertensive, initially with left pupil dilatation and then bilateral fixed dilated pupils. There was loss of pupillary, cough, and gag reflexes. CT and MRI of the brain demonstrated findings consistent with global hypoxic ischemic injury.

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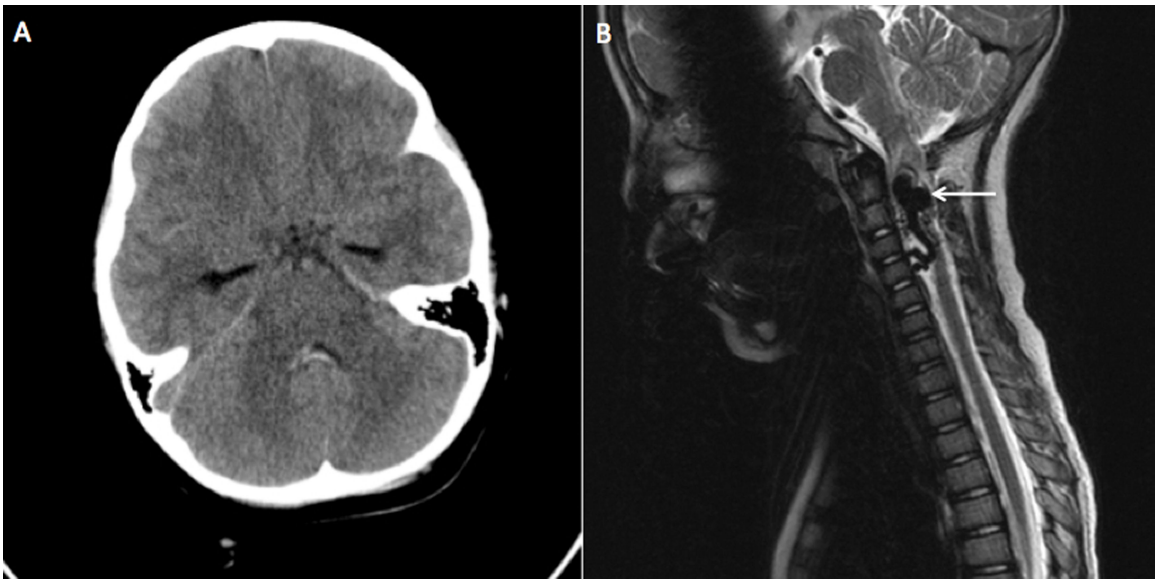

Figure 1. Cervical Gpavf. Panel a demonstrates presence of blood within the fourth ventricle on axial Ct scan without contrast that was performed at presentation. the temporal horns are apparent. Panel B is a T2 weighted Mri of the cervical spine showing a 4 cm × 1 cm serpiginous flow void consistent with a dural arteriovenous malformation at the cervicomedullary junction.

Discussion

GPAVFs are extremely rare vascular malformations. Cervical GPAVFs in particular are so rare that an incidence has not been tabulated.[1] According to the classification of Bao *et al*,3 GPAVFs are classified as type 3 perimedullary arteriovenous fistulae because of their large size, multiple dilated arterial supply, epidural venous drainage and high flow system.[3–6] GPAVFs were first described as intradural extramedullary spinal arteriovenous malformations fed predominantly by ASA.[7] The giant lesions show a higher frequency in children in contrast to the more typical perimedullary arteriovenous fistulae that are seen more commonly in middle-aged men.[8] Additionally, these GPAVFs have a higher tendency toward hemorrhage than their more common thoracolumbar counterparts.[8] Consistent with previous literature about bleeding predilection in cervical spinal GPAVFs, our patient also presented with hemorrhage into the ventricular system.[1,6] Similar to the type 1 and type 2 fistulae, type 3 can present with hemorrhage from venous hypertension and vessel rupture, as well as symptoms from steal phenomenon, or direct compression from ectatic veins. The absence of aneurysms on feeding arteries of the GPAVF was specifically noted in previous literature.[6] Our report is the first time such an aneurysm has been found. The presence of such an arterial aneurysm, lends yet another potential source for hemorrhagic presentation. We postulate that in contrast to the GPAVF and ICA aplasia that are congenital, the arterial aneurysm was acquired consequent to the hemodynamic stress placed upon the feeding artery by a high flow demand. Such aneurysms are known to occur on feeding arteries subjected to high flow, for example, in cerebral arteriovenous malformations.[9,10] In addition to headaches, patients may also present with progressively worsening myelopathy or radiculopathy. If left untreated, complete spinal transection has been shown to occur over a course of 7–9 years.[11,12]

The angiographic findings are indicative of developmental anomalies. It is known that a lack of capillary formation from the primitive vascular plexus during second embryological week leads to arteriovenous malformations. Development of the large cranial vessels occurs between 5th and 7th week. Aplasia of internal carotid artery is rare with prevalence estimated at 0.01%.[2] The presence of the right carotid canal, albeit hypoplastic, and absence of rete mirabile, indicate that our patient had aplasia rather than agenesis of right ICA. In other words, the right ICA development did commence, followed by involution.[13,14] The right ICA aplasia, as seen in our patient, is three times less likely to occur compared with the left side.[15] The significance of the left side predilection for ICA aplasia is unclear. The presence of a normal ipsilateral external carotid artery (ECA) in face of carotid aplasia has been repeatedly

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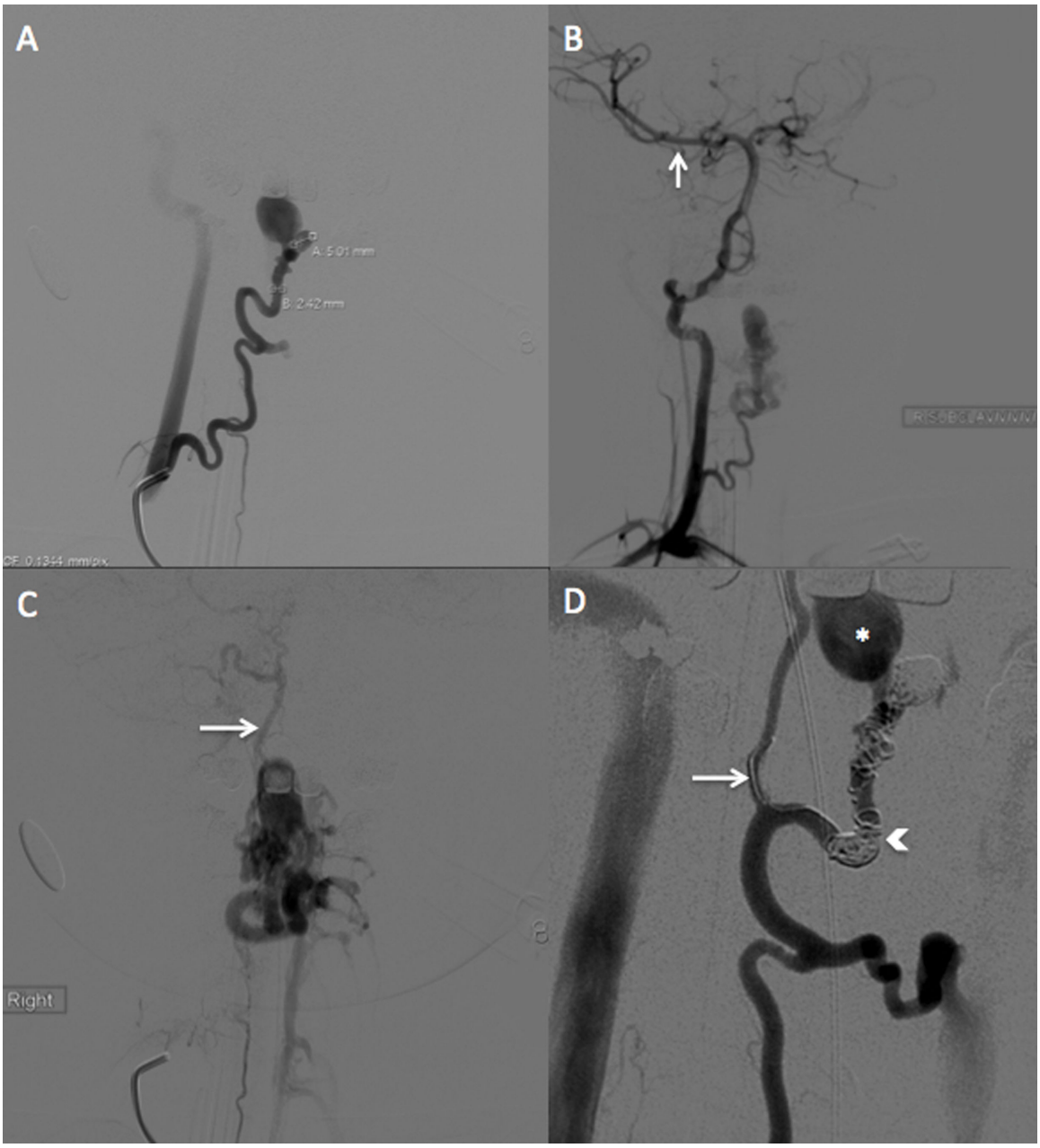

Figure 2. Angiography demonstrating Gpavf and aneurysm on feeding artery. Panel a is a right vertebral angiogram showing an enlarged branch from the Va supplying the Gpavf. an aneurysm (with measurements) is appreciated on the feeding artery, just proximal to a large venous pouch or aneurysm. the latter is known to occur in Gpavf. Panel B demonstrates that the vertebral artery also supplies anterior circulation. the right Mca (arrow) is more obvious than left. Again noted is the Gpavf located further caudally. Panel C shows the Gpavf in late arterial phase. Asa appears hypertrophied rostrally (arrow). Panel D shows selected catheterization of the enlarged feeding vessel arising off right Va in early arterial phase. It demonstrates the Gpavf, coil embolization of the arterial aneurysm (arrow) and partial embolization of the feeding artery. the venous varix (*) continues to fill robustly. the distal end of the coil is noted in the Asa (arrowhead).

noted. This observation supports the likelihood that the ECA does not share a common origin with ICA from the third aortic arch. Rather, similar to the common carotid artery (CCA), the ECA originates from the aortic

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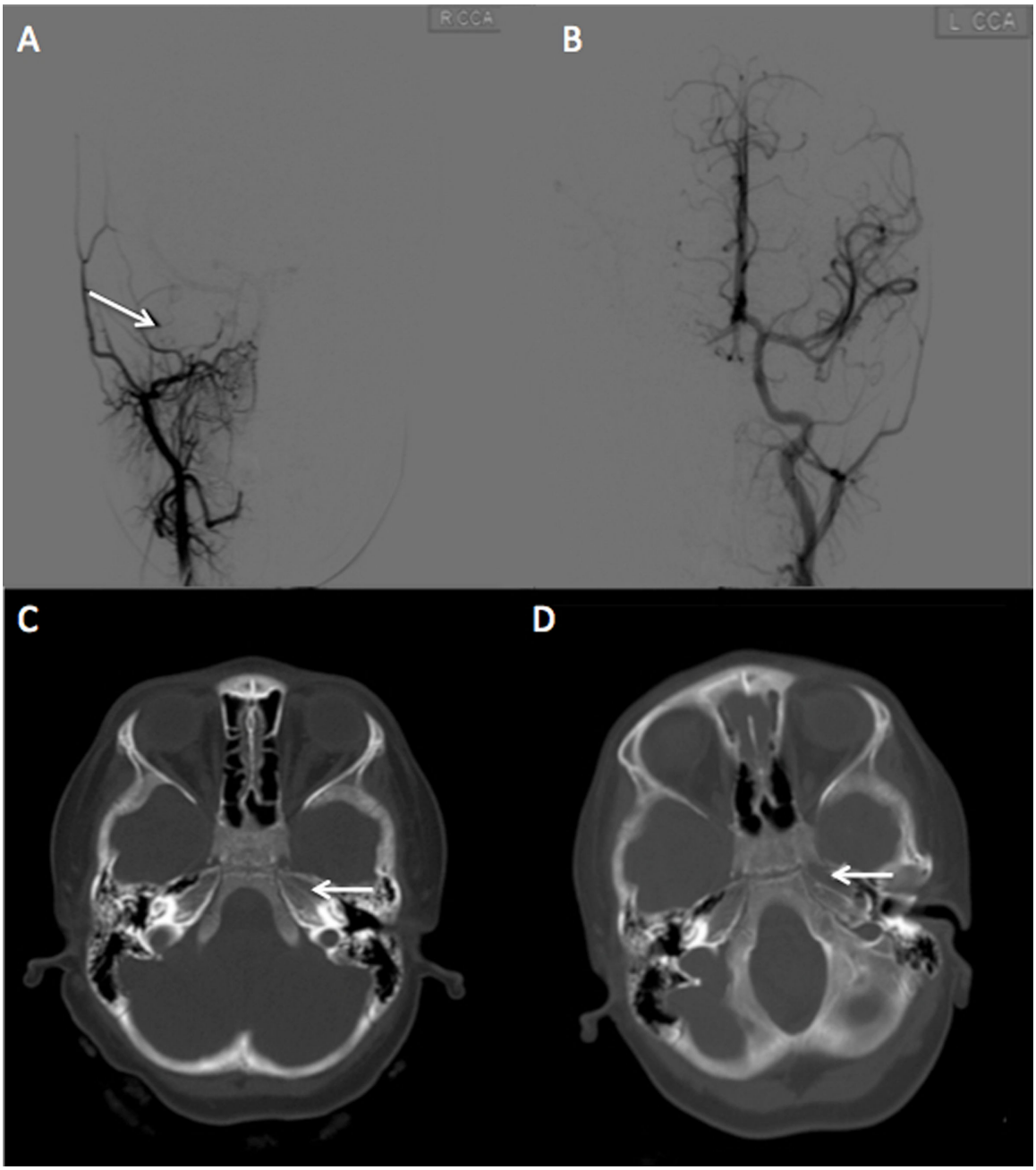

Figure 3. Aplasia of right internal carotid artery. Panel a shows right common carotid angiography anteroposterior view, demonstrating the absence of the right ICA. the CCA solely supplies the ECA and its branches. the left carotid injection (Panel B) demonstrates cross-flow across the anterior communicating artery to the contralateral side. Therefore, the right anterior cerebral artery is obvious almost through its entire extent. Panels C and D are CT head (bone windows), axial section. the hypoplasia of right carotid canal is obvious when compared with its normal counterpart (arrow).

sac.[16,17] In absence of the internal carotid artery, the basilar artery and posterior communicating arteries are enlarged and supply the anterior circulation.[13] The contralateral normal internal carotid artery may also contribute via the anterior communicating artery.[13] Other collateral pathways in absence of ICA have also been described.[16] The fetal pattern of collateral circulation from the anterior and posterior communicating arteries

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proved significant in this case because it enabled embolic strokes involving both hemispheres, following endovascular intervention.

GPAVFs have been associated with coexistent angiodysplastic disorders such as Cobb syndrome, hereditary hemorrhagic telangiectasia, Klippel-Trenaunay syndrome, and Osler-Weber-Rendu disease.[4–6] However, our patient did not have any associated anomalies on physical exam or MRI of the cervical spine to suggest coexistent disorders. Autopsy was declined by the patient's family.

Because of the rarity of its occurrence, a standard treatment remains to be established for cervical GPAVFs. Surgical, endovascular, or combination of the two treatments has been described with variable results.[5,6,12,18] The presence of large feeding arteries may be advantageous from treatment perspective, because it renders type III GPAVFs amenable to endovascular intervention.[8] Treatment with detachable balloons, coils, NBCA, Onyx, or a combination of these agents is possible.[18,19] Because of its unique "lava like" characteristics and nonadhesiveness, Onyx, alone or in combination with coils, may be the most suitable amongst current available agents to treat GPAVF.

The concurrence of GPAVF and aneurysm on a feeding artery or, GPAVF and ICA aplasia has previously not been reported. This case is presented for its unique anatomical attributes and their clinical implications.

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None

Author Justification

Paul E. Kaloostian MD: First Author, Involved in preparation of manuscript and research

Han Chen MD: Second Author, Involved in care of patient and research

Shah Naz-Khan MD: Second Author, Treating Attending, conception and design of manuscript, manuscript editing, critical revision, and final approval.

Christopher Taylor MD: Attending, Treating Attending

References

1. Aviv RI, Shad A, et al. 2004;Cervical dural arteriovenous fistulae manifesting as subarachnoid hemorrhage: report of two cases and literature review. *AJNR Am J Neuroradiol* 25(5):854–8.

2. Choh NA, Choh SA, et al. 2008;Right internal carotid artery agenesis. *Indian J Pediatr* 75(12):1269–71.

3. Bao YH, Ling F. 1997;Classification and therapeutic modalities of spinal vascular malformations in 80 patients. *Neurosurgery* 40(1): 75–81.

4. Biondi A, Merland JJ, et al. 1990;Embolization with particles in thoracic intramedullary arteriovenous malformations: long-term angiographic and clinical results. *Radiology* 177(3):651–8.

5. Mourier KL, Gobin YP, et al. 1993;Intradural perimedullary arteriovenous fistulae: results of surgical and endovascular treatment in a series of 35 cases. *Neurosurgery* 32(6):885–91.discussion 891

6. Ricolfi F, Gobin PY, et al. 1997;Giant perimedullary arteriovenous fistulas of the spine: clinical and radiologic features and endovascular treatment. *AJNR Am J Neuroradiol* 18(4):677–87.

7. Mascalchi MF, Scazzeri, et al. 1996;Dural arteriovenous fistula at the craniocervical junction with perimedullary venous drainage. *AJNR Am J Neuroradiol* 17(6):1137–41.

8. Sasamori T, Hida K, et al. 2008;Cervical perimedullary arteriovenous fistula in an infant presenting with subarachnoid hemorrhage--case report. *Neurol Med Chir (Tokyo)* 48(9):409–13.

9. Hayashi S, Arimoto T, et al. 1981;The association of intracranial aneurysms and arteriovenous malformation of the brain. Case report. *J Neurosurg* 55(6):971–5.

10. Shenkin HA, Spitz EB, et al. 1948;Physiologic studies of arteriovenous anomalies of the brain. *J Neurosurg* 5(2):165–72.

11. Gueguen B, Merland JJ, et al. 1987;Vascular malformations of the spinal cord: intrathecal perimedullary arteriovenous fistulas fed by medullary arteries. *Neurology* 37(6):969–79.

12. Mayumi O, M K. 2008Endovascular Treatment of Spinal Vascular Malformations. New YorkInforma Healthcare

13. Given CA 2nd, Huang-Hellinger F, et al. 2001;Congenital absence of the internal carotid artery: case reports and review of the collateral circulation. *AJNR Am J Neuroradiol* 22(10):1953–9.

14. Ta L. 1968Congenital Anomalies of the Carotid Arteries. AmsterdamExcerpta Medica

15. Teal JS, N M, Hasso AN. 1980Total agenesis of the internal carotid artery. *AJNR Am J Neuroradiol* :435–42.

16. Lie TA. 1968Congenital Anomalies of the Carotid Arteries. Amsterdam, Excerpta Medica

17. Quint DJ, Boulos RS, et al. 1989;Congenital absence of the cervical and petrous internal carotid artery with intercavernous anastomosis. *AJNR Am J Neuroradiol* 10(2):435–9.

18. Casasco A, Guimaraens L, et al. 2012;Endovascular treatment of cervical giant perimedullary arteriovenous fistulas. *Neurosurgery* 70(1):141–9.discussion 149

19. Mandzia JL, terBrugge KG, et al. 1999;Spinal cord arteriovenous malformations in two patients with hereditary hemorrhagic telangiectasia. *Childs Nerv Syst* 15(2–3):80–3.