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CASE REPORT

Two consecutive dural arteriovenous fistulae in a child: a case report of successful treatment with gamma knife radiosurgery

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Abstract
Introduction The occurrence of dural arteriovenous fistulae in children is quite rare. Endovascular embolization is typically the first line treatment. In general, Gamma Knife radiosurgery is used as adjuvant treatment and seldom performed as the first line treatment in children.
Discussion We report a case of a 27-month-old girl who presented with an initial dural arteriovenous fistula (AVF) located at anterior base of the left middle cranial fossa. She subsequently developed another dural AVF over the left transverse-sigmoid sinus region 2 years later.
Conclusion Both fistulae were successfully obliterated with Gamma Knife radiosurgery.

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Keywords Dural arteriovenous fistula · Gamma Knife · Radiosurgery · Embolization · Transverse-sigmoid sinus · Middle cranial fossa

Introduction

A dural arteriovenous fistula (AVF) is a vascular malformation which rarely occurs in the pediatric population [1]. Endovascular embolization is typically the first line of treatment for dural AVF. However, after embolization, a recurrent dural AVF adjacent or remote to the original fistula is occasionally observed because of recruitment of smaller arterial feeders or rerouting of venous outflow [2, 3]. Gamma Knife radiosurgery (GKRS), which causes progressive intimal hypertrophy of the vascular wall with subsequent thrombosis of the dural AVF, is considered to be a reasonable alternative treatment in pediatric patients who harbor dural AVF without concurrent life-threatening conditions such as heart failure or venous infarction [4–6]. GKRS affords obliteration of many dural AVF, but such obliteration typically occurs over 1 to 3 years after radiosurgery. Although there are reports of new dural AVF's after embolization, there are no known reports of this occurring after successful obliteration with GKRS [7–10]. We present the case of a 27-month-old girl who developed a subsequent transverse-sigmoid sinus fistula after successful Gamma Knife radiosurgery of an initial left middle cranial fossa dural fistula. In addition, the potential mechanism of development of this second dural AVF is addressed.

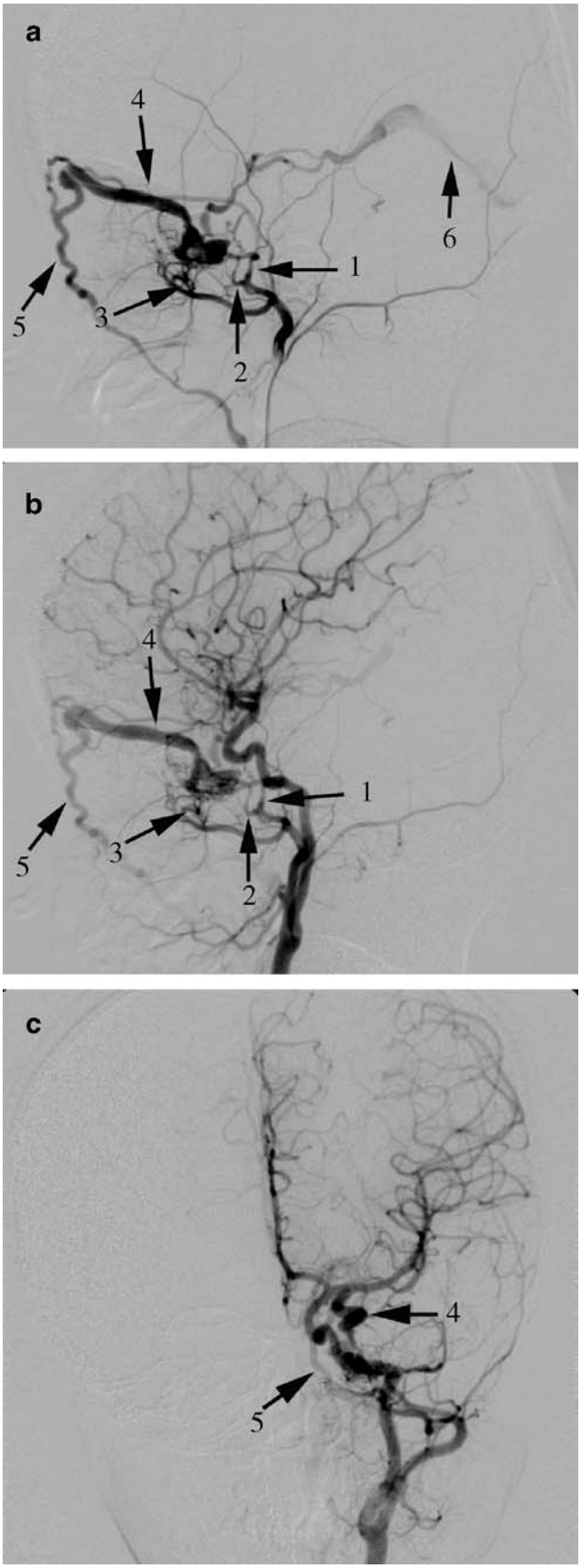

Fig. 1 **a** A 27-month-old girl presented with a protruding left eye and chemosis. She underwent a cerebral angiogram and a MRI/A study. The external carotid angiogram in lateral view shows a dural AVF located at anterior base of left middle cranial fossa. The fistula's feeding arteries included the accessory meningeal artery, middle meningeal artery, and artery of foramen rotundum. It drained into the left ophthalmic vein and straight sinus. **b** Left common carotid angiogram in lateral view demonstrated this fistula did not involve the cavernous sinus. **c** Left common carotid angiogram in anterior–posterior projection revealed this fistula was located at anterior base of the middle cranial fossa. *1* Middle meningeal artery, *2* accessory meningeal artery, *3* artery of rotundum, *4* superior ophthalmic vein, *5* facial vein, *6* straight sinus

Case report

A 27-month-old girl presented with a 2-month history of a protruding left eye and chemosis. The onset of these symptoms coincided with her having an upper respiratory infection. Physical examination revealed a ciliary injection, proptosis, and a bruit over left eye.

She underwent a cerebral angiogram which demonstrated a dural AVF over anterior base of left middle cranial fossa. The left external carotid angiogram revealed arterial feeders entering the fistula from the middle meningeal artery, accessory meningeal artery, and artery of the foramen rotundum. Venous drainage occurred via the left ophthalmic vein and straight sinus (Fig. 1).

In November of 2001, the patient underwent Gamma Knife radiosurgery of the dural AVF. The targeted volume was 0.2 cc, and this was treated to the 50% isodose configuration with a peripheral dose of 20 Gy (Fig. 2). The patient's symptom began to improve 2 months after the GKRS, and total symptomatic relief was achieved at 6 months post-operatively.

Unfortunately, the patient developed recurrence of chemosis in her left eye 2 years after her initial GKRS. As such, she underwent a cerebral angiogram, which confirmed the obliteration of the initial dural AVF but revealed the occurrence of another one located along the left transverse-sigmoid sinus. This new fistula had arterial feeders from left meningohypophyseal trunk, occipital artery, posterior auricle artery, the branch of posterior cerebral artery, vertebral artery, and its drainage was into the transverse and sigmoid sinuses. A magnetic resonance imaging (MRI) confirmed the location of this de novo left transverse-sigmoid sinus dural AVF (Fig. 3).

In October 2003, the patient underwent a second Gamma Knife radiosurgery for this new dural AVF. The treatment volume was 4.2 cc, and a peripheral dose of 20 Gy was

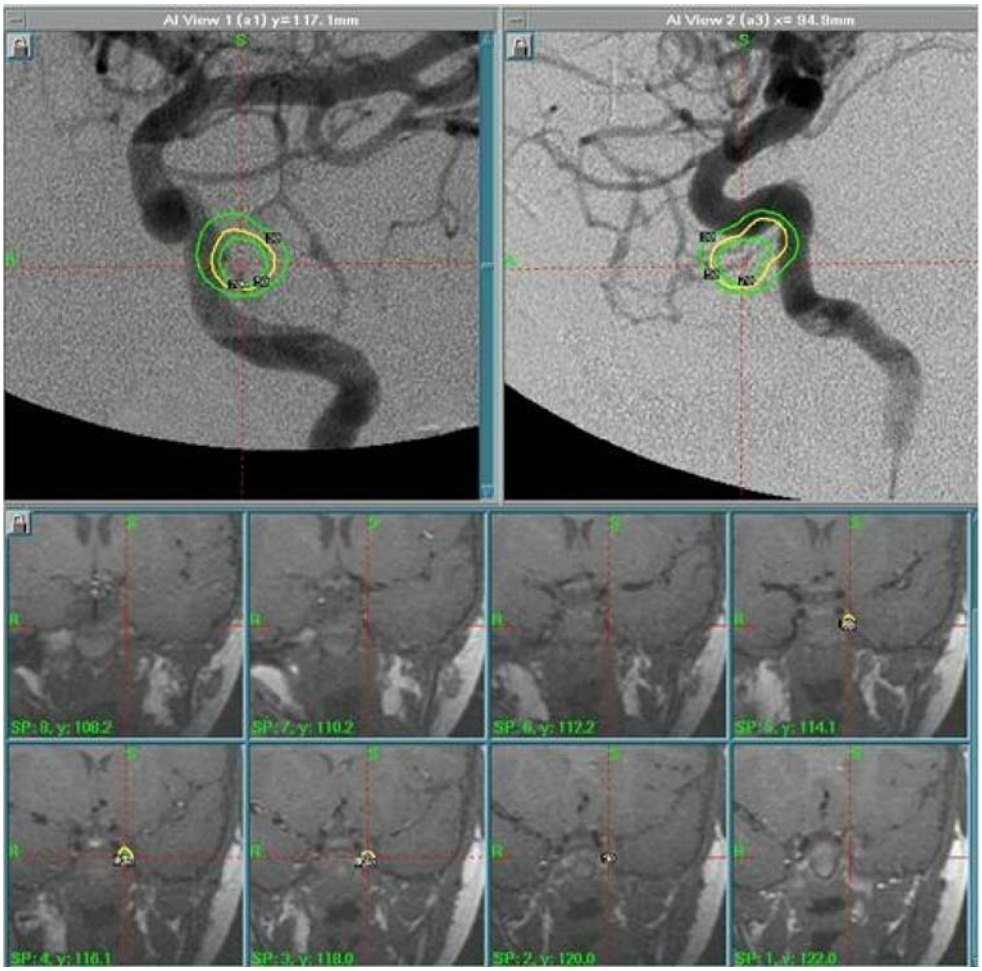

Fig. 2 The dose plan in the first Gamma Knife radiosurgery showed a treatment volume of 0.2 cc, and 20 Gy to the 50% isodose line was delivered

delivered to the 50% isodose line (Fig. 4). Again, the patient's symptoms resolved 6 months after her second GKRS. An angiogram performed 18 months after the second GKRS demonstrated complete obliteration of both fistulae (Fig. 5).

Discussion

There are few case reports of intracranial dural AVF presenting in pediatric patients. When occurring in children, dural AVF has a greater tendency to be multifocal. They may also demonstrate an aggressive clinical course including congestive heart failure or venous hypertension [1, 7].

The endovascular approach can immediately ameliorate symptoms and is typically the first line of treatment. Kincaid et al. [2] reported that 57% of dural AVF could be cured by an endovascular approach and that 28% of patients ultimately died due to uncontrolled intracranial pressure.

Gamma Knife radiosurgery is a reasonable treatment option for dural AVF's when immediate devascularization is not required. Pan and Chung [4] and Guo et al. [5] reported success obliteration rates of 70 and 90%, respectively, using Gamma Knife radiosurgery for transverse-sigmoid sinus and cavernous sinus dural AVFs. The choice of embolization, Gamma Knife radiosurgery, or a combination of the two in the treatment of a particular patient with a dural arteriovenous fistula should be made according to the clinical presentation and imaging findings.

In our patient who presented with proptosis and chemosis, angiograms disclosed the first dural AVF in anterior base of the middle cranial fossa and the subsequent one at the transverse-sigmoid sinus junction. Neither had evidence of cortical vein drainage or varices, and the absence of such drainage was often regarded as an ominous sign of venous hypertension [11]. Furthermore, the multiple feeding arteries for both dural arteriovenous fistulae suggested that total obliteration by transarterial embolization would be unlikely.

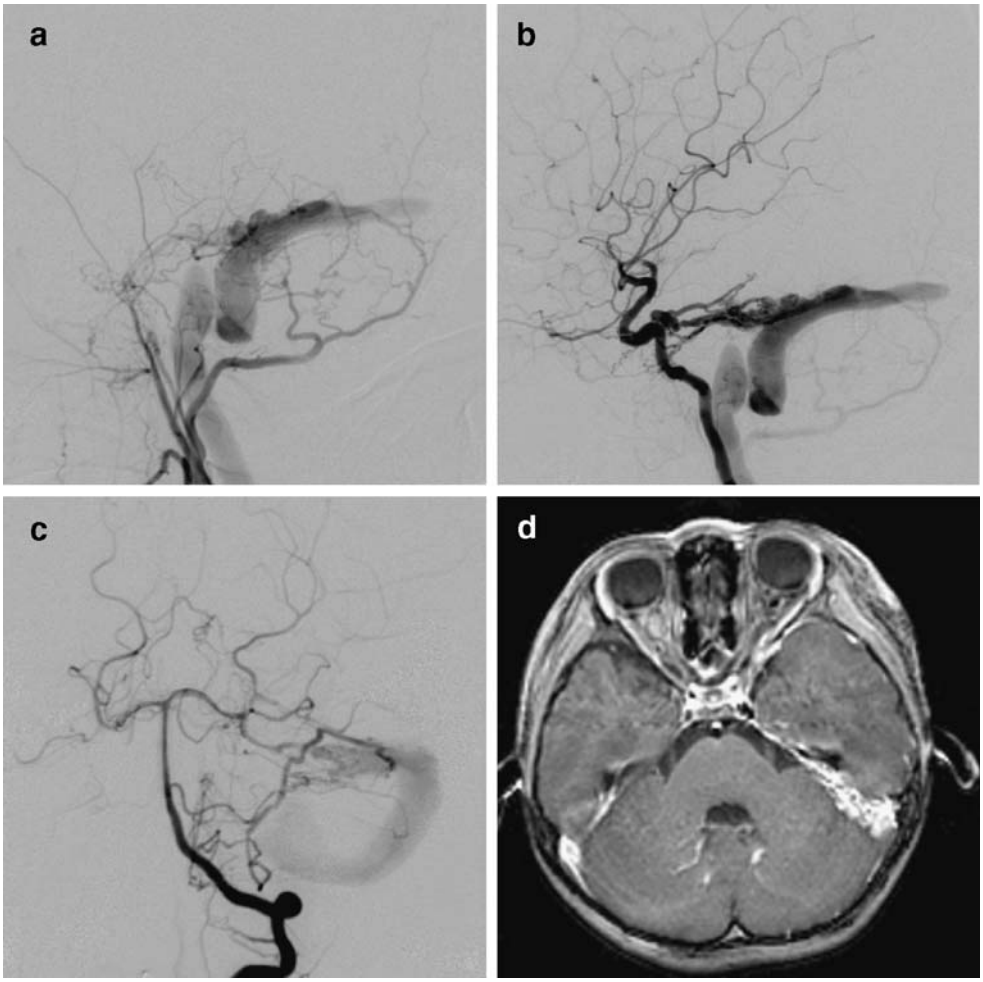

Fig. 3 **a** Twenty-four months after the initial Gamma Knife radiosurgery, a left external carotid angiogram revealed obliteration of the first fistula and the occurrence of another one situated at left transverse-sigmoid sinus junction. External carotid angiogram in lateral view shows the feeders from posterior auricle and occipital arteries. **b** The new fistula recruited the arterial supply from the meningohypophyseal trunk as demonstrated by this lateral view internal carotid angiogram. **c** This new fistula also had feeders from the posterior cerebral artery and meningeal artery of vertebral artery. **d** T1 weighted MRI showing the well-enhanced lesion over left transverse-sigmoid sinus

Some have advocated the possibility of transvenous embolization for dural AVFs. Due to the mild clinical symptoms of the patient, Gamma Knife radiosurgery was chosen as the first line treatment for both of her dural AVFs. Both fistulae were completely obliterated and all clinical symptoms resolved.

There are limited reports in the literature describing the occurrence of another dural AVF after total or partial occlusion of the first one by either transvenous or transarterial embolization [3, 8–10]. Several possible mechanisms described by Gupta et al. [10] could explain these phenomena. One theory is that the manipulation of venous anatomy with microcatheters may injure the vein, leading to sinus thrombosis, venous obstruction, and subsequent development of another dural AVF. A second possibility is that the entire architecture of a dural AVF was present but not visible at the time of angiography. Because subsequent embolization alters the venous outflow, the occult fistula might have become clinically and angiographically apparent. Furthermore, initial embolization can result in venous turbulence, thereby initiating thrombosis that can lead to development of an AVF as the thrombosis is recanalized.

The typical angiograms in these anecdotal reports showed that the second dural AVF involved branches of ECA draining into either the transverse sinus or jugular bulb. It is well known that the Gamma Knife radiosurgery usually takes at least 3 to 6 months to produce an effect on the carotid-cavernous or transverse-sigmoid sinus [4, 5]. Hence, a repeat angiogram shortly after the first GKRS was not performed. The first dural AVF in this case was located at anterior base of left middle cranial fossa with feeding arteries from the branches of the external carotid artery and draining into the ophthalmic vein and straight sinus. The new fistula's feeding artery was from the meningohypophyseal trunk, occipital artery, posterior auricle artery, a branch of the posterior cerebral artery, meningeal artery of the vertebral artery, and it drained into the transverse-sigmoid sinus junction. Obviously, part of this new fistula obtained feeding arteries from branches of the external carotid artery and drained into the transverse sinus. Such an occurrence is very

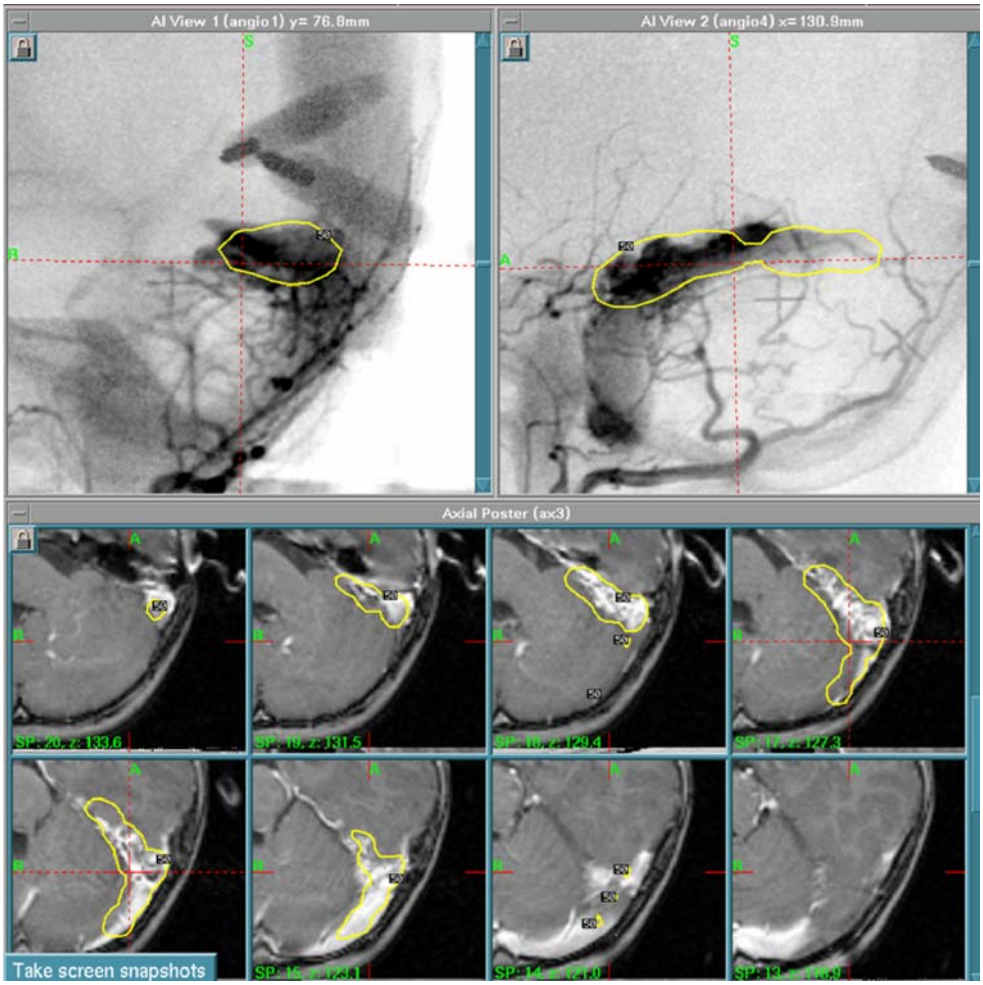

Fig. 4 The dose plan for the second Gamma Knife radiosurgery revealed a treatment volume of 4.2 cc. A peripheral dose of 20 Gy was delivered to the 50% isodose line

similar to the previous reports from the embolization literature. Typically, there is no immediate hemodynamic change observed in Gamma Knife radiosurgery. In our case, progressive obliteration of the first fistula likely redirected the venous return, thereby aggravating the flow to the transverse sinus and resulting in the development of the second fistula.

It is well known that Gamma knife radiosurgery cannot produce the immediate hemodynamic change in dural arteriovenous fistula. This raises the possibility that the second fistula comes from the existing pathological condition such as sinus thrombosis, coagulopathy, and high expression of angiogenic growth factors [12–15]. Based on the angiographic findings and laboratory data, sinus thrombosis and coagulopathy should be excluded as the possible reasons. Uranishi et al. [14] reported that surgically resected specimens from dural AVF demonstrated higher expression of angiogenic growth factors, such as basic fibroblast growth factors and vascular endothelial growth factor, than those in dural sinus wall of cadavers with unrelated central nervous system disease. They postulated that angiogenic growth factors produced in healing process of sinus thrombosis may participate in the genesis of dural AVF. Concluded from the possible predilection factors, endogenously produced angiogenic growth factors in sinus wall may also contribute to the development of second dura AVF after obliteration of the initial one.

Conclusion

Gamma Knife radiosurgery represents an important treatment modality for dural AVF. Just as with embolization, formation of another dural AVF may occur after successful obliteration of an initial one with GKRS. A new dural AVF may result from progressive hemodynamic changes accompanying the initial AVF obliteration.

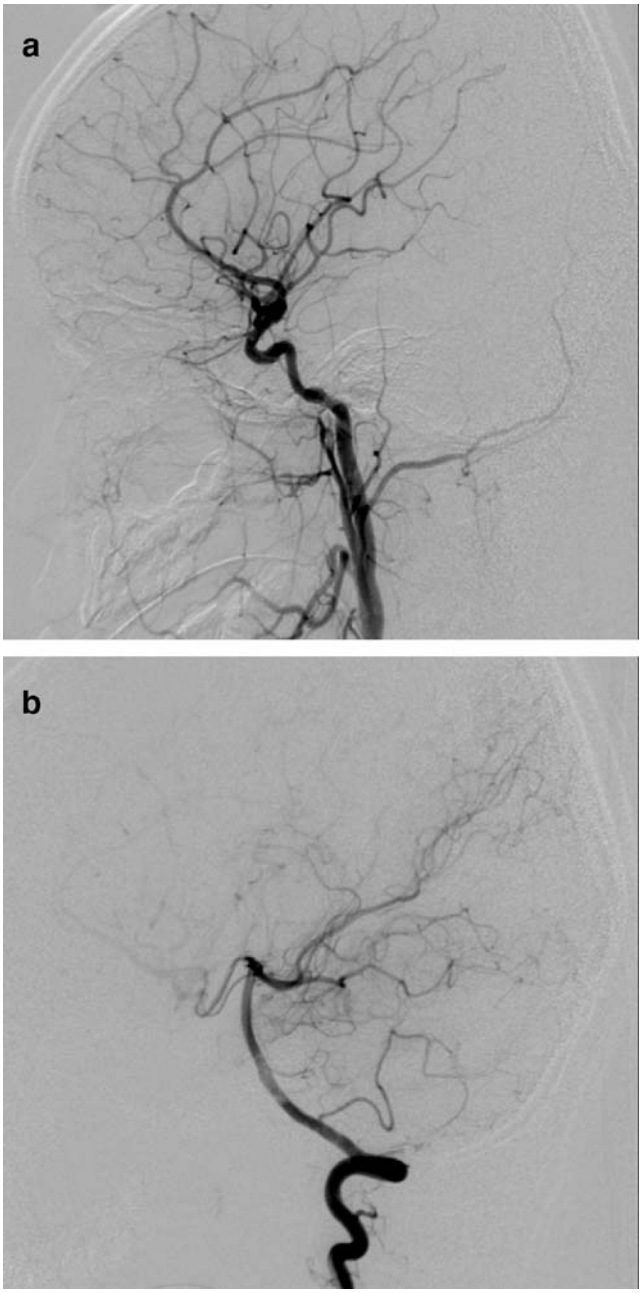

Fig. 5 **a** Left common carotid angiogram in lateral view demonstrates complete obliteration of the first and second fistulae. **b** The vertebral artery angiogram confirms obliteration of the second dural AVF

References

1. Lasjaunias P, Magufis A, Goulao R, Suthipongchai S, Rodesch R, Alvarez H (1996) Anatomical and clinical aspects of dural arteriovenous shunts in children. Interv Neuroradiol 2:179–191
2. Kincaid PK, Duckwiler GR, Gobin YP, Vinuela F (2001) Dural arteriovenous fistula in children: endovascular treatment and outcomes in seven cases. Am J Neuroradiol 22:1217–1225
3. Kiyosue H, Tanoue S, Okahara M, Yamashita M, Nagatomi H, Mori H (2000) Recurrence of dural arteriovenous fistula in another location after selective transvenous coil embolization: report of two cases. Am J Neuroradiol 23:689–692
4. Pan DHC, Chung WY (2002) Stereotactic radiosurgery for the treatment of dural arteriovenous fistula involving the transverse-sigmoid sinus. J Neurosurg 96:823–829
5. Guo WY, Pan DHC, Wu HM (1998) Radiosurgery as a treatment alternative for dural arteriovenous fistula of the cavernous sinus. Am J Neuroradiol 19:1081–1087
6. Payne BR, Prasad D, Steiner M, Bunge H, Steiner L (2000) Gamma surgery for vein of Galen malformation. J Neurosurg 93:229–236
7. Chaloupka J (1994) Endovascular therapy of dural arteriovenous fistulae. Semin Intervent Radiol 11:1–13
8. Nakaqawa H, Kubo S, Nakajima Y, Izumoto S, Fujita T (1992) Shifting of dural arteriovenous malformation from the cavernous sinus to the sigmoid sinus to the transverse sinus after transvenous embolization. Surg Neurol 37:30–38
9. Kubota Y, Ueda T, Kaku Y, Sakai N (1999) Development of a dural arteriovenous fistula around the jugular valve after transvenous embolization of cavernous dural arteriovenous fistula. Surg Neurol 51:174–176
10. Gupta R, Horowitz M, Tayal A, Jovin T (2005) De novo development of a remote arteriovenous fistula following transarterial embolization of a carotid cavernous fistula: case report and review of literature. Am J Neuroradiol 26:2587–2590
11. Halbach VV, Hieshima GB, Higashida RT, Reicher M (1987) Carotid cavernous fistulae: indication for urgent treatment. Am J Neuroradiol 8:627–633
12. Kutluk K, Schumacher M, Mironov (1991) The role of sinus thrombosis in occipital dural arteriovenous malformation-development and spontaneous closure. Neruochirurgica (Stutt) 34:144–147
13. Martinelli I, Rosendaal FR, Vandenbrocke JP, Mannucci PM (1996) Oral contraceptive are a risk factor for cerebral vein thrombosis (letter and comment). Thromb Haemost 76:477–478
14. Uranishi R, Nakase H, Sakaki T (1999) Expression of angiogenic growth factors in dural arteriovenous fistula. J Neurosurg 91:781–786
15. Koizumi T, Shiraishi T, Hagihara N, Tabuchi K, Hayashi T, Kawano T (2002) Expression of vascular endothelial growth factors and their receptors in and around intracranial arteriovenous malformation. Neurosurgery 50:117–126