Cerebral arteriovenous malformation (CAVM) associated with arteriovenous fistula (AVF) is rare. It may be difficult to identify hemodynamic details of mixed CAVM and AVF, even when using x-ray cerebral angiography (digital subtraction angiography). We report on a 37-year-old male patient with headache that led to an initial diagnosis of deep frontotemporal CAVM. The first DSA revealed engorged, tortuous, and high-flow venous drainage in addition to clusters of vasculature niduses. The patient was initially treated using γ-knife radiosurgery (GKRS), which resulted in partial nidus obliteration, documented by a series of follow-up magnetic resonance imaging (MRI). However, the high-flow venous drainage remained, seen on MRI as engorged venous pouches. Clinically, the patient was bothered by persistent headache and bruits after GKRS. Follow-up DSA 3 years after GKRS confirmed a small remnant CAVM nidus and a nearby AVF, separated from and lateral to the original CAVM nidus in the ipsilateral deep temporal lobe. When the initial DSA was reviewed, it revealed that the AVF was difficult to define because of superimposition of the nidus and engorged drainage vessels. Embolization of the AVF using electrodetachable coils resulted in total occlusion of the AVF. The patient’s symptoms resolved immediately after embolization. This case suggests that superselective angiography using a microcatheter may be necessary for the initial diagnosis of CAVM associated with AVF with high-flow and engorged venous drainage. For CAVM patients with persistent symptoms after radiosurgery and engorged venous drainage when CAVM is expected to be cured, a microcatheter and superselective endovascular approach may offer diagnosis. Immediate embolization for associated AVF in the same angiographic session may thereby improve neurologic deficits and reduce hemorrhagic risk during the latency after GKRS.

**Key Words:** arteriovenous malformation, arteriovenous fistula, embolization, radiosurgery

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The goal of treatment for cerebral arteriovenous malformation (CAVM) is to completely obliterate the CAVM and normalize both cerebral vascular anatomy and hemodynamics. Microsurgery, embolization, and radiosurgery are the therapeutic strategies currently used for CAVM. γ-Knife radiosurgery (GKRS) is effective, with a cure rate of 60–85% at 2 years [1–3]. Incomplete cure of GKRS-treated CAVM, regardless of the percentage of obliteration, is classified as unsuccessful treatment. Patients with incomplete cure of CAVM have similar survival rates as untreated patients. The risk of repeat hemorrhage remains, although the re-bleeding rate is lower in partially obliterated CAVM. Failed CAVM GKRS is multifactorial [4,5], and should failure occur, efforts to find possible underlying causes should be undertaken. If feasible, a second GKRS or endovascular treatment is offered for the residual CAVM.
Combined endovascular treatment and GKRS for associated arteriovenous fistula (AVF) and CAVM has not been previously reported. Herein, we report a rare case, where the CAVM nidus and engorged venous drainage were superimposed on the associated AVF before GKRS, masking the AVF. Endovascular treatment for the AVF was performed 3 years after GKRS.

CASE PRESENTATION

A 35-year-old male, who had a 13-year history of headache and bruit over his left temporal region, was admitted to hospital on March 20, 2003. He had been admitted because of deterioration in his clinical symptoms on July 3, 1998. Left temporal CAVM was found during that admission. There was neither a family history of CAVM nor a history of seizures, hemiparesis, or symptoms suggestive of intracranial hemorrhage. Clinical examination disclosed an impaired visual field with right hemianopsia. Brain computed tomography and magnetic resonance imaging (MRI) revealed multiple tortuous and engorged vessels over the left temporal and basal ganglion regions, as well as dilated cortical veins and superficial venous system, leading to an initial diagnosis of CAVM. Cerebral angiography (digital subtraction angiography; DSA) disclosed a CAVM in the left temporal and basal ganglion regions, fed by temporal branches of the left middle cerebral artery (MCA) and lenticulostriate arteries with early venous drainage into the frontal cortical veins and vein of Labbe (Figure 1). There also were tortuous venous pouches. The CAVM was treated using GKRS with irradiation doses of 30.91 Gy at the target center and 17 Gy at the periphery. The volume of the CAVM was calculated to be 7.4 mL and

Figure 1. (A, B) Initial left carotid angiograms reveal a cerebral arteriovenous malformation in the left temporal and basal ganglion (arrowheads), supplied by the lenticulostriate arteries and draining into the frontal cortical vein and vein of Labbe. Engorged and tortuous venous outflow with venous ectasia are present as well.
the radiation volume was 9 mL. After GKRS, regular clinical and MRI follow-up at 6-month intervals revealed progressive obliteration of the CAVM, but clinical symptoms persisted. The last MRI revealed shrinkage of the CAVM nidus, but tortuous venous pouches remained in the left temporal lobe. Because of persistent symptoms, DSA was performed and revealed a small residual CAVM in the left basal ganglion and an AVF in the ipsilateral temporal lobe, supplied by the temporal branch of the MCA (Figure 2). It drained primarily into the frontal cortical vein and joined the superior sagittal sinus. Secondary drainage took place through the prominent vein of Labbe and later into the transverse sinus. Several tortuous venous pouches were demonstrated, as in the initial DSA. Embolization of the AVF was performed under general anesthesia using a femoral approach; a Tracker-18 microcatheter (Boston Scientific, Fremont, CA, USA) was introduced and easily advanced into the fistula site. Electrodetachable coils were selected because of the potential risk of gluing nearby normal branches of the MCA, and probable distal migration of liquid adhesive to the venous side in this high-flow shunt. Five coils with a total length of 110 cm were lodged in the venous pouch just beyond the fistula site, resulting in total occlusion of the shunt flow and reduction of the venous hypertension (Figure 3). A small residual CAVM remained in the left basal ganglion. The pre- and post-procedure courses were smooth. Follow-up MRI and DSA 1 week later confirmed complete occlusion of the AVF, as well as thrombosis in the venous pouches. The patient recovered uneventfully. His headache and bruit resolved 2 weeks after embolization. His small residual CAVM was treated by a second session of GKRS. At the time of writing, the residual CAVM has further regressed to a tiny remnant.

Figure 2. (A, B) Follow-up angiograms demonstrate occlusion of most of the cerebral arteriovenous malformation 3 years after γ-knife radiosurgery. An arteriovenous fistula is disclosed in the left temporal region (arrow), with prominent draining veins.
DISCUSSION

Embolization, microsurgery, and GKRS are established treatments for CAVM. Embolization alone has little chance of completely occluding a CAVM, unless the lesion is small and/or has few feeding vessels. Combining embolization with microsurgery or GKRS is promising. The rationale for combined treatments is that the CAVM is reduced in size so that the residual nidus can be irradiated with a better cure rate and fewer side effects; the unstable weak points of the CAVM, such as flow-related and dysplastic arterial as well as intranidal aneurysms, can be selectively occluded so that the risk of bleeding in the time lag between radiosurgery and cure decreases; and the associated AVF of a plexiform CAVM can be occluded by embolization either before or after GKRS [6,7].

Pre-therapeutic detailed analysis of the angio-architecture, particularly the maximum diameter, location, and angiographic shape of the CAVM nidus, intranidus aneurysm, and any associated AVF, is important to predict each patient’s prognosis and the response to GKRS. In general, smaller and compact CAVMs seem to respond better, and high-flow CAVMs and/or fistulas seem to respond less to GKRS. Most hemodynamic information is provided by DSA. However, in the absence of microcatheter superselective angiography (MSA), a number of associated vascular or unstable lesions probably go undetected because of superimposition by CAVM, or by engorged and tortuous draining veins. To detect associated AVF and to facilitate the success of subsequent GKRS, Pollock et al used MSA as part of the pre-therapeutic evaluation [5]. MSA can clarify the presence of dysplastic aneurysms in feeding vessels,

Figure 3. (A, B) Endovascular embolization was performed with a microcatheter positioned at the fistula site and a total of four electrodetachable coils placed in the venous pouch just beyond the fistula site (arrowhead). Control angiograms document total occlusion of the AVF with significant reduction of the venous hypertension. A small residual cerebral arteriovenous malformation remains in the left basal ganglion.
differentiate intranidal aneurysms from venous ectasia, and delineate any intranidal AVF. The second advantage of MSA is the feasibility of immediate treatment of intranidus aneurysms and/or AVF in the same angiographic session with an acceptable low risk of morbidity and mortality. Nevertheless, MSA is time-consuming and more costly than regular diagnostic DSA. In the current case, the associated AVF was only diagnosed 3 years after GKRS, when most of the CAVM was obliterated. It was initially overlooked mainly because of superimposition by the nidus and nearby engorged drainage vessels. The exact incidence of CAVM associated with AVF in the same anatomic location remains unknown.

Gallina et al reported a series of 120 patients with CAVM who underwent radiosurgery; complete obliteration was not possible in eight patients due to intranidal high-flow shunts [4]. It seems reasonable to assume that 6.7% of CAVMs are associated with AVF. Nevertheless, multiple CAVM/AVF with intervening normal brain tissue is rare, and this relation is mostly documented in reports on patients with known hereditary hemorrhagic telangiectasis, Wyburn-Mason disease, or other systemic CAVM [8,9]. In a series of 203 CAVM patients, only 18 (9%) had multiple CAVM [10]. The incidence of multiple CAVM/AVF is likely to have been underestimated, due to the failure to recognize small AVFs associated with large CAVMs.

The treatment goal in patients with these multiple lesions is total obliteration of all CAVM/AVF. However, this is not always possible using a single treatment modality, such as surgical removal, GKRS, or endovascular embolization. Each modality has its own unique, inherent problems, so a combination seems a promising solution. Surgically accessible lesions can be removed by microsurgery. Medium to large plexiform deep-seated CAVMs are managed by combined embolization and radiosurgery with a reasonable risk of complications.

The selection of embolic agents is crucial and depends on the anatomic location and flow characteristics of the fistula. A permanent occlusion agent is preferred. N-butyl-2-cyanoacrylate (NBCA), a liquid adhesive embolic agent, is widely used by neurointerventionalists for occlusion of CAVM/AVF, with good results. However, it is dangerous when used alone for high-flow fistulas like the one in the current case, because of the risk that distal migration of NBCA through the shunts may cause distal venous occlusion. The other disadvantage is that unintentional gluing of the nearby normal branches of the MCA can result in ischemic stroke. Stainless steel coils and platinum microcoils are another option. They have been successfully used for occluding AVF. The coils work by creating a thrombosis, but they may also migrate through the fistula if they are not the correct size. The risk of involuntary migration can be diminished by using electrodetachable coils, as illustrated in the current case.

In conclusion, CAVM associated with AVF may not be discerned even in a thorough global angiographic workup. This is due to superimposition of the CAVM and its engorged draining veins. MSA is important in the pre-GKRS imaging workup for CAVM with engorged, tortuous venous pouches, as well as a high-flow venous shunt. In post-GKRS follow up, if clinical symptoms persist or even deteriorate after partial obliteration of the CAVM, the possibility of an associated AVF should be suspected. A partially obliterated CAVM will, sometimes, make the detection of associated AVF easier. Furthermore, MSA has the advantage of offering detailed angioarchitecture of the CAVM and may allow the immediate treatment of intranidus aneurysms or AVF with an acceptable low risk of morbidity and mortality, thereby reducing the risk of neurologic deficit and hemorrhage during the latency after GKRS.

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REFERENCES


以血管內栓塞治療多發性腦動靜脈畸形
經加馬刀放射手術後的殘餘動靜脈瘻管

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多發性腦動靜脈畸形或動靜脈瘻管非常少見，這罕見的多發性血管畸形即使以血管攝影有時也不易同時診斷。本文報告一例因頭痛及頭部雜音求診的37歲男性患者，經檢查診斷患左側顯著腦動靜脈畸形合併擴大彎曲及快速的靜脈血流。此腦動靜脈畸形接受加馬刀放射手術治療，術後3年追蹤期間磁振造影顯示動靜脈畸形逐漸變小，但擴大彎曲的快速靜脈血流仍在，且病患症狀並未獲得明顯改善。此時的追蹤血管攝影發現另一動靜脈瘻管於同側大腦顱葉深部，三年前此瘻管因與擴大彎曲的靜脈血管及腦動靜脈畸形重疊而被忽略。針對此動靜脈瘻管我們使用可分離式線圈銅血管內栓塞治療成功地將此瘻管堵住，病患並獲得立即症狀緩解。由此例經驗顯見有時使用微小導管做超選擇血管攝影是必須的，它有利於檢視瘻管是否與腦動靜脈畸形並存，特別是針對腦動靜脈畸形合併擴大彎曲及快速靜脈血流及部位治療後症狀仍持續的患者，尤其診療價值。此外以微小導管做超選擇血管攝影的同時，如因病情需要，亦能提供立即栓塞治療腦動靜脈畸形的瘻管部分；之後以加馬刀放射手術治療剩餘的動靜脈畸形部分，應可加速放射手術療效及改善神經症狀並減少治療後的出血機會。

關鍵詞：動靜脈畸形，動靜脈瘻管，栓塞，放射外科

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