The association of an adult pial AVM and high-grade stenosis of the SSS is unusual. Adult pial high-flow AVMs are uncommon, observed in only 2.4% of Yaşargil’s surgical series of 419 brain AVMs. When anomalies of the major sinuses (typically agenesis and stenosis or occlusion) are associated with pial AVMs, however, they are usually observed in vein of Galen AVMs, and large parietal, occipital, or posterior fossa high-flow AVMs. Although the pathogenesis of these associated venous anomalies is unknown, when present, they typically involve the straight, transverse, and sigmoid sinuses. Acquired high-flow arteriopathy and venopathy have been described as possible causes of arterial and venous stenoses that are associated with brain AVMs. In the present case we documented chronological angiographic findings that support the hypothesis that high-flow venopathy may be the cause of associated major sinus stenosis or occlusion in certain pial AVMs. The stenosis of the posterior SSS suggests that atypical locations distal to the AVM nidus can be involved. In addition to discussing the implications that this case has for our understanding of morphological and hemodynamic changes in the venous system of brain AVMs, we describe the technical and clinical details of angioplasty and stent placement for the treatment of severe cortical venous hypertension caused by nonthrombotic SSS stenosis.

Case Report

History. Five years ago, this 69-year-old man suffered a Hunt and Hess Grade III subarachnoid hemorrhage from a ruptured anterior communicating artery aneurysm and underwent craniotomy for aneurysm clip ligation. During his hospitalization he was also found to have a high-flow left parietal pial AVM with superficial venous drainage into a patent SSS (Fig. 1). The patient’s family declined treatment of the AVM. After rehabilitation, the man was able to function independently, despite short-term memory deficit and mild diplopia. For several months before his recent admission, he experienced seizures and progressive mental decline without focal neurological deficit and was sent to a skilled nursing facility where he eventually became unresponsive.
Examination. On admission, the patient received intubation and ventilation therapy. He displayed no spontaneous eye or extremity movement and did not respond to voice. He grimaced in response to painful stimuli but did not open his eyes, and he slightly flexed his upper extremities, the left more than the right, and withdrew his lower extremities. The patient’s brainstem reflexes were intact. He displayed hyperreflexia, more so on the right side than on the left. Computerized tomography scans did not reveal any intracranial hemorrhage, hydrocephalus, or infarcts. Magnetic resonance imaging was contraindicated because the patient had a pacemaker. Cerebral angiograms revealed the presence of a high-flow AVM in the left parietal region with superior superficial cortical venous drainage (Fig. 2). In addition, focal severe stenosis of the posterior SSS was present, with evidence of severe bilateral cortical venous hypertension (Fig. 3). This was manifested on the cerebral angiogram as a delayed arteriovenous transit time, a prolonged capillary phase, and an irregular and retrograde filling of bilateral cortical veins that eventually drained into the cavernous sinus. An examination of cerebral angiograms obtained in this patient at the time of his previous hospitalization showed that the venous hypertension and focal high-grade stenosis of the posterior SSS, located away from the AVM nidus and away from the entrance of the dominant superficial cortical draining vein into the SSS, were not present 5 years previously.

Operation. The large single-hole direct middle cerebral artery fistula was embolized with coils and acrylic. The patient’s neurological status remained unchanged after embolization. Four days later, angioplasty and stent placement (Fig. 4) in the SSS stenosis was performed, as well as additional embolization of the left parietal AVM with acrylic. A 6 × 13-mm stent (Corinthian High Q stent; Cordis Neuro-
vascular, Miami Lakes, FL) premounted on a balloon catheter (Jupiter; Cordis Neurovascular) was delivered via a retrograde left internal jugular vein approach and was inflated to 8 atm (nominal pressure 12 atm) for 30 seconds. Postembolization angiography revealed a significantly improved arteriovenous circulation time. Routine follow-up head computerized tomography scanning revealed a left parietal ICH adjacent to the AVM nidus. Based on a careful retrospective analysis of the cerebral angiograms obtained in this patient, particularly with respect to the mass effect on perinidal vessels, we concluded that the ICH had occurred after the first embolization and not during the SSS venous angioplasty and stent placement.

Postoperative Course. After the procedure, the patient’s neurological status improved to slight eye opening and weak localization of his upper extremities to painful stimuli. Subsequently, he deteriorated neurologically. His family decided to withdraw care and the patient died 2 weeks later. The family declined the request for autopsy.

Discussion

Morphological and hemodynamic changes in the venous system of brain AVMs are complex and remain incompletely understood, despite their important clinical and surgical implications. Despite their low incidence, large high-flow parietal, occipital, and posterior fossa AVMs can be associated with stenotic or occlusive anomalies of major sinuses, typically involving the straight, transverse, or sigmoid sinus. Our case is unusual in that a superficial pial high-flow AVM was associated with a distal focal severe
SSS stenosis that developed within 5 years and caused morbid cortical venous hypertension.

The cause and pathogenesis of major sinus occlusive disease associated with pial AVMs are not well established. In an angiographic analysis of the nidus and venous system in 53 patients with deep pial AVMs, Viñuela, et al.,14 found major venous wall irregularities or stenoses of the galenic venous system in 14 cases. They postulated that the angiographic changes were related to local endothelial damage secondary to "turbulent, unstable, and irregular flow," a theory supported by other authors.6,9,14 Duckwiler and colleagues3 described delayed venous occlusion that developed 15 months after embolization of a vein of Galen malformation in a newborn. These authors theorized that the progressive venous occlusion was due to high-flow venopathy characterized by hyperplasia of vessel wall elements, presumably as a result of shear forces related to turbulent arterIALIZED venous flow, which has been observed in animal models.10,12 In support of their hypothesis, autopsy findings revealed smooth-muscle cell hyperplasia of stenotic transverse and sigmoid sinuses as the primary pathological change.3

An angiographic documentation of the development of focal high-grade SSS stenosis within 5 years in our patient with a superficial high-flow pial AVM supports the hypothesis that high-flow venopathy may be the cause of associated major sinus stenosis or occlusion and that these lesions are acquired rather than congenital or developmental. We can infer from the stenosis of the posterior SSS that atypical locations distal to the AVM nidus can be involved, possibly related to local areas of turbulent arterIALIZED venous flow caused by normal inflow of adjacent cortical veins. Flow in the SSS anterior to the area of stenosis may have been laminar. High flow and associated shear stress may alter endothelial production of nitrous oxide and release of cytokines. This can affect local production of tissue-type plasminogen activator,1,4,5,7 which may, in part, explain the patency of the SSS adjacent to the AVM nidus and in the segment more proximal to the site of stenosis.

It is unknown if any metabolic, endocrinological, or hemodynamic factors may have triggered this relatively abrupt venoocclusive event or, given the development of stenosis within 5 years, why this progressive occlusive venopathy did not occur earlier in life. Theoretically, de novo focal thrombosis formation and partial recanalization within an SSS may alternatively account for the angiographic features; however, we considered this unlikely. Thrombosis of the SSS is associated with a wide variety of disorders including infections, forms of vasculitis, and hypercoagulation states,11 but it has not been associated with a high-flow state within the SSS and has not been reported as a focal disease process. Moreover, during angioplasty no evidence of thrombus was noted, and a high inflation pressure was necessary to dilate the stenotic SSS mechanically to its normal caliber. Thrombus formation in the SSS anterior to the area of stenosis may have possibly related to local areas of turbulent arterialized venous flow.

Stent placement for stenosis or occlusion of an intracranial dural sinus has been rarely reported and only in the transverse, sigmoid, and occipital venous sinuses.7,8 We describe the first case in which mechanical dilation with balloon angioplasty and stent placement in the SSS was performed for the treatment of morbid diffuse cortical venous hypertension. Nevertheless, given our patient’s poor outcome and the anticipated high risk of thrombosis and restenosis in a low-flow state, the indications for and efficacy of intracranial venous stent placement remain uncertain.