Tortuous, Engorged Pial Veins in Intracranial Dural Arteriovenous Fistulas: Correlations with Presentation, Location, and MR Findings in 122 Patients

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BACKGROUND AND PURPOSE: Tortuous, engorged veins can be identified on the venous phase of the brain circulation in patients with venous congestion related to an intracranial dural arteriovenous fistula (DAVF). The term pseudophlebitic pattern (PPP) has been used to describe this finding. The purpose of this study was to determine the prevalence of PPP in patients with intracranial DAVF and to analyze the relationship of this sign to presentation, location of the fistula, presence of retrograde leptomeningeal venous drainage, and MR findings.

METHODS: We retrospectively reviewed the charts and imaging findings of 122 patients with intracranial DAVF. In 122 patients the venous phase of the brain circulation was adequately assessed. The PPP was graded as mild, moderate, or severe.

RESULTS: PPP was found in 51 patients (42%). Thirty-two (73%) of the 44 patients who had a hemorrhage, neurologic deficit, or seizure had PPP as compared with 16 (21%) of the 75 who had a bruit or orbital signs. The three patients with either congestive heart failure or increasing head circumference had PPP. Fourteen (88%) of the 16 who had fistula of the superior sagittal sinus, straight sinus, or superior petrosal sinus had PPP. PPP was seen in 46 (81%) of 57 patients who had retrograde leptomeningeal venous drainage and in five (8%) of the 65 who had only sinosal drainage. Fourteen (88%) of the 16 who had white matter T2 hyperintensity on MR images had severe PPP.

CONCLUSION: The PPP reflects venous congestion and is associated with an aggressive presentation with or without retrograde leptomeningeal venous drainage. PPP may be a useful prognostic indicator and should be considered in treatment decisions.
cumference. Hemorrhage was either subdural, intracerebral, or subarachnoid; some resulted in a neurologic deficit. Deficit included a transient or progressive nonhemorrhagic neurologic deficit, excluding cranial nerve palsies related to the cavernous sinus. Bruit included tinnitus and chronic headaches; patients who were asymptomatic at presentation were also grouped into this category. Orbital signs included proptosis, chemosis, conjunctival injection, and a cavernous sinus syndrome. Congestive heart failure/increasing head circumference was found in 27% of the patients with cavernous sinus DAVFs. Table 3 highlights the relationship between presentation and location of PPP and presentation, RLVD, and location of the intracranial DAVF, respectively. Table 1 shows that 32 (73%) of the 44 patients who presented with a hemorrhage, deficit, or seizure had PPP. In contrast, 16 (21%) of the 75 patients who presented with orbital symptoms or bruit had PPP. All three patients with congestive heart failure or an enlarging head had PPP. Table 1 shows that presentation with a deficit or hemorrhage accounted for 13 (93%) of the 14 adult patients with severe PPP. Table 2 shows that nine (22%) of 41 patients with cavernous sinus DAVFs had PPP, whereas 14 (88%) of 16 patients with superior sagittal, superior petrosal, or straight sinus locations had PPP. RLVD was found in 27% of the patients with cavernous sinus DAVFs, compared with 100% of those with ten- torial, anterior cranial fossa, superior petrosal, and straight sinus DAVFs. Table 3 highlights the relationship between presentation and location of PPP in five patients who did not have RLVD.

MR imaging was done in 52 patients, 42 of whom had prominent pial vessels (Fig 2). Thirty-eight of the 42 patients had RLVD and 31 had PPP. PPP was present in two of the four patients who had prominent pial vessels but no RLVD. MR findings were normal in two patients with RLVD. Hy-
drocephalus and hindbrain herniation were evident on MR images in four adults and one child, four of whom had severe PPP. The four adults presented with a deficit, the child with increasing head circumference. MR images showed hyperintensity on T2-weighted sequences in 11 patients, 10 of whom presented with a deficit. Ten of these 11 had PPP, and in six it was severe.

Seventy-four patients were treated either by embolization alone (n = 32), by embolization and surgery (n = 23), or by surgery alone (n = 19). In 51 patients, follow-up angiography within 1 month of treatment showed no change in the PPP. Angiography showed less marked PPP in 11 of the 17 patients who had follow-up studies more than 6 months after treatment (Fig 3).
In patients with intracranial DAVFs, RLVD and venous ectasia were first associated with intracranial hemorrhage by Houser et al (7) in 1972 and was later supported by Castaigne et al (8). In 1984, a metaanalysis by Malik et al (9) found RLVD, venous ectasia, and lesion location “outside a major sinus” to be associated with intracranial hemorrhage on presentation. In 1986, Lasjaunias et al (10) showed that focal neurologic deficits were related to the territory of the venous reflux. In 1987, Ishii et al (5) identified a subgroup of patients who were at high risk for hemorrhage and dementia because of venous overload exacerbated by occlusive changes in the transverse sinus. In their report, Ishii et al noted that engorged pial veins on angiography were related to the venous overload. Awad et al (11), in a review of 360 cases from the literature prior to 1990, found that RLVD, venous ectasia, and vein of Galen drainage correlated with intracranial hemorrhage and nonhemorrhagic neurologic deficit at presentation. In 1993, Lalwani et al (12) developed a grading system for transverse/sigmoid sinus DAVFs based on the severity of the venous restrictive disease.

A comprehensive classification of intracranial DAVFs based on angioarchitecture was first proposed by Djindjian et al (13) in 1977. In 1995, this scheme was modified by Cognard et al (14), who, in a review of their own series of 205 patients, were able to show a relationship between type of intracranial DAVF and presentation. A similar but simplified version of this classification was proposed by Borden et al (15). In 1996, Davies et al (16) confirmed the validity of the Borden and Cognard classification systems by showing a highly significant correlation with presentation.

The presentation of benign intracranial DAVFs often relates to the location of the fistula, and includes tinnitus, cranial nerve palsies, and/or signs related to venous congestion in the orbit (1, 11). Intracranial DAVFs with RLVD may have a similar presentation to the benign type or may present with an intracranial hemorrhage, neurologic deficit, or seizure (2, 5, 10–12, 17–21). Direct RLVD has a greater prevalence of hemorrhage as compared with sinosal drainage with RLVD (14, 22). Aneurysmal pial venous drainage has a greater prevalence of hemorrhage on presentation than does such drainage without aneurysmal enlargement (11, 14, 17). Sinosal drainage with retrograde flow in the venous sinuses but without RLVD can result in raised intracranial pressure (14, 23–25). The study by Davies et al (1, 2) of the natural history of intracranial DAVFs confirmed the benign course of those with sinosal drainage only and the aggressive course of those with RLVD.

The pathophysiology of nonhemorrhagic neurologic deficits and seizures in intracranial DAVFs relates to venous congestion (5, 6, 10, 19, 23). Venous congestion may be evident on MR images as
bilateral or unilateral diffuse T2 hyperintensity in the white matter of either the cerebral or cerebellar hemispheres. These MR findings may be partially reversible after treatment. Thalamic and brain stem venous congestion can result from these fistulas refluxing into the straight sinus and deep veins. This clinicopathologic picture has been referred to as venous congestive encephalopathy. PPP represents a response to venous congestion with the development of enlarged, tortuous collateral veins and venous rerouting. Rerouting may be into dilated transosseous venous channels or may be retrograde into the orbital veins.

Many reports have noted an association between presentation and location of intracranial DAVFs. An aggressive neurologic course is seen least often with transverse/sigmoid and cavernous sinus locations and most often with tentorial DAVFs. This corresponds to the frequency with which we found RLVD in each location (Table 2). RLVD was present in 100% of our tentorial, anterior cranial fossa, superior petrosal, and straight sinus locations. Cavernous and transverse sinus/torcular DAVFs were associated with RLVD in 27% and 42% of patients, respectively. A similar proportion of our patients had RLVD and PPP at each location (Table 2). Fourteen (88%) of our 16 patients with DAVFs located within the superior sagittal, straight, and superior petrosal sinuses had PPP, as compared with nine (22%) of 41 patients with cavernous sinus DAVFs. No location of an intracranial DAVF was immune to either RLVD or PPP.

Five patients with PPP had no RLVD (Table 3). In the literature there are a number of reports of intracranial DAVFs with only sinosal drainage producing raised intracranial pressure. From these reports and our observation that five patients had PPP with sinosal drainage only, it could be suggested that these patients may have had venous hypertension within the involved sinuses, resulting in venous congestion. Four of these five patients had moderate or severe PPP, and their fistulas were all located in the transverse sinus/torcular. Two of the five were children who presented with congestive heart failure. Since PPP indicates venous congestion, its presence may be a factor to consider in management, especially in those patients without RLVD and in whom conservative management may be an option. Presentations with a hemorrhage or deficit in two of these five patients support the hypothesis that PPP without RLVD may be a sign that aggressive symptoms can occur despite the absence of RLVD.

Hydrocephalus and hindbrain herniation are not well-known findings in intracranial DAVFs; they are thought to be related to venous hypertension in the superior sagittal sinus, interfering with CSF absorption. Previous subarachnoid hemorrhage could also interfere with CSF absorption. Lasjaunias and Berenstein referred to this pathophysiology as a hydrovenous disorder. Five of our 130 patients had hydrocephalus and hindbrain herniation. Two of the five had white matter hyperintensity on T2-weighted MR images. Their presentations included dementia in three, ataxia in one, and increasing head circumference in a 4-month-old child. All five patients with hydrocephalus had PPP, and in four it was severe. Hydrocephalus and hindbrain herniation are more commonly seen in children with a high-flow fistula, such as a vein of Galen aneurysmal malformation. Treatment of this hydrovenous disorder should be directed toward eliminating the fistula instead of ventricular shunting.

Cognard et al reported worsening of venous drainage in seven partially treated patients with intracranial DAVFs and advised close follow-up of these patients. Davies et al did not encounter any worsening of symptoms in 55 untreated intracranial DAVFs with sinosal venous drainage only (Borden type 1), which were followed up for a mean of 33 months. Long-term follow-up is required for conservatively managed or partially treated patients and for those treated by surgical disconnection alone. The presence of moderate or severe PPP may be useful in highlighting a subgroup of patients in whom aggressive symptoms or signs may develop. Interval improvement in PPP may be a reassuring angiographic sign in the follow-up of partially treated patients.

Catheter angiography in the investigation of intracranial DAVFs must include good visualization of the venous phase of the brain circulation. The ideal study would be done under general anesthesia to obtain perfect subtractions; however, in most patients, this is not practical. Excellent visualization of the venous phase is needed to detect subtle findings, which may include pial or medullary collateral veins, focal regions of delayed circulation, and venous rerouting to the orbit or to transosseous veins. Objective assessment of the circulation time would be a useful adjunct in the assessment of venous congestion.

Conclusion

The PPP is a common finding in intracranial DAVFs that can be identified if the venous circulation of the brain is carefully scrutinized. Analysis of PPP in relation to presentation, RLVD, location of the DAVF, and MR findings shows that the severity of PPP reflects the venous congestion. PPP tends to be associated with aggressive presentation and RLVD. PPP may be a useful angiographic sign to consider in treatment decisions.

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