NEUROIMAGING CLINICS OF NORTH AMERICA

Neuroimag Clin N Am 13 (2003) 13–26

Venous variations of the brain and cranial vault

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The cerebral developmental venous anomaly (DVA), previously termed "venous angioma," is believed to represent a functional adaptation to absence of a normal cortical or deep venous pathway [1–6]. The anomaly is a relatively frequent finding on imaging studies of otherwise normal, asymptomatic patients. It is also known to be associated with symptoms in a minority of patients (presumably caused by relative obstruction to venous outflow) [6] and is seen in association with other intracranial vascular anomalies eg, cerebral arteriovenous malformations (AVMs), dural arteriovenous fistulas (AVFs), and cavernous malformations, as well as craniofacial vascular anomalies [7–12]. In this article, common and uncommon abnormalities of cerebral and cranial venous drainage will be illustrated.

Some relevant terminology will be reviewed first. The biologic classification of vascular anomalies proposed by Mulliken and Glowacki has two major categories: hemangiomas and vascular malformations [13]. This classification is supported by clinical, histologic, histochemical, and biochemical differences, as well as by features on angiography and cross-sectional imaging. Hemangiomas are proliferative endothelial cell tumors that typically present in infancy, and they have a characteristic period of rapid growth in the first year of life followed by slow involution. Almost all lesions involute spontaneously by age 9 years. Therapeutic interventions that speed up involution include pharmacologic treatment with steroids, interferon alpha, and vincristine [14]. Vascular malformations are considered to be developmental anomalies and are further classified according to the channel abnormalities (arterial, arteriovenous, capillary, venous, lymphatic, and mixed) or flow characteristics (high flow, low flow). They are believed to be present at birth but may become symptomatic later in life; they usually grow proportionate to the growth of the child and have no cellular proliferation or involution. The lesions can be readily

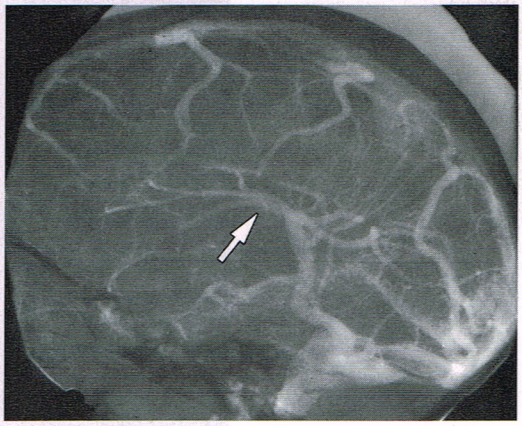

Fig. 1. Cerebral hemispheric developmental venous anomaly (DVA) in a child with a lymphatic malformation of the ipsilateral orbit. The right carotid angiogram demonstrates absence of the deep cerebral veins and vein of Galen. Periventricular veins (*arrow*) drain via a collector that courses in the perimedullary cistern.

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PII: S1052-5149(02)00061-8

distinguished by characteristic findings on CT, MR imaging, and angiography. Interventional radiologic techniques, such as embolization and direct intralesional sclerosant injection, play an important role in the management of these lesions.

Infantile hemangiomas infrequently involve the brain, and, when they do, they are usually on the brain surface (pial or dural) and are usually seen in association with multi-organ hemangiomatosis. They can be symptomatic by their associated mass effect (eg, hydrocephalus). They appear to involute at the same rate as cutaneous hemangiomas.

Among vascular malformations, all forms except the lymphatic malformation may involve brain and cervicofacial areas. Involvement of the face and brain simultaneously is well known in Wyburn-Mason syndrome [15], a rare condition in which facial and retinal vascular malformations are associated with an AVM of the ipsilateral intracranial optic pathway. The face and brain may also be targeted in hereditary hemor-

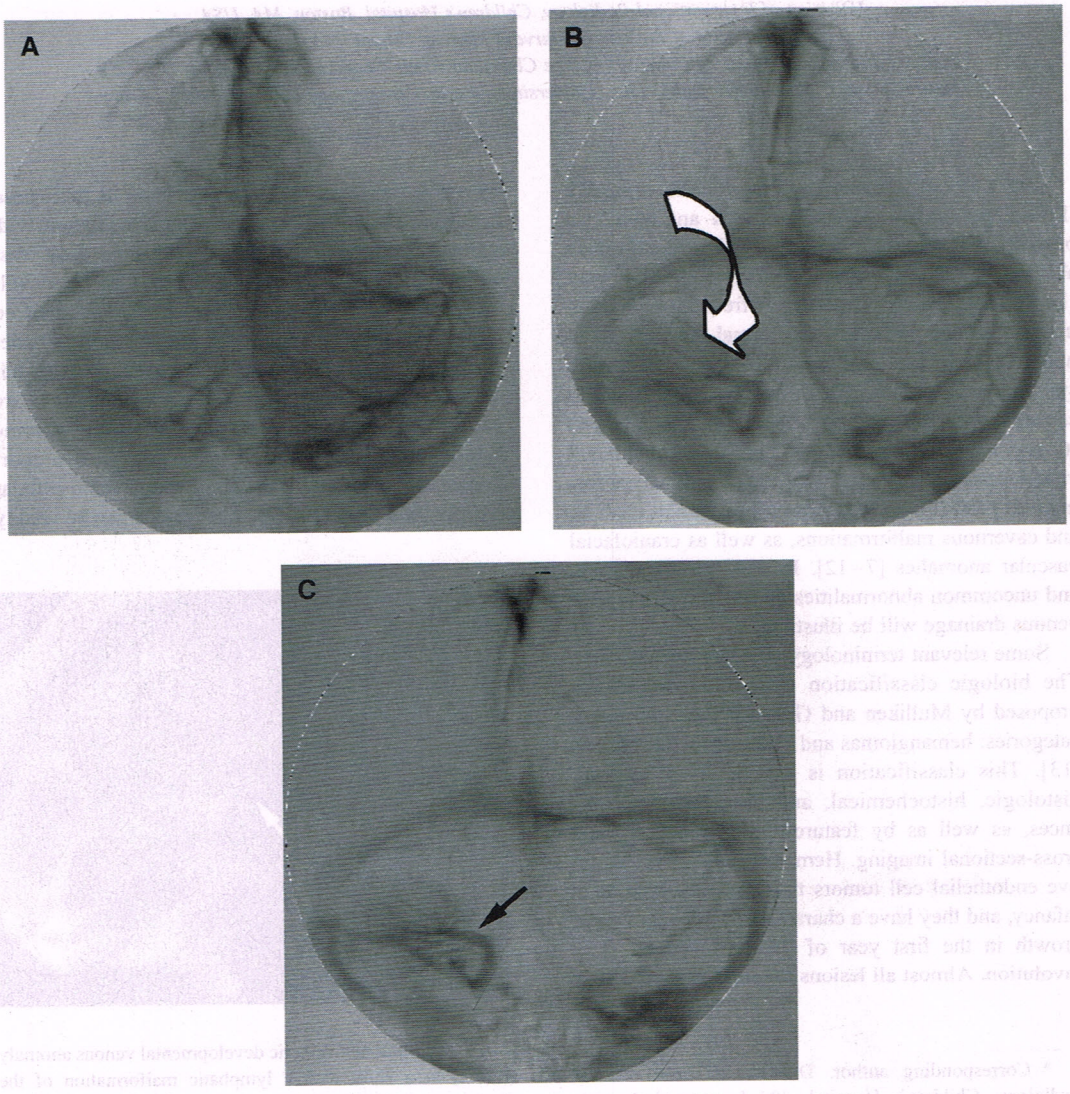


Fig. 2. Right cerebellar DVA associated with hemorrhage. Note the slower passage of contrast medium through the vessels of the right hemisphere, compared with the left, presumably on the basis of relative obstruction of the anomalous vein. (A) Early venous phase of left vertebral artery injection, Townes projection, shows a relatively hypovascular area in the drainage distribution of the anomalous right cerebellar vein. (B) Mid venous phase, showing a "parenchymal blush" around the DVA (*arrow*). (C) Late venous phase, shows persistent contrast opacification of the DVA (*arrow*).

rhagic telangiectasia and Sturge-Weber syndrome [16]. Other combined lesions are less well known.

Developmental venous anomaly

Intracranial DVA (previously called "venous angioma") represents a congenital alteration in the distribution in venous drainage of an arterial territory caused by focal underdevelopment of superficial or deep cerebral veins [1–6]. Most DVAs have a typical appearance on MR imaging and angiography. The umbrella or "candelabra" appearance of dilated intramedullary veins converging on an extraparenchymal collector vein is well known (Figs. 1 and 2). Cavernous malformations of the brain coexist in 8–33% of patients with DVA [8,11]. Though most are asymptomatic, DVAs have been associated with intracranial hemorrhage, seizures, focal neurologic deficits, and headaches [6]. Symptoms are presumably related to a relative obstruction of venous outflow, or an associated developmental brain anomaly (Fig. 2).

Cerebral high-flow arteriovenous malformations

Venous and dural sinus anomalies frequently coexist with intracranial high-flow vascular malformations, including vein of Galen aneurysmal malformations (VGAM), dural AVF (Figs. 3 and 4), and pial AVM [17–21] (Fig. 5). These anomalies include occlusive changes (Fig. 4), often felt to be acquired secondary to intimal hyperplasia induced by high flow, and anomalies of distribution (DVA) (Fig. 5). The venous occlusive changes are frequently responsible for hydrocephalus, cerebral atrophy, or focal neurologic (ischemic) symptoms, caused by the aggravation of cerebral venous hypertension.

Facial capillary malformation

The port-wine stain, a capillary or venular malformation, is most often an isolated lesion, but it may be associated with other deep vascular malformations, including AVM or lymphatic malformation (LM) [22].

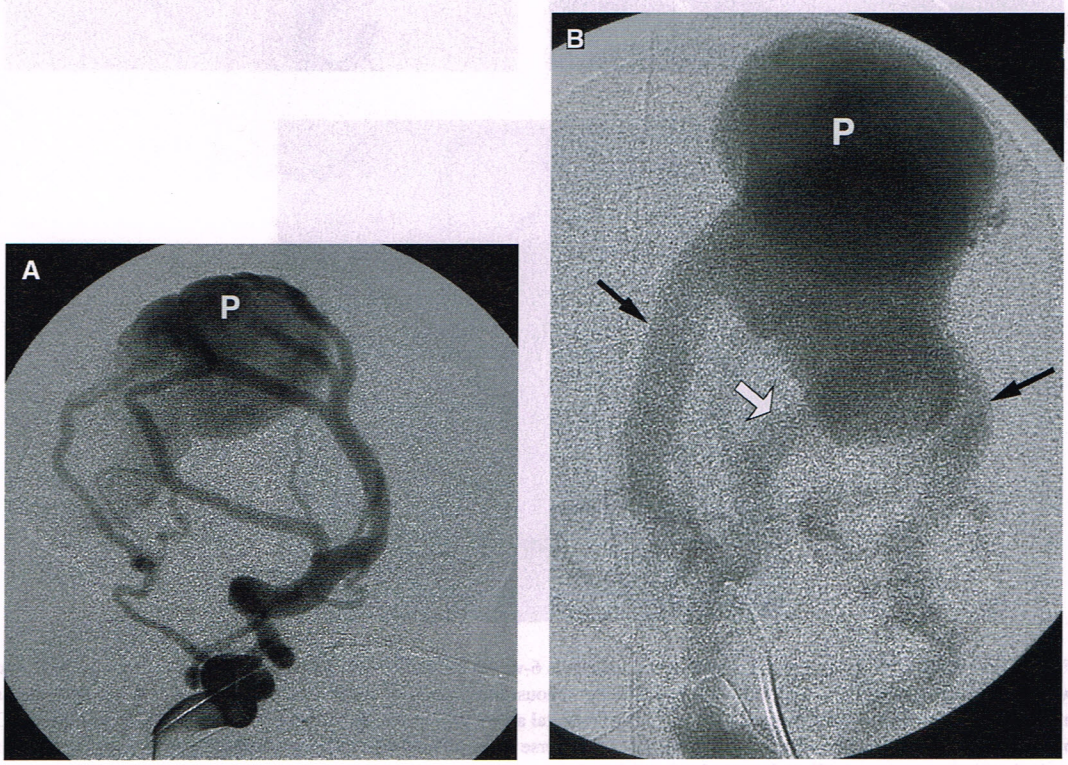


Fig. 3. Dural sinus anomaly in a preterm neonate with a high flow dural ateriovenous malformation (AVM). (A) The common carotid angiogram, lateral projection, shows a direct middle meningeal artery AVF with a varix or pouch (P) arising from the torcular. (B) The frontal projection shows patency of the dilated dural sinuses (*arrows*), including accessory occipital sinuses (*open arrow*). These accessory sinuses are frequently patent in neonates with high flow lesions. (*P* = dural pouch)

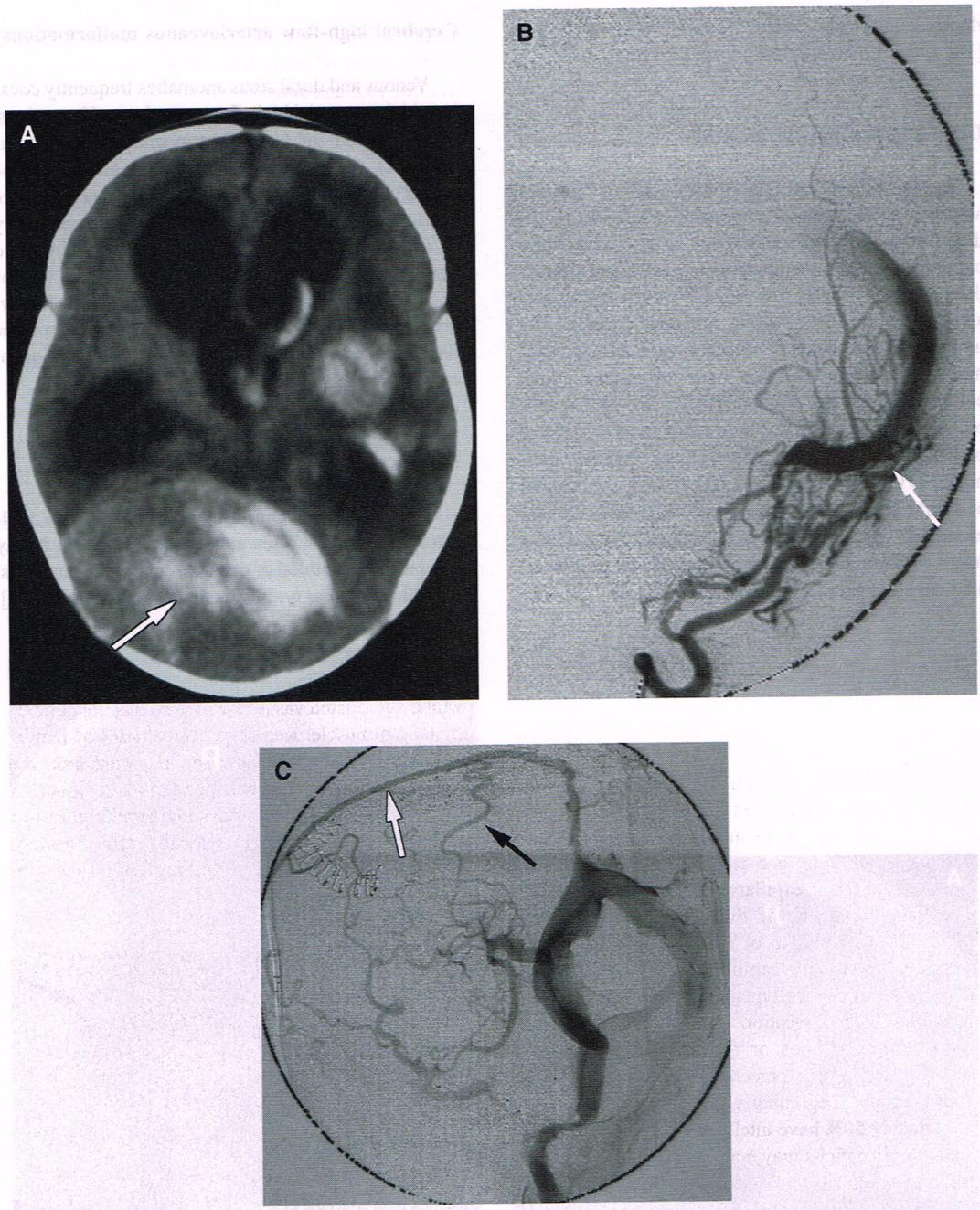


Fig.4. Dural AVM with thrombosed dural sinus anomaly, in a 6-week- old infant with macrocephaly and lethargy. (A) CT scan with contrast shows the large, partly thrombosed torcular venous pouch (*arrow*), hydrocephalus (secondary to cerebral venous hypertension), and hemorrhagic venous infarcts. (B,C) Occipital artery angiogram, lateral projection, arterial (B) and venous (C) phases, demonstrates the AVF into the partly occluded transverse sinus (*arrow*) (B), and retrograde opacification of the superior sagittal sinus (*open arrow*) (C) and cortical veins (*closed arrow*) (C).

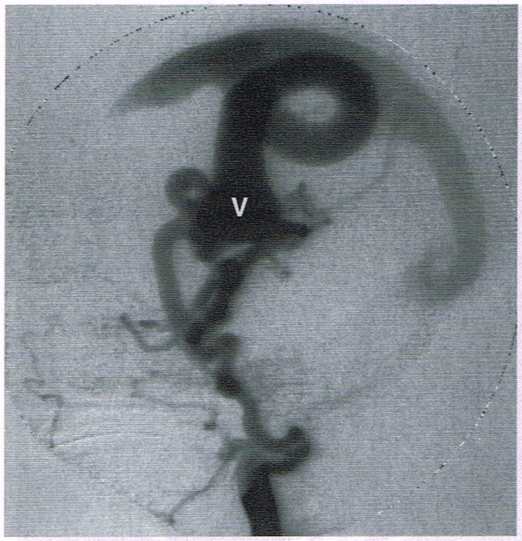


Fig. 5. Neonate with large right cerebral pial ateriovenous fistulas (AVFs) and absence of the deep cerebral venous drainage (confirmed at postmortem examination). Right common carotid arteriogram, lateral projection, shows direct shunting between sylvian branches of the middle cerebral artery and an anomalous sylvian varix (V), which drains into the superior sagittal sinus.

Sturge-Weber disease (encephalotrigeminal angiomatosis) represents a complex vascular anomaly of neuroectodermal layers, including the brain and its coverings, skull, skin, and retina [16,23]. The malformation includes a capillary or venular malformation (port-wine stain) of the facial skin, always involving the distribution of the first division of the trigeminal nerve, with capillary-venous or venular malformation of the leptomeninges, most commonly in a parieto-occipital location. Choroidal involvement leading to buphthalmos or glaucoma is present in 30% of cases. Ninety percent of the patients have epilepsy, usually beginning early in childhood, and approximately 50% have intellectual impairment. Focal neurologic deficits may occur, often in an acute or step-like fashion.

Characteristic CT and MR imaging findings include focal cerebral atrophy with gyriform calcification, diffuse leptomeningeal enhancement of the affected area, and enlargement and enhancement of the ipsilateral choroid plexus. The cortical atrophy and calcification are likely caused by venous ischemia. It may be present at birth but usually appears in early childhood. Leptomeningeal enhancement, with enlargement of the choroid plexus, appears earlier. Cerebral angiographic findings are variable but include a capillary blush, contrast stasis, and delayed opacification of abnormal cortical veins (Fig. 6). Cortical vein and sinus thrombosis has been reported, although the mechanism of this venous occlusive process is not known. Trans-medullary venous collaterals are typical. External carotid angiography may demonstrate high-flow vascular malformations of the calvarium and meninges.

Cervicofacial venous malformation

Venous malformation (VM) is the most common symptomatic vascular malformation involving the head and neck. All VMs are present at birth but may not be evident until later. They usually grow proportionately with the patient. Characteristic clinical features include bluish skin discoloration with compressible soft tissue swelling that increases with dependency or increased venous pressure. The most common clinical presentation of a cervicofacial VM is facial deformity, followed in decreasing order by pain, drooling, dental distortion, speech difficulty, sleep apnea, and visual obstruction [24]. On histopathologic examination, the classic VM is composed of thin-walled and dilated vascular channels that are deficient in smooth muscle. Recent genetic studies of familial VM have shown gain of function mutations involving Tie-2, the angiopoietin receptor [25–27]. Most VM are solitary or focal, although some familial forms are multi-focal. Diffuse VMs typically affect multiple tissue layers and consist of dysplasia of all the veins

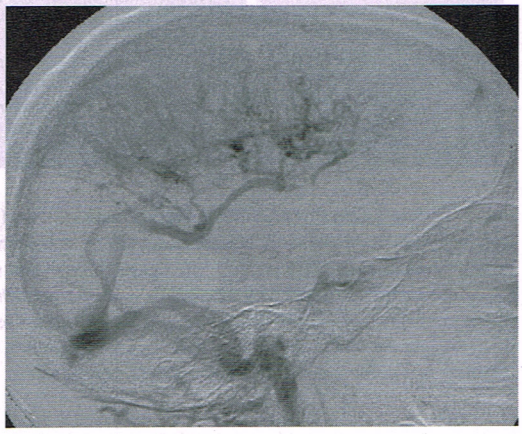


Fig. 6. Sturge-Weber syndrome. Left internal carotid angiography, lateral (A) and frontal (B) planes, shows tortuous medullary collateral veins draining into the deep venous system, with no filling of the cortical veins.

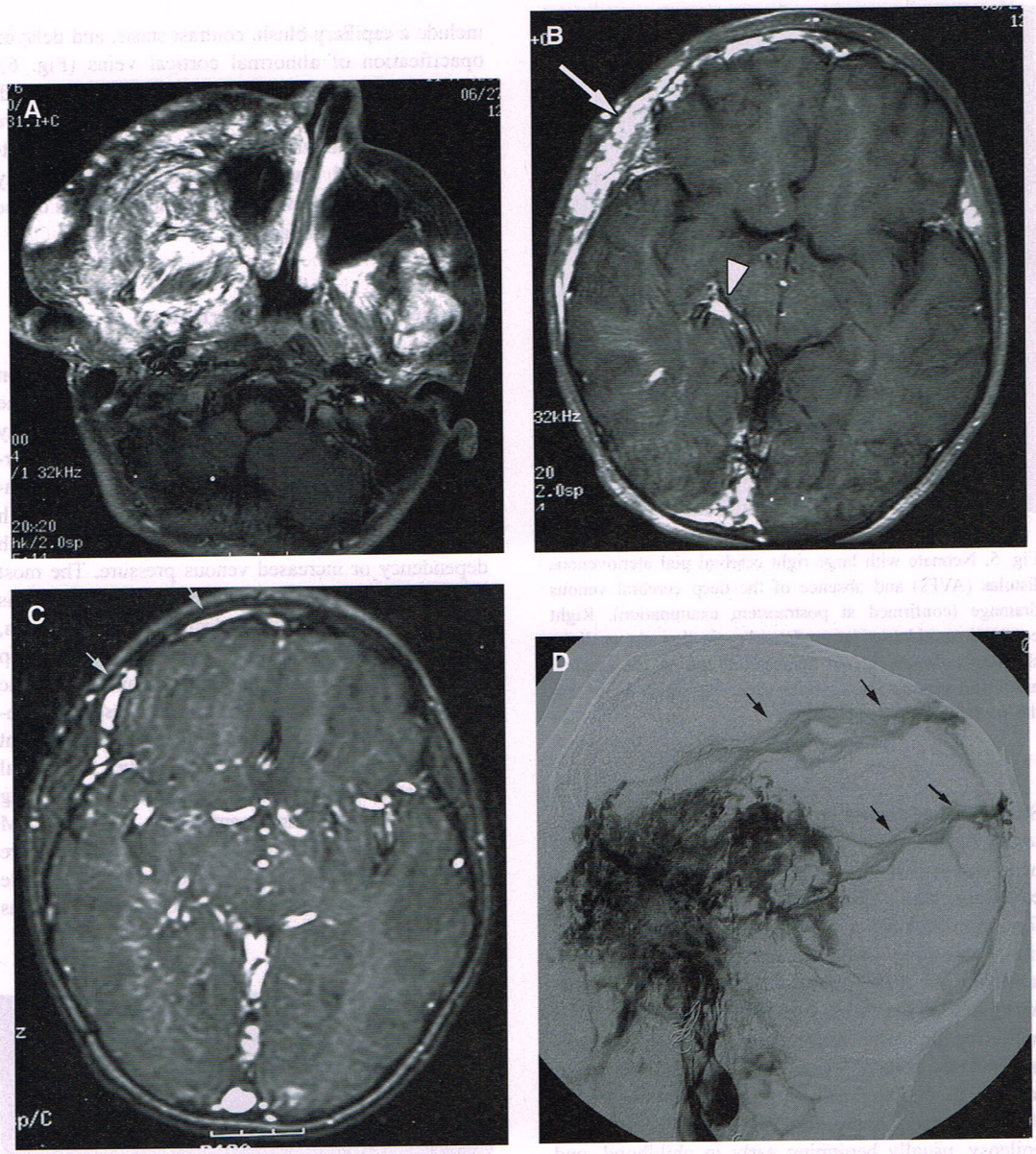


Fig. 7. Diffuse bilateral cervicofacial venous malformation (VM) with DVA, calvarial VM, and sinus pericranii. (A) Axial T1-weighted MR image post-gadolinium shows diffuse enhancing soft-tissue masses and abnormal enhancement of the zygoma. (B) Postgadolinium axial T1-weighted MR image through the midbrain shows abnormal enhancement of the right frontoparietal calvarium (*arrow*), a right perimesencephalic DVA (*arrowhead*), and abnormal signal in the torcular. (C) Gradient echo MR image confirms that the enhancing areas in the calvarium seen in Fig. 7B are high flow channels (*arrows*) (D) Direct contrast injection into the VM in the right temple, digital subtraction, lateral projection, shows drainage in irregular intraosseous channels (*arrows*) that communicate with the superior sagittal sinus.

in the involved area. These lesions often contain varicosities of superficial conducting veins [24].

Most cervicofacial VMs are isolated sporadic lesions. Less common hereditary or syndromic forms occur, including glomangioma [glomovenous malformation (GVM)], familial cutanomucosal VM, Maffucci syndrome, and blue-rubber bleb nevus syndrome (BRBNS). GVM is characterized by painful nodular skin VM, whereas the lesions in familial cutaneomucosal VM typically are dome-shaped and develop progressively over time. The distinguishing feature of BRBNS is the additional presence of multiple gastrointestinal VMs resulting in chronic gastrointestinal bleeds. Maffucci syndrome, on the other hand, is characterized by bony exostoses and enchondromas with cutaneous VM-like lesions containing spindle cell hemangioendothelioma.

Within the head and neck, facial involvement (particularly buccal, periorbital, and labial regions) is most common. VMs range in extent from a few millimeters in diameter to a diffuse involvement of the entire head and neck that causes significant distortion of cervicofacial anatomy. Some VMs are limited to subcutaneous tissue or isolated muscles, whereas diffuse lesions may involve all tissue layers including the calvarium, dural sinuses, and cerebral veins (Figs. 7 and 8).

The most helpful imaging test in diagnosing and evaluating the extent of VMs is MR imaging [23,28–30]. VMs typically present as a high-signal soft tissue abnormality on spin echo T2-weighted images and demonstrate inhomogeneous contrast enhancement. High-flow vessels are characteristically absent on gradient echo (GE) sequences. T2-weighted images are commonly used to assess the extent of a VM. Postgadolinium images help to differentiate VMs from LM and GE images are used to rule out a high-flow vascular anomaly (eg, AVM). Other imaging modalities have a limited role in the diagnostic work-up of these abnormalities, although ultrasonography is useful to determine flow characteristics, and direct intralesional contrast injections are commonly used to outline the vascular spaces and drainage pattern during sclerotherapy. Plain radiographs and CT may demonstrate any osseous involvement, and, in some cases, characteristic phleboliths within the lesion. Soft tissue phleboliths are highly suggestive of VM and are rarely found in other slow-flow vascular anomalies (eg, LM, LVM).

Calvarial venous malformation

Calvarial VMs are rarely isolated; most are associated with diffuse adjacent VM (Fig. 7). Calvarial involvement is particularly common in large VMs that are located in the temporal-parietal regions [31]. The thickness of the involved calvarium may be normal or increased. Increased calvarial thickness is usually caused by the extension of the VM within the diploic space, which may split the inner and outer tables. When the calvarium is involved, palpation of the area may reveal a bony defect or irregularity. Involvement of the calvarial diploic space in extensive cervicofacial VMs are frequently associated with sinus pericranii and intracranial developmental venous anomalies.

Isolated calvarial VM is commonly referred to as "osseous hemangioma" and often classified erroneously as a tumor in the literature. It has been reported that these isolated lesions are somewhat more common in women, and they have characteristic "sunburst" appearance without definite margins on plain radiographs, causing thickening of the outer table without displacing the inner table or prominent scalloping in the inner or outer table [32]. Sessile and globular types have been described. These patients usually present with a palpable painless scalp mass, or the lesion is incidentally detected during an imaging evaluation. Common locations in the skull are reported to be frontal and temporal regions, and 15% of patients have multiple lesions. Because of its rare occurrence in the calvarium, histopathologic confirmation has been suggested.

On MR imaging, the calvarial involvement is noted as hyperintense signal abnormality on T2-weighted MR images, and abnormal contrast enhancement on the post-contrast T1-weighted MR images. The T2-weighted spin-echo images are particularly sensitive to detecting any calvarial involvement. The involvement of the calvarium may be seen as a small area of MR signal change only within the outer surface of the bone that is adjacent to the craniocervical VM, or it may be the entire thickness of the calvarium, with or without extending into the intracranial extra-axial space. CT is less sensitive but may show a bony defect or an osseous expansion on the bone window images.

Sinus pericranii

Sinus pericranii (SP) represents a communication between the intracranial and extracranial venous circulations, often associated with an extracranial vascular malformation or varix-like abnormality of the forehead [4,31,33–37] (Fig. 9). The imaging features and appropriate management of these patients, either surgically or by percutaneous interventional means, is

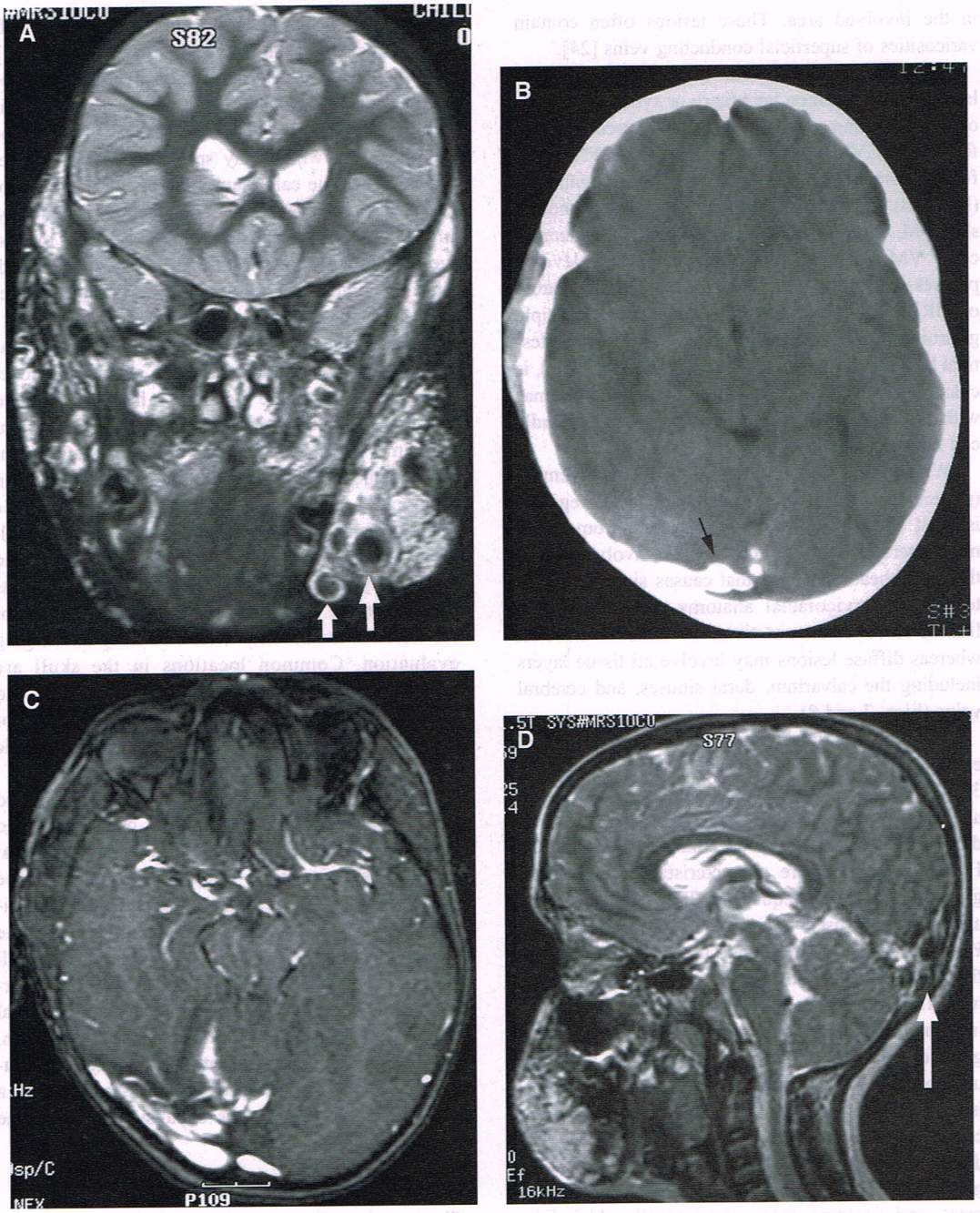


Fig. 8. Ten-year-old girl with bilateral cervicofacial VM, anomalous right transverse sinus, and right skull base sinus pericranii. (A) Coronal T2-weighted MR image demonstrates bilateral VM, with phleboliths (*arrows*). (B) Axial CT image without intravenous contrast medium show calcifications (*arrow*) in the region of the right transverse sinus. (C) Gradient echo recalled MR image confirms the abnormally dilated right transverse sinus with an intraluminal calcification. (D) Abnormal torcula (*arrow*) is seen on the sagittal T2-weighted MR image. (E) Left vertebral angiogram, Townes projection, confirms the dural sinus anomalies, as well as cerebellar DVA (*arrow*).

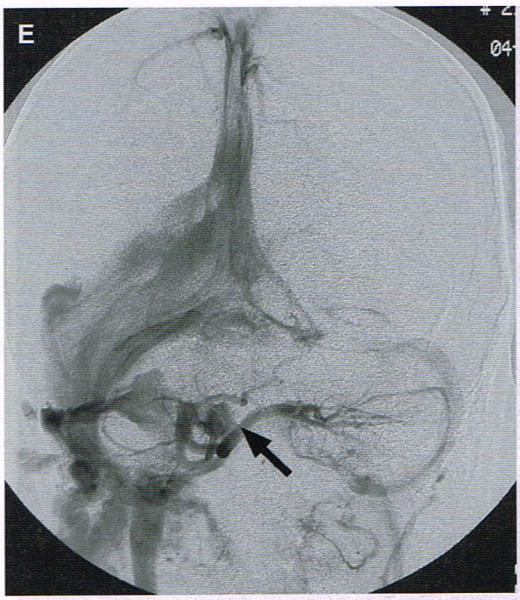


Fig. 8 (*continued*).

not well known. The terminology used to describe the extracranial vascular anomaly associated with this condition, and the exact nature of the associated extracranial vascular abnormality, is unclear in most published reports. Most of the confusion arises from the misnomer or improper use of "angioma" or "hemangioma" as a synonym for VM. The classic presentation of this condition has been described as a round, fluctuant, nonpulsatile soft-tissue prominence or mass that disappears with compression and increases in size when the head is down or during crying and coughing. It is our experience that the associated extracranial vascular anomaly is a VM in most cases; it is rarely a lymphatic malformation or arteriovenous malformation. SP is seen in 11% of patients who present for treatment of cervicofacial VM [31]. Twenty percent of patients with SP have VMs elsewhere in the body and include patients with BRBNS. SP has also been described in patients with craniosynostosis and other skull base anomalies [12,38] (Fig. 10).

Some patients with SP have vague symptoms such as nausea and headache. It can also be postulated that these patients are prone to more serious complications if any direct impact occurs to the SP, or if the associated extracranial abnormality is removed surgically without confirming the adequacy of the intracranial circulation. In our practice, most SP patients seek treatment because of facial deformity, pain, and discomfort, and the symptoms related to the SP are minimal if any.

SP has been classified as "closed" versus "drainer," and as true SP versus pseudo SP. The most useful classification, in terms of management of these patients, is the classification based on flow dynamics (closed vs. drainer). In the closed type, the blood flows from the intracranial venous circulation to the sinus and extracranial vascular anomaly without draining into the extracranial venous circulation, whereas in the drainer-type, drainage into the extracranial venous circulation occurs. In our experience, most SPs associated with VM are drainer type (63%) [31].

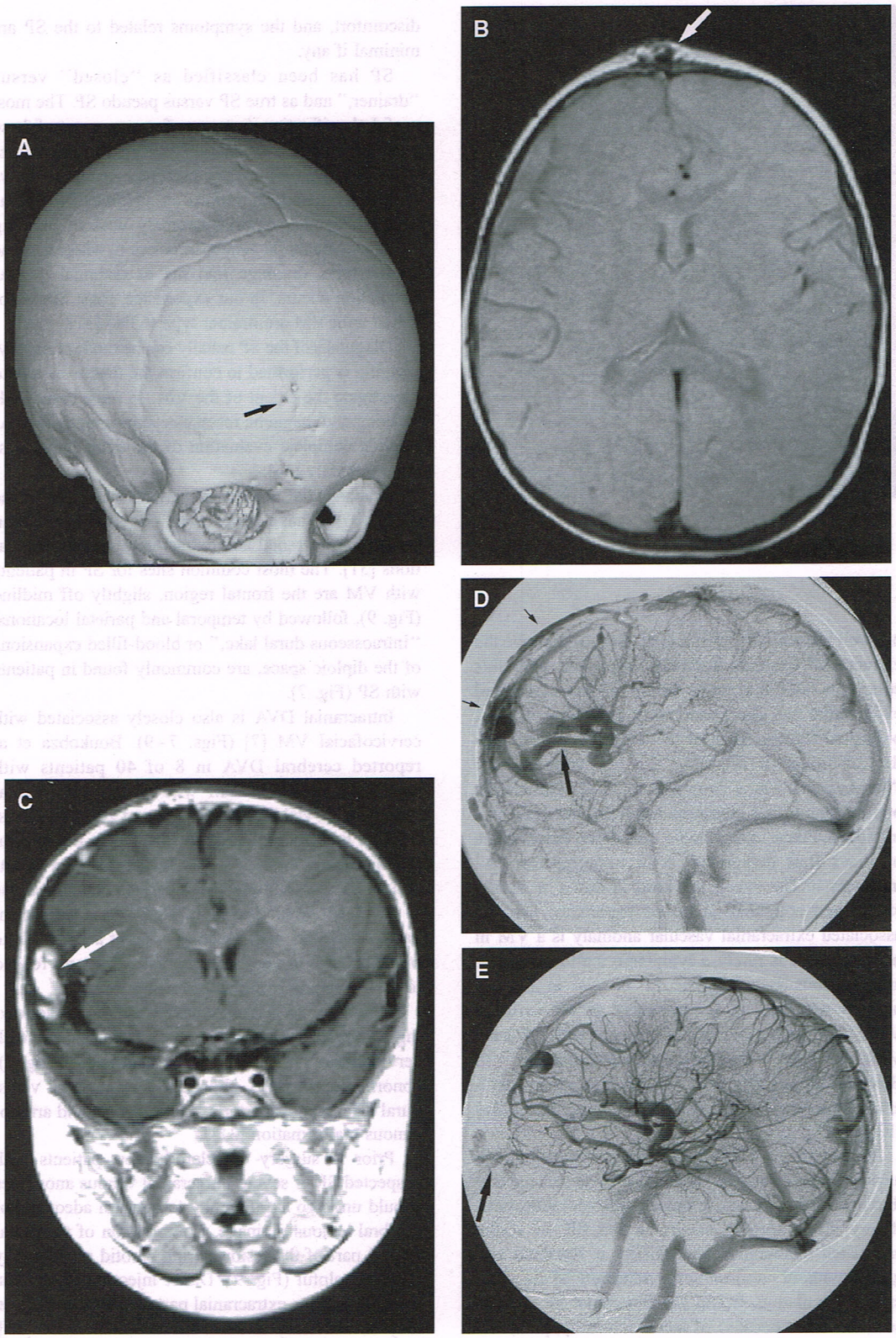
Diagnosis of the SP usually can be made clinically; imaging is performed to confirm the diagnosis and to investigate the extent of the VM. In most cases, MR imaging and magnetic resonance angiography (MRA) provide complete evaluation of the extracranial vascular anomaly [34].

A substantial number of patients with VM of the scalp (37%) have more than one communication between intracranial and extracranial venous circulations [31]. The most common sites for SP in patients with VM are the frontal region, slightly off midline (Fig. 9), followed by temporal and parietal locations. "Intraosseous dural lake," or blood-filled expansions of the diploic space, are commonly found in patients with SP (Fig. 7).

Intracranial DVA is also closely associated with cervicofacial VM [7] (Figs. 7–9). Boukobza et al reported cerebral DVA in 8 of 40 patients with extensive superficial VM who underwent cerebral angiography, yielding a prevalence of 20% in contrast with an incidence of less than 0.25% in the general population [7]. In their series, the DVAs were unilateral in 7/8 and multiple in 5/8 patients. Drainage was to the deep circulation in 14/18 DVAs. One patient had a cavernous malformation without a DVA. None of the patients had any documented symptoms related to the intracranial venous anomalies.

In addition to DVA, we have observed the following intracranial vascular anomalies in patients with cervicofacial VMs: dural sinus abnormalities (Fig. 8), abnormal dural veins, abnormal deep cerebral veins, dural calcifications, arteriovenous fistula, and arteriovenous malformation [31].

Prior to surgery or sclerotherapy, patients with suspected SP or severe intracranial venous anomalies should undergo angiography to confirm adequacy of cerebral venous drainage. Compression of the extracranial part of the lesion during carotid angiography may be helpful (Fig. 9). Direct injection of contrast material into the extracranial part of the malformation may be necessary to show the communication [23].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Standard treatment of sinus pericranii involves excision of the extracranial lesion and closing of the calvarial defects with bone wax [36]. Small or single communications can be closed percutaneously by venous embolization of the extracranial draining veins using coils and sclerosants, or theoretically using adhesive polymer.

Orbital lymphatic malformation

Lymphatic malformations are uncommon vascular anomalies consisting of dilated lymphatic channels or cysts [13,39–45]. Histologic features include thin-walled endothelium-lined channels with loose junctions and varying stromal components. Seventy-seven percent involve the head and neck. They are more common in the neck region than in the orbit. In the general population, the incidence of orbital LM is approximately 3.5 per 100,000 persons. Most patients are symptomatic with proptosis, orbital nerve compression, and recurrent hemorrhage. Ocular complications are common in patients with orbital LM and include marked astigmatism, proptosis, hyperopia secondary to pressure on the posterior aspect of the globe, strabismus, glaucoma, amblyopia, ptosis, cosmetic deformity, compressive optic neuropathy, and cellulitis. The size of the LM may fluctuate with concomitant upper respiratory infection.

Though the channels of LM may be fully formed at birth, the lesion can remain inconspicuous until it expands. The major growth of LM is caused by expansion of the channels by intrinsic hemorrhage or infection. Bleeding, most likely from small nutrient vessels within the fragile walls, may occur spontaneously or after trauma. Aside from intermittent enlargement, the LM growth generally parallels the growth of the child.

At MR imaging, orbital LM consist of T2-weighted hyperintensities with enhancing septae [23,29,30,46]. In many cases, discrete cysts, often with fluid/fluid levels, can be discerned. The contents of the cysts may enhance, especially after surgical intervention. The orbital and frontal bones are often abnormal, and, in the presence of intraconal or postseptal involvement, the orbit is expanded. Angiography may show some hypervascularity without arteriovenous shunting.

A few patients with combined extracranial LM, and intracranial vascular anomalies have been reported. The anomalies described include noncontiguous intracranial anomalies [9], complex DVAs [18], ipsilateral dural AVF [1], and intracranial AVM [19] (Figs. 1, 11, and 12).

The embryogenesis of DVA is unclear, as is the embryogenesis of cervicofacial LMs. Recent work with vascular endothelial growth factors (VEGFs) and their specific cell surface receptors may shed some light on the origin of the lymphatic system, its relationship to the venous system, and ultimately abnormalities that arise in vasculogenesis [27]. Conclusions from recent genetic experiments support theories of the venous origin of lymphatic vessels as originally proposed by Sabin and later expanded on by Van der Putte [13]. The origin of facial vasculature from neuroectoderm presumably explains the coexistence of extracranial and intracranial venous anomalies. Padget's work [47] on the development of the circulatory system indicated that abnormal development at an appropriate gestational stage could result in an anomalous draining vein supplementing the usual drainage.

Summary

Vascular anomalies involving both intra- and extra-cranial structures are more common than previously thought. It is important to evaluate the brain and its coverings carefully when imaging cervicofacial vascular malformations. Scientific knowledge regarding developmental mechanisms responsible for blood vessel formation is increasing rapidly and, hopefully, will contribute to better understanding of these clinical and imaging "patterns."

Fig. 9. Two-year-old boy with midfrontal VM, underlying SP and cerebral DVA. (A) Three-dimensional CT scan shows calvarial defects to the right of the midline (*arrow*). (B) Proton density axial MR image, showing flow voids (*arrow*) in the scalp, corresponding to the subcutaneous VM. Normally, VM do not appear as flow voids, but rather as stagnant fluid. (C), T1-weighted coronal MR image shows an enhancing cortical varix in the right frontal convexity, part of the DVA. (D) Right internal carotid angiogram, lateral projection, demonstrates the right frontal DVA and subsequent filling of the VM of the scalp (*arrows*). (E) Repeat carotid angiogram, with compression of the extracranial VM confirms adequate venous outflow of the DVA. Note that the VM also fills from the superior ophthalmic vein.

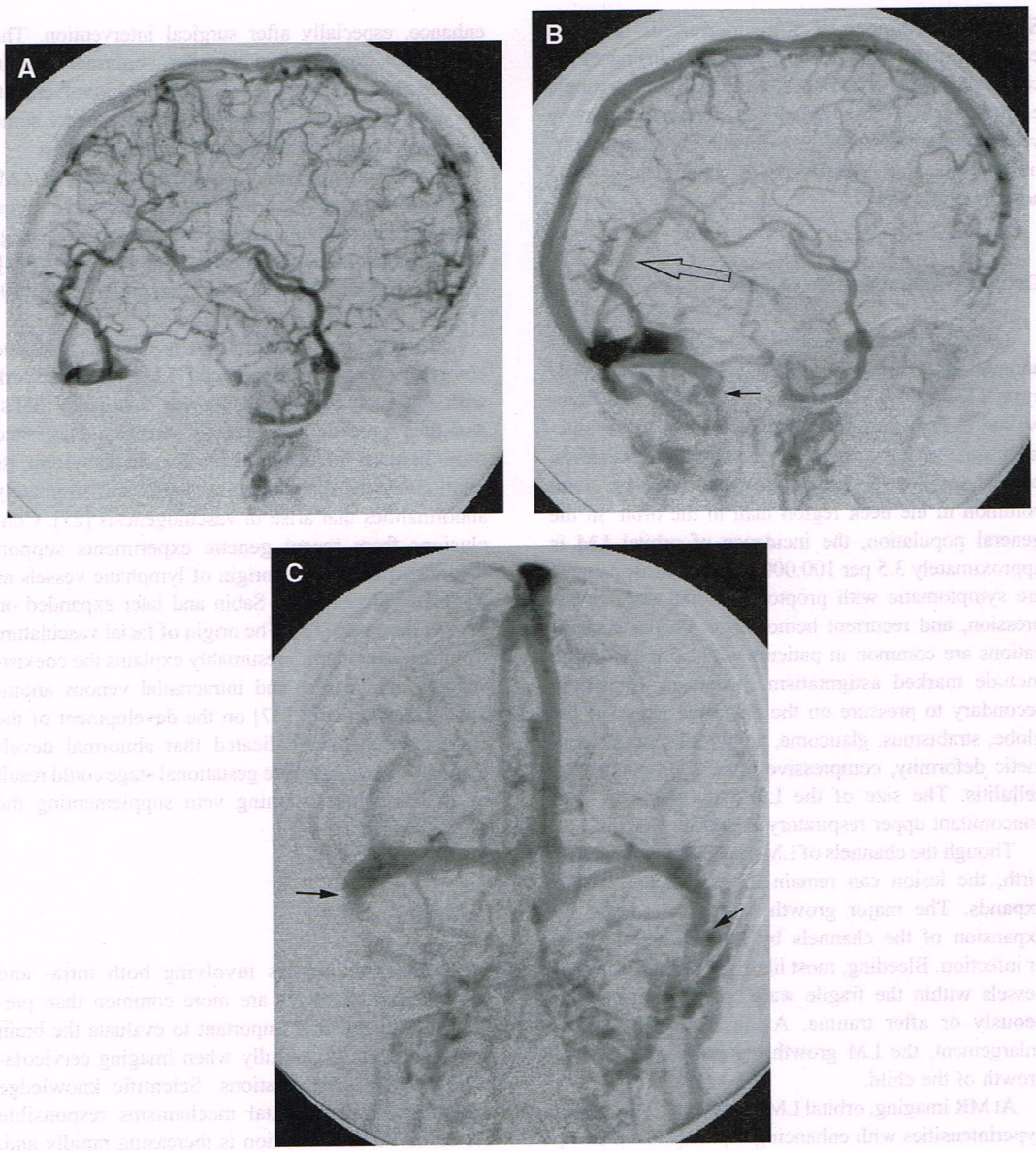

Fig. 10. Dural sinus obstruction associated with craniosynostosis. This 5-year-old boy with Apert's syndrome developed severe headaches and hydrocephalus after cranioplasty and ligation of a "pulsatile mass" in the right neck. (A and B) Right internal carotid angiogram, venous phases, digital subtraction, lateral projection, shows congestion of the cerebral veins and obstruction of the sigmoid-jugular junction (*closed arrow*). Note the elevation of the straight sinus caused by the foreshortened cranial vault (*open arrow*). (C) The frontal projection confirms the bilateral dural sinus obstructions (*arrows*) with emissary veins or SP on the left. Corresponding veins on the right were presumably ligated.

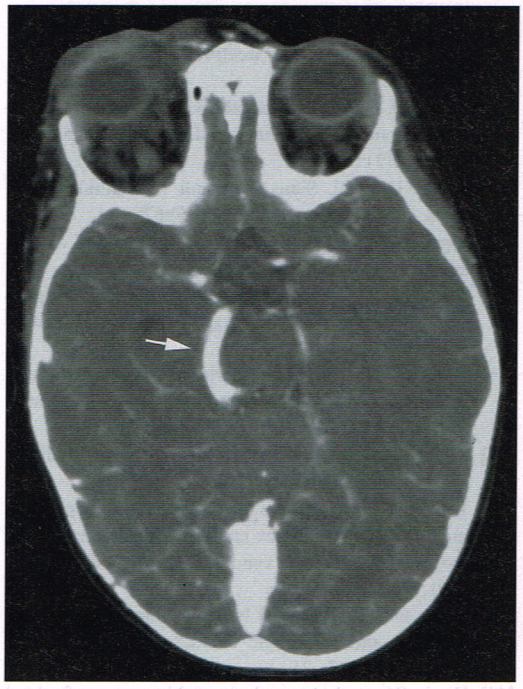

Fig. 11. Right orbital LM with DVA draining into ipsilateral perimesencephalic collector (*arrow*).

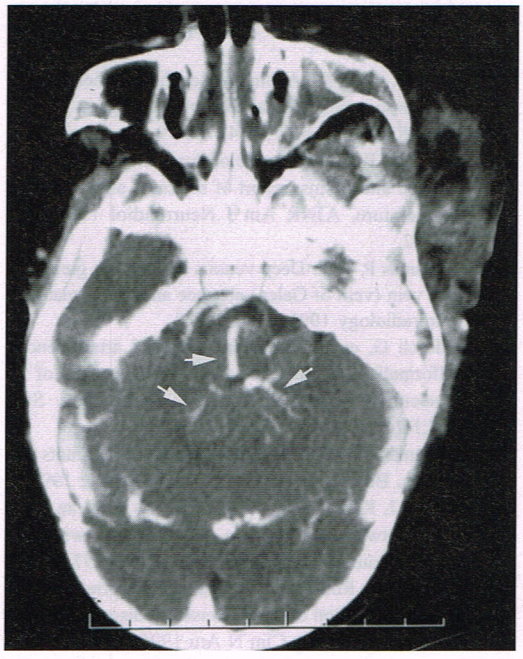

Fig. 12. Left orbital LM with cerebellar DVA, draining into a transpontine collector.

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