

# Venous angiomas of the brain — a review

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## Introduction

Venous angiomas of the brain, also termed venous malformations or developmental venous anomalies (DVA) are commonest of the intracranial vascular malformations comprising between 50% and 63% of all intracranial vascular malformations. They may be found in as many as 2 - 2.5% of the general population.<sup>1-3</sup>

The term 'venous angioma' was introduced by Russel and Rubinstein in 1951.<sup>4</sup> In 1963 Courville<sup>5</sup> first suggested the concept that venous angiomas represent compensatory venous drainage pathways in the brain. Later reports indicated that these venous angiomas seemed to drain normal brain tissue and were to be considered as anomalous venous drainage systems.<sup>6</sup> Huang *et al.*<sup>7</sup> described these anomalies as being purely venous with no associated arterial or capillary abnormalities and suggested the term 'medullary venous malformation' rather than venous angioma.

Finally in 1986 Lasjuanias *et al.*<sup>8</sup> coined the term 'developmental

venous anomaly' or DVA, pointing out that these abnormalities actually represented an extreme anatomical variant of the normal venous drainage of the brain.

## Pathology

The theory of the development of venous angiomas is that there is failure of regression of normal embryonic transmedullary venous channels. These persistent transmedullary veins run axially through the white matter to drain into a single larger calibre collecting venous trunk. The dilated terminal collecting vein then penetrates the cortex to drain either superficially into cortical veins or dural sinuses, or deeply in the subependymal veins and deep venous system. In the posterior fossa collecting veins may drain into the pontomesencephalic or cerebellar veins. A further effect of this apparent halt in venous anatomical development is that there is regional hypoplasia or aplasia of normal pial veins in the same area of the brain. The result of this is that the anomalous vein drains the normal brain tissue in the area affected.

In the cerebral hemispheres, Valavanis *et al.*<sup>9</sup> described both superficial and deep types. With the superficial type there is drainage of the subependymal and deeper medullary regions into the cortical veins, and with the deep type the blood from the subcortical white matter drains into the subependymal and deep venous

systems.<sup>9</sup> Venous angiomas may be quite small, draining a limited region of the brain, or may be very large, sometimes draining an entire hemisphere. They can be single or multiple, and even bilateral.<sup>10,11</sup> The commonest sites of occurrence are in the frontal and parietal lobes of the cerebral hemispheres and in the cerebellum.<sup>11,12</sup> They can also be found in the occipital and temporal lobes, basal ganglia and pons.<sup>13</sup>

## Imaging

The classical radiographic appearance of these abnormalities accurately reflects the anatomical picture with multiple enlarged transmedullary veins radiating in a wedge or radial pattern toward the larger collecting vein producing the pathognomic 'caput medusae' or 'spoke wheel' appearance during the venous phase of a cerebral angiogram (Figs 1, 2).<sup>14,15</sup> A similar appearance is often seen on computed tomographic (CT) or magnetic resonance (MR) scanning of the brain (Fig. 1), although with smaller angiomas often only the collecting vein is seen (Fig. 3). These angiomas are best seen on CT scanning following contrast administration. Similarly, they are also usually best seen on MR imaging on contrast-enhanced T1-weighted images.<sup>12,16</sup> The collecting veins are seen as either straight or serpiginous enhancing structures traversing the brain parenchyma on CT or MR. They may be seen as flow voids on standard unenhanced T1 and proton density MR sequences (Fig. 2), and due to slow flow the collecting veins may also show as hyperintense on T2-weighted images. The enhancement of the collecting veins with gadolinium on MR scanning in keeping with other normal venous

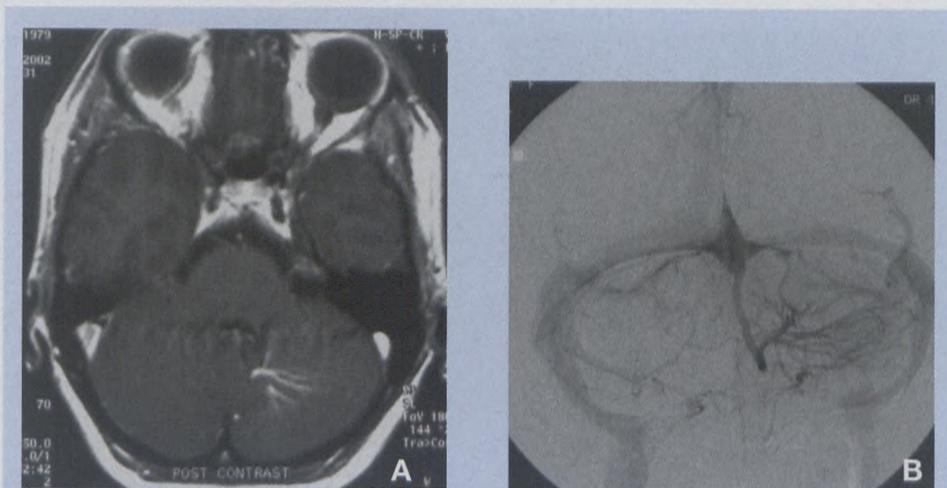


Fig. 1. Cerebellar venous angioma: characteristic appearance. Axial contrast-enhanced T1-weighted MR image (Fig. 1a) with corresponding venous return phase image during a selective left vertebral arteriogram (Fig. 1b) both showing the typical 'caput medusae' appearance of a venous angioma within the cerebellum.

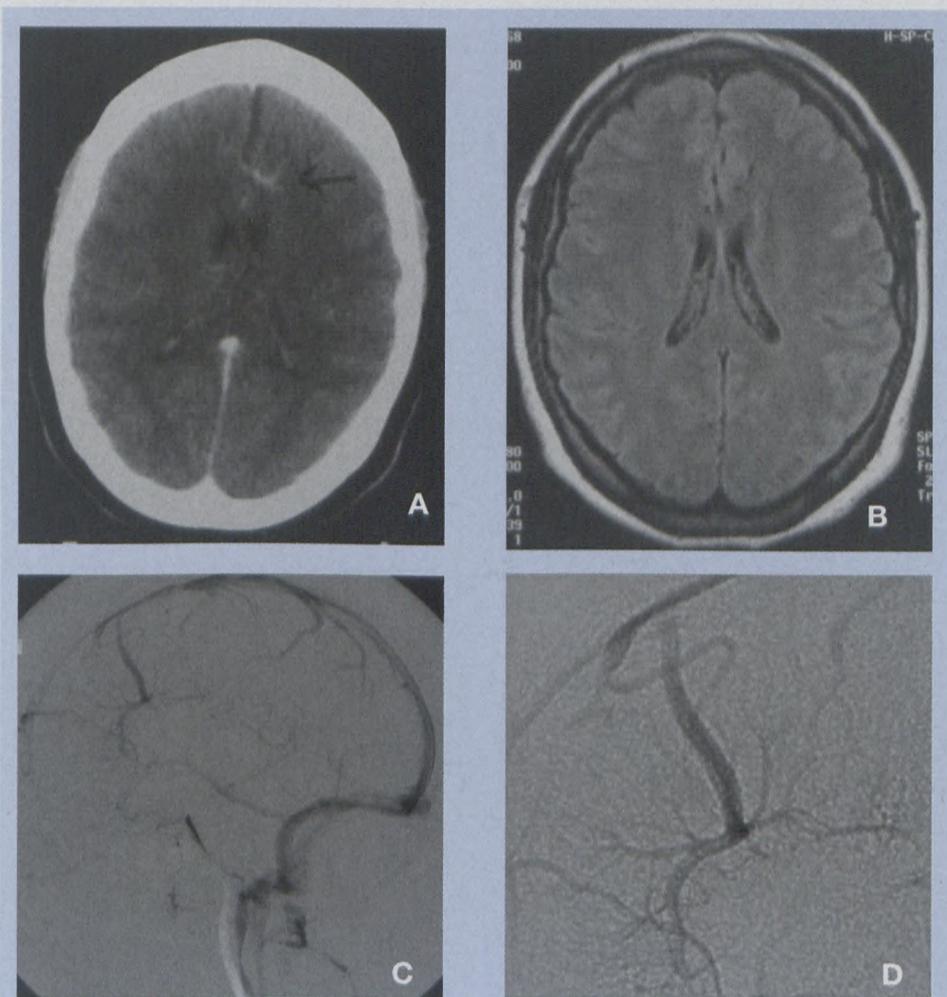


Fig. 2. Frontal venous angioma: characteristic appearance. Contrast-enhanced axial CT (Fig. 2a) and axial proton-density MR (Fig. 2b) images at the same level showing a venous angioma in the left frontal lobe. It shows as an enhancing structure on the former and as a flow-void on the latter. The venous return phase images of a selective left internal carotid arteriogram (Figs 2c, 2d) show the same deep venous angioma draining superficially.

structures of the brain is in contrast to the appearance of dilated veins in a high-flow situation such as is seen with the veins draining an arteriovenous malformation or fistula, where these veins tend to show as flow voids on all sequences despite contrast administration.

Occasionally decreased signal intensity may be seen in venous angiomas on gradient-echo MR images due to magnetic susceptibility resulting from deoxyhaemoglobin in the veins.<sup>17</sup> Venous angiomas can be seen on MR venography but this seldom adds any more information than standard multiplanar MR imaging.

During angiography these venous angiomas are seen during the venous return phase of a selective cerebral angiogram. The arterial phase is generally normal although occasional regional arterial supply variations may be seen. A prominent late capillary blush may be seen in the area drained by a venous angioma followed by filling of the medullary veins and later the collecting vein producing the characteristic angiographic appearance.

## Pathological associations

Although most venous angiomas occur in isolation, usually found as incidental findings, they are known to have associations with other cranial vascular and developmental abnormalities. These include cavernous malformations, arteriovenous malformations, capillary telangiectasias, sinus pericranii and other facial vascular malformations, and cortical dysplasias and other migrational brain abnormalities. Venous angiomas may be associated with other intracranial vascular malformations in up to 9% of cases.

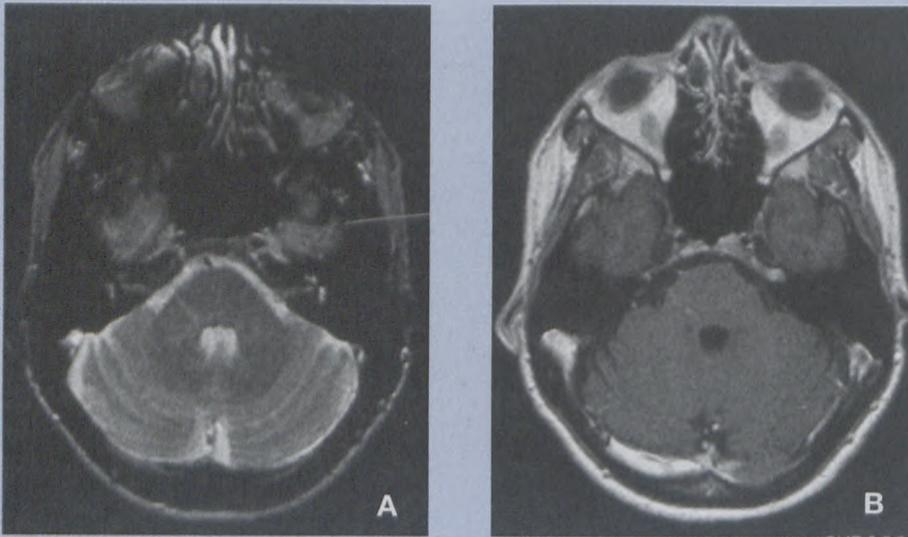


Fig. 3. Probable brainstem venous angioma. Axial T2-weighted (Fig. 3a) and contrast-enhanced T1-weighted (Fig. 3b) images showing a linear vascular structure traversing the brainstem on the right at the level of the pons in a patient who presented with an ipsilateral trigeminal neuralgia. The linear structure is probably the collecting trunk of a brainstem venous angioma. This was not confirmed angiographically. Its relationship to the clinical presentation is questionable.

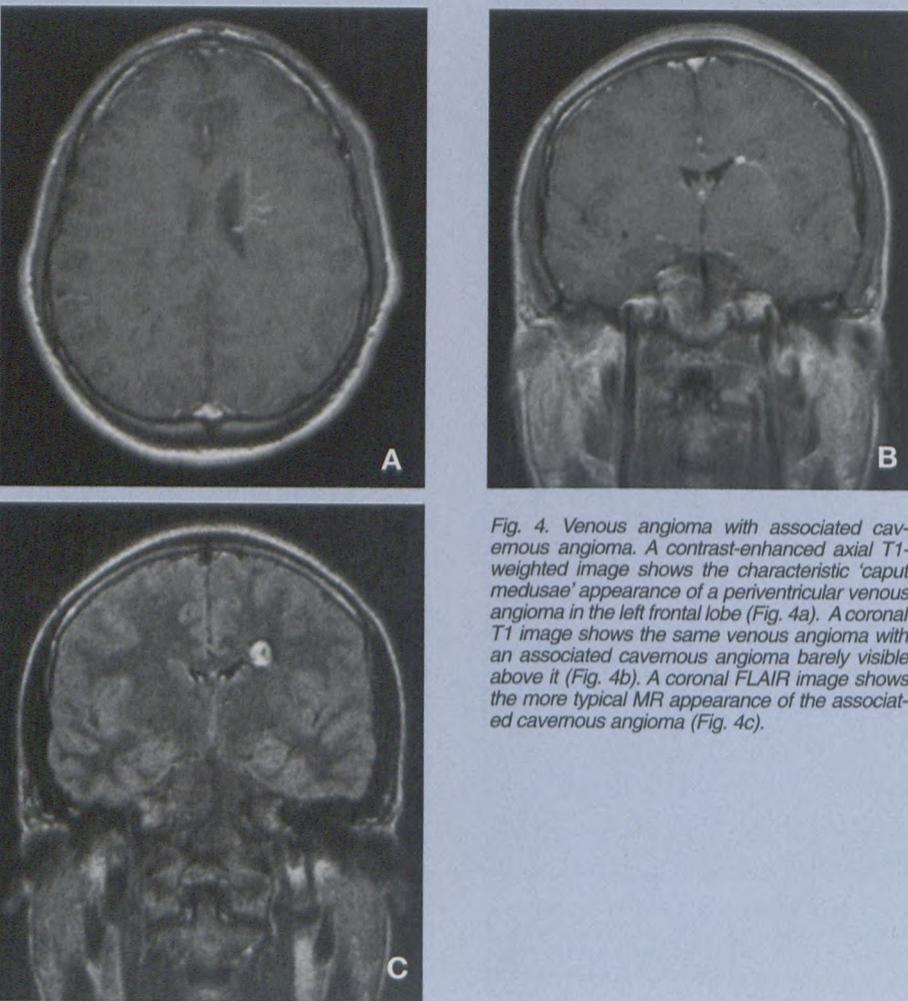


Fig. 4. Venous angioma with associated cavernous angioma. A contrast-enhanced axial T1-weighted image shows the characteristic 'caput medusae' appearance of a periventricular venous angioma in the left frontal lobe (Fig. 4a). A coronal T1 image shows the same venous angioma with an associated cavernous angioma barely visible above it (Fig. 4b). A coronal FLAIR image shows the more typical MR appearance of the associated cavernous angioma (Fig. 4c).

The association of venous angiomas with cavernous malformations suggests a common malformative mechanism, possibly with the presence of the cavernous malformation somehow triggering the development of the venous anomaly (Fig. 4). Lasjaunias<sup>18</sup> has disputed this, postulating that the haemodynamic differences created by the anomalous venous drainage may, in fact, trigger the formation of the cavernoma at a later stage. Occasionally a true arteriovenous malformation may drain into the collecting vein of a venous angioma. In a very small percentage (< 5%) of venous angiomas, associated tiny arteriovenous fistulas can be identified at angiography where small, slightly dilated arteries are seen to drain directly into the medullary veins. Mullan *et al.*<sup>19</sup> have postulated that cerebral venous malformations and arteriovenous malformations share a related malformative mechanism, and the presence of these microfistulas seen with some venous angiomas seems to lend credence to this theory. Venous angiomas may also occasionally be associated with intracranial varix with stenotic narrowing of the outlet of the collecting vein leading to ectasia of the collecting vein.<sup>20</sup>

Two groups of anomalies also found in association with venous angiomas in children include craniofacial vascular malformations and cortical dysplasias. The craniofacial vascular anomalies that have been described in association with venous angiomas include facial port-wine stains, lymphatic malformations, venous malformations, frontal varices, prominent transcranial veins and sinus pericranii.<sup>18,21</sup> The association with port-wine stains and lym-

phatic malformations appears to be coincidental as no direct causative link has yet been established. An equally rare situation is the presence of a large hemispheric venous angioma draining transcranially through a sinus pericranii defect.<sup>8</sup> A sinus pericranii defect is an osteodural venous anomaly where the intracranial venous drainage joins a midline sinus before draining externally through a skull defect directly into subcutaneous veins. The association of venous angioma with cortical migration and sulcation defects such as cortical dysplasia, pachygyria, polymicrogyria or schizencephaly suggests a link between arrested neuronal migration and failed transmedullary venous regression during early embryonic life (Fig. 5).<sup>22</sup> Yet another rare association is with the blue rubber bleb nevus syndrome where multiple venous angiomas are associated with cutaneous naevi.<sup>23</sup>

## Clinical features

The clinical behaviour of venous angiomas remains somewhat controversial. Venous angiomas have been associated with a number of clinical presentations including headache, haemorrhage, epilepsy and progressive neurological deficits. In many cases their presence is probably coincidental but in some cases may be causative. Headache is the commonest symptom associated with venous angiomas, found in up to 43% of cases.<sup>14</sup>

Their association with haemorrhage has also been somewhat controversial with reported haemorrhage rates of between 0% and 43%.<sup>17,24</sup> Historically venous angiomas have been known to be associated with haemorrhage, particularly those

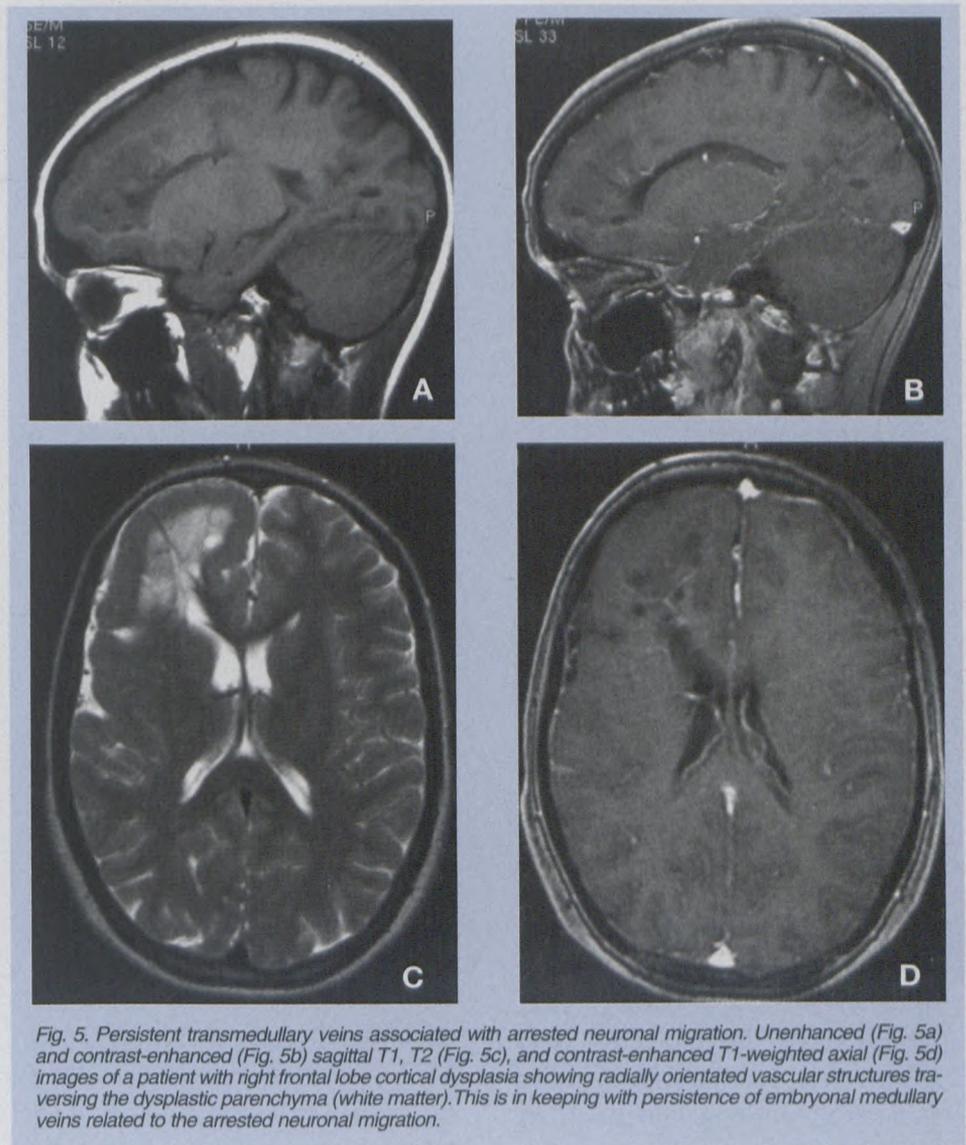


Fig. 5. Persistent transmedullary veins associated with arrested neuronal migration. Unenhanced (Fig. 5a) and contrast-enhanced (Fig. 5b) sagittal T1, T2 (Fig. 5c), and contrast-enhanced T1-weighted axial (Fig. 5d) images of a patient with right frontal lobe cortical dysplasia showing radially orientated vascular structures traversing the dysplastic parenchyma (white matter). This is in keeping with persistence of embryonal medullary veins related to the arrested neuronal migration.

occurring in the posterior fossa.<sup>25,26</sup> Higher rates of haemorrhage were undoubtedly reported in the days prior to the use of cross-sectional and in particular MR imaging. The widespread use of MR imaging has shown that the majority of venous angiomas detected are found as incidental lesions.<sup>17,27</sup> MR imaging has also shown the association of venous angiomas with other angiographically occult cerebrovascular malformations including cavernous malformations and capillary telangiectasias, particularly within the posterior fossa.<sup>28-30</sup>

Because of these associations it is felt that many of the haemorrhages related to venous angiomas probably arise from other undetected associated occult vascular malformations and not the venous angiomas themselves.<sup>30-32</sup>

There are a number of reports describing haemorrhage directly attributable to a venous angioma itself resulting from spontaneous thrombosis of the collecting vein.<sup>33,34</sup> Very rarely they may present with subarachnoid haemorrhage.<sup>1</sup>

Spontaneous thrombosis of the

collecting vein can result in venous infarction of the area drained by a venous angioma.<sup>35,36</sup>

A small number (0.4%) of patients presenting with epilepsy are shown to have venous angiomas. Although some of these are almost certainly incidental findings, the possible associated presence of cortical dysplasia or the vascular malformation may have a more direct aetiological relationship with seizure activity.<sup>37,38</sup>

Other clinical features that have been ascribed to the presence of a venous angioma include motor deficits and sensory disturbances,<sup>32</sup> trigeminal neuralgia,<sup>32,39</sup> hemifacial spasm<sup>40</sup> and hydrocephalus.<sup>41</sup> These symptoms are thought to be due to direct pressure effects of the associated venous angioma.

## Differential diagnosis

In many cases the imaging appearance of venous angiomas is relatively straightforward and pathognomic, regardless of the modality used. Dilated veins may be seen related to an arteriovenous malformation or highly vascular tumour but the associated lesion is usually apparent on sectional imaging. Dilated medullary veins may be seen in some cases of dural arteriovenous fistula with cortical venous reflux.<sup>17</sup> Visible persistent medullary veins may be seen in the area of a cortical dysplasia or other migrational disorder.<sup>22</sup>

## Treatment

The general consensus is that by themselves venous angiomas are very benign lesions. The risk of haemorrhage is generally that of any associated cavernous or capillary malformation rather than any risk from the

venous angioma itself. Occasional spontaneous thrombosis of these lesions has been described leading to venous infarction of the brain in keeping with the fact that these venous angiomas are anatomical variants which drain normal brain tissue. For this reason attempted resection of a venous angioma generally has a catastrophic outcome.<sup>16</sup> Similarly, radiosurgery is contraindicated for the management of venous angiomas.<sup>42</sup> If indicated, any direct surgical or other treatment should rather be aimed at relevant associated pathologies such as cavernous malformations or capillary telangiectasias which have a clear risk of haemorrhage, or cortical dysplasias in intractable epilepsy.

## Conclusion

Venous angiomas are extreme variants of normal brain venous anatomy, representing failure of regression of normal early embryological venous drainage pathways. They are generally benign lesions, often found coincidentally on CT or MR scans or at angiography. They can be seen in association with other intracranial or craniofacial vascular abnormalities or cerebral dysplasias. Their appearance on imaging is generally pathognomic regardless of the modality used. Confirmation of a venous angioma by angiography is generally not required, although MR scanning is probably required in most cases to exclude an associated angiographically occult vascular malformation or focal cortical dysplasia. The risk of haemorrhage is generally that of any associated occult vascular malformation. Venous angiomas may be associated with other neurological abnormalities but are very rarely the direct cause thereof. Surgical or other

direct treatment of these lesions is contraindicated and should rather be directed at any other relevant associated amenable pathology.

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