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Multiple intracranial developmental venous anomalies associated with complex orbitofacial vascular malformation

Sir,

Cerebral venous angiomas are one of the most common intracranial vascular abnormalities encountered during the course of imaging. Most are uncomplicated and surgery is unwarranted. Multiple intracranial developmental venous anomalies associated with craniofacial venous malformations are uncommon. The presence of deep venous angiomas (DVAs) in both supra and infratentorial compartments and that too in association with cervico-facial hemolymphangioma is rare. We describe the imaging of one such case.

A 16-month-old baby presented with a gradually progressive soft tissue swelling on the left fronto-temporal region extending towards the left orbit causing proptosis and facial asymmetry. On physical examination the swelling was soft and boggy, with no bruit or thrill.

Color Doppler, CT scan and MRI suggested the possibility of a slow flow venous malformation [Figure 1]. In addition, venous angiomas were detected in the posterior fossa. Sclerotherapy was attempted, with no free back flow of blood and hence the procedure was abandoned.

Six-vessel diagnostic cranial angiogram was done on which the forehead lesion did not show any abnormal vascularity. Intracranial circulation showed three venous angiomas, one in the right cerebellar hemisphere and two in the supratentorial compartment in the left parietal region [Figure 2]. All these three lesions had enlarged medullary veins converging centrally towards

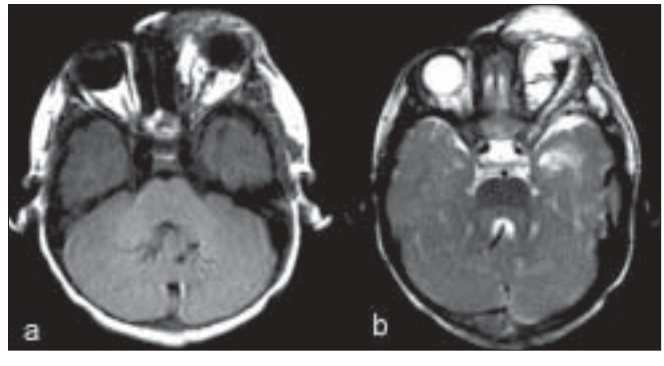

Figure 1: T1 (a) and T2 (b) weighted MR images showing left orbitofacial complex vascular malformation and right cerebellar venous angioma. The supratentorial angiomas were occult on MR imaging

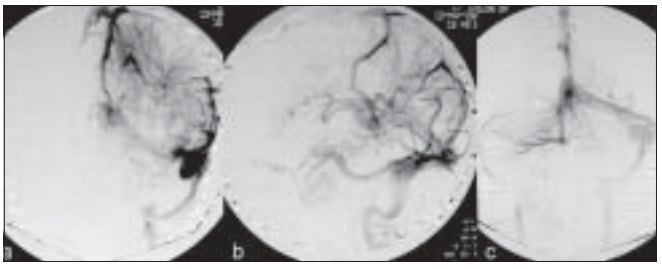

Figure 2: Venous phase of digital subtraction angiography of supratentorial (a, b) and infratentorial (c) compartments showing enlarged medullary veins converging centrally towards a deep venous structure with a typical caput medusae appearance suggestive of a venous angioma

a deep venous structure with a typical caput medusae appearance suggestive of a venous angioma. On the venous phase some under-filling of the posterior part of the superior sagittal sinus was noted with dilated veins of the galenic system suggestive of a predominant central drainage. The extracranial lesion was superficially excised with an intraoperative impression of mixed hemangio-lymphangioma, which was subsequently proved on histopathology.

Developmental venous anomalies also known as venous angiomas are the most commonly encountered intracranial vascular lesions.[1] Cerebral deep venous angiomas are frequently found in patients with vascular lesions of face orbit, including arterio-vascular malformations (AVM), capillary malformations, venous vascular malformations, orbital venous varices, sinus pericranii and some vascular tumors e.g. hemangio-lymphangioma[2]. In patients exhibiting cervicofacial venous malformation 20% have concurrent cerebral DVA.[3] The coexistence of various vascular lesions and anomalies suggests that whatever developmental factors favor one may spawn others.[2]

Bilateral DVA and DVA in both supra and infratentorial compartment are uncommon and that too in association with a complex orbitofacial vascular malformation i.e. a hemangio-lymphangioma as described in our case, is extremely rare.

The angiographic picture of venous angioma is characterized by one or more radial peripheral medullary veins converging towards an enlarged transcerebral-draining vein.[4] On MRI this lesion is hypo/isointense on T1WI and hyperintense on T2WI.

The clinical significance of DVA lies primarily in recognizing this entity as a variation of normal rather than a pathological process to prevent iatrogenic exacerbation of symptoms. Because hemorrhage is not a clinical concern and the venous structures drain normal brain parenchyma, no further therapy surgical or intervention is warranted.[5] The other concern of DVA is that planned or inadvertent occlusion during treatment of associated lesions frequently leads to venous infarction of the surrounding normal brain.[2]

In view of a notable pervasiveness of 20%, of DVA in patients with venous malformations it is important to look for a cerebral developmental venous anomaly when we come across an orbitofacial vascular malformation.

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