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Clinical manifestations and imaging findings of thrombosis of developmental venous anomalies

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ARTICLE INFORMATION

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AIM: To determine clinical manifestations, imaging findings and outcome of patients with thrombosed developmental venous anomalies (DVAs).

MATERIALS AND METHODS: The radiology database was searched retrospectively for thrombosed DVAs between 01/01/2000 to 07/01/2016. Demographic variables, associated risk factors, clinical manifestations, imaging findings, treatments, and follow-up were recorded.

RESULTS: Six patients were found (four female and two male; age range 16–45 years with mean age, 21.3 years). The most common clinical presentation was headaches followed by neurological deficits and seizures. Venous infarction, parenchymal haemorrhage, venous congestive oedema were noted as the radiological findings. Clinical outcome was favourable in all patients with complete recovery or persistence of mild neurological symptoms.

CONCLUSION: Thrombosed DVAs may occur under rare circumstances, which lead to variable symptoms. Familiarity with this entity and early recognition of associated findings including venous infarction, parenchymal haemorrhage, and venous congestive oedema would help early diagnosis and prompt treatment.

Introduction

Developmental venous anomalies (DVAs), otherwise known as venous malformations or venous angiomas, are the most common cerebral vascular malformation, reported in approximately 0.4–2.6% of the general population.[1,12] DVAs are generally regarded as benign incidental findings on imaging,[2] and are considered a variation in normal medullary veins drainage where multiple radiating medullary veins converge into a transcerebral collecting vein and ultimately drain into either deep pial or superficial cortical veins.[3] Unlike other congenital malformations, DVAs rarely produce symptoms; therefore, in most cases they are detected incidentally with little or no clinical significance.[4] A DVA can also be associated with a regional cavernous malformation (CM) in 13–40% of cases.[5,6] These CMs are responsible for the vast majority of regional complications previously attributed to DVAs themselves.[7,8] However, DVAs can occasionally give rise to symptoms secondary to various circumstances,[8] including thrombosis of the drainage vein. Thrombosis of the draining vein of a DVA is rare, and thus far only 29 cases have been reported in the literature.[9] Various complications have been reported from thrombosed DVAs including venous ischaemic infarction, parenchymal haemorrhage, as well as subarachnoid and intraventricular haemorrhage.[10]

Data regarding the clinical manifestations of thrombosed DVAs are predominately based on sporadic case reports in

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the literature. The aim of this article was to describe the clinical manifestations, imaging findings, and long-term outcome of patients with thrombosed DVAs.

Materials and methods

The study was performed following the approval of the Institutional Review Board and was compliant with the Health Insurance Portability and Accountability Act. A radiology information system keyword search was performed using MONTAGE Search and Analytics software (Montage Healthcare Solutions, Philadelphia, PA, USA) to retrospectively identify patients with thrombosed DVAs described at brain magnetic resonance imaging (MRI) examinations performed between 01/01/2000 to 07/01/2016. Search terms for DVA included: "venous angioma," "developmental venous anomaly," "venous malformation," and "DVA."

Data regarding demographic variables, such as age and gender, associated risk factors, including hypercoagulable states, and clinical/imaging follow-up were analysed. The angioarchitectural features of the DVA, including size, venous drainage, and morphology of medullary veins and collector vein were also examined. Associated imaging findings in the region of DVA including haemorrhage, parenchymal signal abnormality on T2-weighted (W) and fluid attenuation inversion recovery (FLAIR) images (vasogenic oedema), or infarction were also recorded.

Patients with at least one cerebral or cerebellar thrombosed DVA identified at MRI were included. The diagnostic criteria consisted of linear hyperdenisty on CT when available, linear susceptibility on gradient-echo (GRE) or susceptibility-weighted imaging (SWI) and lack of enhancement on contrast-enhanced T1W sequences.

Patients with other vascular malformations or other confounding lesions, such as a tumour, old infarction/haemorrhage, or surgery in the region of the DVA with indeterminate effects on radiological changes were excluded. Only vascular complications that directly linked to the thrombosed DVA and that could not be associated with other pathologies were included.

A review was conducted by two neuroradiologists to confirm thrombosed DVA and exclude other anomalies that did not fit the diagnostic criteria for thrombosed DVA.

Imaging

Computed tomography (CT) was performed on various multidetector CT systems (Siemens, Erlangen, Germany), and MRI was performed on 1.5 or 3 T systems (Siemens, Erlangen, Germany) with standard clinical protocols used at the time of the study. All MRI protocols included FLAIR, GRE/SWI, and pre- and post-contrast T1W sequences. Axial CT sections were reconstructed at 5-mm thicknesses.

All patients had routine MRI images, and unenhanced CT images. In addition, MR angiography (MRA) and MR venography (MRV) were performed in all patients to evaluate vasculature. Cerebral angiograms were obtained in three patients for verifying the presence of thrombosed DVA.

Results

Six cases were found with the principal diagnosis of thrombosed DVA. Four patients were female and two were male; ages ranged from 16 to 45 years (mean age, 21.3 years). A total of six supratentorial thrombosed DVAs were found in these patients. There were five subcortical and one periventricular or deep DVA, located in the frontal (n=3), parietotemporal (n=1), and occipital (n=2) regions. The imaging findings in case no. 3 involved the right medial parieto-occipital lobe with a DVA predominately coursing in the medial occipital lobe. DVA in case no. 3 ultimately drained into the straight sinus, and in case no. 6 into the transverse sinus. The remainder of the DVAs drained to the superficial sagittal sinus (Table 1).

Depending on DVA locations, clinical manifestations varied from asymptomatic to headache (n=4 [66.6%]), seizure (n=3 [50%]), and neurological deficits including numbness, aphasia, and visual field cut (n=3 [50%]). A hypercoagulable state was identified in three patients including oral contraceptive ingestion, pregnancy, and antiphospholipid syndrome. In addition, case no. 3 was affected by Crohn's disease. Thrombosed DVA was incidentally found in case no. 4 during work-up for concussion. The percentage of different imaging findings are summarised in Figs 2 and 3.

Of note, CT findings included normal, regional hypodensity, transmantle tubular hyperdensity, and gross intraparenchymal haemorrhage.

MRI findings consisted of linear T1/T2 shortening, regional signal abnormality, lack of enhancement in transcerebral collecting vein, FLAIR signal abnormality in converging medullary veins, engorgement of medullary veins, susceptibility blooming in collecting vein and proximal subependymal medullary veins, diffusion restriction about the draining vein, varix at the drainage point, and intra parenchymal haemorrhage.

In three cases (nos. 1, 2, 6) associated signal abnormality was attributed to a combination of cytotoxic and vasogenic oedema. Cytotoxic oedema was predominately found immediately along the thrombosed collecting vein. Intraparenchymal haemorrhage was the main feature in case no. 3, mimicking brain tumours. An underlying DVA was detected on follow-up imaging after resolution of haemorrhage. In case no. 5, the findings were initially limited to vasogenic oedema and DVA was found on follow-up after resolution of oedema. Of note, no associated findings were identified in this case on concurrently performed CT. The surrounding signal abnormality was minimal in case no. 4 with incidentally found transcerebral susceptibility blooming/thrombosis.

MRA and MRV helped to exclude other possible causes of signal abnormality and haemorrhage. On cerebral angiogram delayed filling and emptying of medullary veins were shown.

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