CASE REPORT

Paediatric presentation of intracranial haemorrhage due to thrombosis of a developmental venous anomaly

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SUMMARY

Developmental venous anomalies (DVAs) are the most commonly encountered cerebral malformations. While generally asymptomatic and discovered as incidental findings, there is a small number that can cause intracranial haemorrhage, usually attributed to associated cavernomas; however, venous thrombosis of the draining vein is a rare cause. A 10-year-old woman presented with seizure episodes. Angiographic evaluation revealed a collection of vessels draining into the superior sagittal sinus via the vein of Trolard, concerning for a DVA. The patient improved clinically with supportive care and antiepileptic treatment. Anatomically, DVAs represent dysplasia of primary capillary beds and smaller cerebral veins, resulting in abnormal venous drainage of the affected parenchyma. Several distinguishing radiological findings can help differentiate a DVA from other pathologies. Early radiological identification can help in the initiation of appropriate therapy and prevent incorrect surgical management leading to further neurological demise.

BACKGROUND

Developmental venous anomalies (DVAs)—formerly known as venous angiomas—are the most commonly encountered cerebral malformations. While generally asymptomatic and discovered as incidental findings, there is a small number that can cause complications such as mechanical compression, venous infarctions, stroke and intracranial haemorrhage.^{1–3} Intracranial haemorrhage is usually attributed to associated

cavernomas; however, venous thrombosis of the draining vein is a rare cause. We present the latter scenario, in a case of a paediatric patient with intracranial haemorrhage secondary to thrombosis of a DVA draining vein.

CASE PRESENTATION

A 10-year-old woman with a history of panhypopituitarism and mild developmental delay presented with episodes of right-sided facial twitching and increasing lethargy while at school, progressing to right hemiparesis over a 4-hour time course. She had previously been diagnosed 2 years prior with a left temporal lesion on imaging obtained for hearing loss, but had never seen a neurosurgeon and further workup was not done at that time ([figure 1](#)). Physical examination revealed facial asymmetry with a right facial droop and profound right hemiparesis. The patient was intubated due to concerns for continued seizure and monitored in the paediatric intensive care unit.

INVESTIGATIONS

Subsequently, CT was obtained, revealing a left frontotemporal hypoattenuation initially thought to be a haemorrhagic tumour, with abnormal tortuous vessels on CT angiography. MRI was similarly concerning for a large thrombosed cortical vein, prompting a catheter cerebral angiogram ([figure 2](#)). Angiographic evaluation revealed a collection of vessels draining into the superior sagittal sinus via the vein of Trolard, concerning for a DVA ([figure 3](#)). At this time, retrospective imaging review of the lesion found 2 years prior in comparison to the current imaging determined that the initial MRI 2 years prior to the acute haemorrhage event ([figure 1](#)) was likely a DVA.

OUTCOME AND FOLLOW-UP

The patient improved clinically with supportive care and antiepileptic treatment. She was treated acutely with propofol infusion for the treatment of her seizures, and subsequently started on 500 mg of levetiracetam, two times a day. Using electroencephalogram recordings, she was able to be weaned off of the propofol, until she was extubated and monitored closely in the paediatric intensive care unit. After evaluation and input from the paediatric neurology team, the most likely cause of seizure was determined to be the haemorrhage as a result of venous occlusion. Ultimately, she improved back

Figure 1 Prethrombosis event MRI scan depicting left frontal peri-insular developmental venous anomaly, found 2 years prior. (A–C) Axial T1 with gadolinium sequences, (D–F) Coronal T1 with gadolinium sequences.

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Figure 2 (A) CT scan with contrast depicting mass lesion in left frontal region. Images 1 and 2 reveal circumscribed area of hyperdensity with vascularity in the left frontal peri-insular cortex and white matter. (B) Images 1–3 are axial T2 sequences revealing areas of hyperintensity involving the left peri-insular cortical and subcortical region, with large vascular channels in the region of the Sylvian fissure. Images 4–6 are axial T1 with gadolinium sequences revealing enhancement in blood vessels throughout the left frontal region with surrounding regions of hypointensity. The white arrow in slice six points to a possible thrombus within vessels in the Sylvian fissure.

to her motor baseline and only mild residual speech deficits, and was discharged home after extensive physical therapy. Evaluation for haematological issues including hypercoagulability has remained negative, and ultimately no cause for this thrombotic event was found. After discussion with our haematology colleagues, no anticoagulation or antiplatelet therapy was started in the acute phase given the risk of acute haemorrhagic expansion and it was agreed that the thrombosis would be followed closely with imaging. Three-month follow-up MRI demonstrated recanalisation of the occluded DVA (figure 4).

DISCUSSION

DVAs are a common and typically benign finding in up to 2.5% of the population.^{1–4} Unlike other vascular malformations, the

Figure 3 Cerebral angiogram, left internal carotid injection. Lateral view depicting arterial phase (A), venous phase (B) and late venous phase (C) revealing venous anomaly. Anteroposterior view depicting arterial phase (D), venous phase (E) and late venous phase (F) revealing venous anomaly. Venous anomaly is best viewed in the venous phases, draining into the vein of Trolard (black arrow). Caput medusae can be seen in the late venous phase (black dashed line). LICA, left internal carotid artery.

Figure 4 Axial (A, B) and coronal (C) T1 with contrast MRI sequences showing occlusion, in comparison to 3-month follow-up (D–F) showing recanalisation.

intervening brain parenchyma is normal, suggesting that DVAs are a consequence of abnormal development and not an acquired pathology.^{1–3} Anatomically, these structures represent dysplasia of primary capillary beds and smaller cerebral veins, resulting in abnormal venous drainage of the affected parenchyma via the DVA. This resultant compensatory mechanism of venous drainage places the patient at risk, as spontaneous thrombosis or surgical ligation can cause venous haemorrhage or infarction due to increased venous pressure.^{1–3} Several risk factors have been identified for the development of spontaneous thrombosis of a DVA, including hypercoagulable states.⁵

Several distinguishing radiological findings can help differentiate a DVA from other pathologies. CT angiography is often the first study obtained on presentation and may show dilated linear enhancements or abnormal vasculature.⁶ MRI is often more useful and can reveal the presence of linear transcerebral flow voids, with satellite arrangement. Cerebral catheter angiography of DVAs classically reveals radiating veins (eg, ‘caput medusae’) emptying into a deep vein or dural sinus. These are seen in the same phase as normal venous drainage.^{2–4 6–9} Early radiological identification can help in the initiation of appropriate therapy and prevent incorrect surgical management leading to further neurological demise.

As with previous reports, we believe thrombosis of the draining vein was the cause for haemorrhage in our patient. While spontaneous thrombosis of the venous drainage of a DVA was first reported in 1986, haemorrhagic conversion as a result remains a rare entity, especially in the paediatric cohort. This can result in misdiagnosis and ultimately problematic management. Quick diagnosis leads to more rapid and judicious management. Treatment is often the initiation of anticoagulation therapy

or expectant observation. In cases of severe haemorrhage and elevated intracranial pressures, surgical intervention in the form of decompressive craniectomy and/or evacuation of the haemorrhage may be necessary.^{1–5 7 8 10}

We present the case of a paediatric patient who presented with intracranial haemorrhage secondary to thrombosis of the draining vein of a DVA. Initially, it was believed this patient had suffered an intratumoural haemorrhage from a primary parenchymal neoplasm. While rare, it is important to recognise that thrombosis of a DVA can result in haemorrhage in a pattern that does not conform to a usual vascular territory from a classic venous thromboembolism. Characteristic radiological features can help differentiate this pathology from a tumour. Early identification can help the initiation of appropriate therapy and prevent progression to more severe neurological deficits. The clinical outcome remains favourable in the majority of reported cases of haemorrhage as a result of spontaneous DVA venous thrombosis.

Learning points

- Developmental venous anomalies are generally asymptomatic lesions but in rare cases can thrombose and haemorrhage.
- Early radiological identification with MRI can be further supported with angiographic evaluation revealing an abnormal collection of venous drainage ('caput medusae').
- Expectant management may be an option even for haemorrhage into complicated malformations if no increased intracranial pressure is suspected.

Contributors All authors have contributed equally to the preparation and submission of this manuscript. All four authors were part of the management team for this patient. With regard to planning, conduct, reporting, acquisition of data, data analysis and interpretation, all four authors (PE, AB, JY and MA) were equally

involved. The background information was primarily researched by PE, while drafting of the text was mostly done by PE and MA. Critical review of the manuscript was done by AB and JY, with all authors reviewing and accepting the manuscript in its final form.

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