Developmental Venous Anomaly Thrombosis

- Trauma-Induced Cerebellar Edema: A Rare Presentation of Infratentorial Developmental Venous Anomaly in a Pediatric Patient
- Developmental venous anomaly coexisting with arteriovenous malformation: a case report
- Developmental Venous Anomaly Thrombosis Presenting with Intracerebral Hemorrhage
- Cavernous Venous Malformation(Archived)
- Magnetic resonance imaging findings of cerebral venous malformations
- Suprarenal interrupted inferior cava with azygos continuation in a patient with pulmonary embolism: a case report
- Draining Vein Thrombosis of Developmental Venous Anomaly in Sickle Cell Trait Patients: A Case Report and a Literature Review
- Cerebral Doppler imaging in neonates: A guide for clinical application and diagnosis

A **Developmental Venous Anomaly (DVA)**, also known as a venous angioma, is a congenital vascular malformation characterized by abnormal cerebral veins that converge into a single draining vein, often described as a “caput medusae” pattern on imaging. While DVAs are generally benign and asymptomatic, thrombosis of a DVA is a rare but potentially serious complication.

Developmental venous anomaly formerly known as venous angiomas is the most commonly encountered Brain vascular 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) 2) 3)].

Intracranial haemorrhage is usually attributed to associated cavernomas; however, venous thrombosis of the draining vein is a rare cause [4)].

Pathophysiology of DVA Thrombosis

- **Venous Outflow Obstruction**: DVAs are dependent on their anomalous venous drainage. Thrombosis in the draining vein can lead to venous congestion, reduced cerebral perfusion, and localized venous hypertension.
- **Resultant Effects**:
  - Ischemia
  - Hemorrhage (typically venous hemorrhage)
  - Edema

Clinical Presentation

Patients with DVA thrombosis may present with:

- **Headache**: Often a nonspecific but common symptom.
- **Neurological Deficits**: Depending on the location of the DVA, symptoms such as hemiparesis, aphasia, or seizures can occur.
- **Seizures**: If the thrombosis affects cortical or subcortical structures.
- **Altered Mental Status**: In cases of severe venous congestion or hemorrhage.

Diagnostic Evaluation

Neuroimaging

- **MRI and MR Venography (MRV)**: Preferred imaging modalities to identify thrombosis and assess venous drainage.
- **CT Venography (CTV)**: Useful for detecting thrombosis and associated hemorrhage.
- **Susceptibility-Weighted Imaging (SWI)**: Highlights venous structures and associated thrombi.

Findings

- Thrombus in the draining vein.
- Evidence of venous infarction or hemorrhage.
- Enlarged draining vein with associated congestion or ischemia.

Management

The management of DVA thrombosis is multifaceted and depends on the clinical presentation and complications:

Anticoagulation

- To prevent propagation of the thrombus and facilitate recanalization.
- Careful consideration is needed if intracerebral hemorrhage is present.

Symptomatic Treatment

- **Antiepileptic Drugs**: If seizures occur.
- **Management of Raised Intracranial Pressure**: Decompressive interventions may be necessary in severe cases.

Surgical Intervention

- Rarely indicated. Surgical removal of a DVA is generally avoided due to the critical nature of venous drainage.

Prognosis

- Prognosis varies based on:
    - Location and size of the DVA.
    - Presence of complications such as hemorrhage or infarction.

    - Timeliness of intervention.
- Most patients recover well with prompt diagnosis and treatment, although residual neurological deficits may persist in severe cases.

Research Directions

Given the rarity of DVA thrombosis, future studies should focus on:

- Identifying risk factors predisposing patients with DVAs to thrombosis.
- Refining guidelines for anticoagulation in cases with associated hemorrhage.
- Long-term outcomes and recurrence rates after an episode of thrombosis.

If you need further details or specific case discussions, feel free to ask!

Case reports

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 [5].

A patient who presented with focal neurological deficits and parathesia due to an infarct associated with a developmental venous anomaly with a thrombosed draining vein [6].

A case of a thrombosed developmental venous anomaly with venous congestion and pontine hemorrhage that improved after anticoagulation therapy [7].

Kiroglu et al., reported imaging findings of posterior fossa DVA with a thrombosed drainage vein in a patient with nonhemorrhagic cerebellar infarct. They also reviewed the relevant literature on the subject [8].

A patient had no associated vascular malformations, but she did have a 1-year history of oral contraceptive use and was also heterozygous for the Factor V Leiden R506Q mutation. It is likely that the combination of these thrombotic risk factors along with sluggish circulation in the DVA drainage

system allowed the thrombus to form.

Oral contraceptives were discontinued. Anticoagulation was considered, but not given due to ICH and patient preference. Long-term aspirin therapy was recommended to prevent further thrombosis. The patient recovered well and at the 3-month follow-up visit had only flattening of her right nasolabial fold [9)].

Abarca-Olivas et al., reported in 2009 two cases of brain hemorrhage secondary to developmental venous anomaly thrombosis treated at Alicante.

The first patient was a 28-year old woman on oral contraceptives treatment for a month who was referred to the Hospital with sudden-onset conscious level deterioration after presenting 24 hours previously with headache, vomits and hemiparesis. Computed Tomography revealed a predominant hypodense area containing hyperdense foci causing mild mass effect and midline shift in keeping with a hemorrhagic infarction occupying almost completely the right frontal lobe. On CT, magnetic resonance (MR) and magnetic resonance angiography (MRA) there was a prominent tubular structure adjacent to the hematoma in keeping with a partly thrombosed vessel. Urgent craniotomy and partial hematoma evacuation was performed. Digital subtraction angiography confirmed the presence of a filling defect within the draining vein of a typical caput medusae pattern developmental venous anomaly (DVA). Systemic anticoagulation was started and four days after surgery sedation was reversed and the patient awoke with normal conscious level although mild (4/5) hemiparesis persisted.

The second patient was a 38-year old male evaluated in the Emergency Department due to tonic clonic seizures in the left side followed by altered sensation in the same distribution. Initial CT revealed an intracranial bleed. After contrast administration there was an anomalous vessel in the same location that was confirmed angiographically represented a partly thrombosed DVA. Conservative management was favoured and the patient was discharged from the hospital without clinical neurological deficits [10)].

Thrombosis of developmental venous anomalies of the brain after liver transplantation [11)].

---

2 cases of spontaneous thrombosis of the draining vein of a DVA depicted on CT and MR imaging. One patient presented with a nonhemorrhagic transient ischemia, which was successfully treated with anticoagulant therapy. The second patient presented with ischemia complicated by hemorrhagic conversion [12)].

References

1)
Walsh M, Parmar H, Mukherji SK, et al. Developmental venous anomaly with symptomatic thrombosis of the draining vein. J Neurosurg 2008;109:1119–22.
2)

Dorn F, Brinker G, Blau T, et al. Spontaneous thrombosis of a DVA with subsequent intracranial hemorrhage. Clin Neuroradiol 2013;23:315–7.
3)

Agarwal A, Kanekar S, Kalapos P, et al. Spontaneous thrombosis of developmental venous anomaly (DVA) with venous infarct and acute cerebellar ataxia. Emerg Radiol 2014;21:427–30
4) , 5)

Entezami P, Boulos A, Yamamoto J, Adamo M. Paediatric presentation of intracranial haemorrhage due to thrombosis of a developmental venous anomaly. BMJ Case Rep. 2019 Jan 17;12(1). pii: bcr-2018-227362. doi: 10.1136/bcr-2018-227362. PubMed PMID: 30659007.
6)

Parker BJ, Sabb BJ. Developmental Venous Anomaly Complicated by Cerebral Venous Infarction. Radiol Case Rep. 2015 Dec 7;2(4):48. doi: 10.2484/rcr.2007.v2i4.48. eCollection 2007. PubMed PMID: 27303486; PubMed Central PMCID: PMC4895773.
7)

Griffiths D, Newey A, Faulder K, Steinfort B, Krause M. Thrombosis of a developmental venous anomaly causing venous infarction and pontine hemorrhage. J Stroke Cerebrovasc Dis. 2013 Nov;22(8):e653-5. doi: 10.1016/j.jstrokecerebrovasdis.2013.04.033. Epub 2013 Jun 19. PubMed PMID: 23791470.
8)

Kiroglu Y, Oran I, Dalbasti T, Karabulut N, Calli C. Thrombosis of a drainage vein in developmental venous anomaly (DVA) leading venous infarction: a case report and review of the literature. J Neuroimaging. 2011 Apr;21(2):197-201. doi: 10.1111/j.1552-6569.2009.00399.x. Review. PubMed PMID: 19555403.
9)

Sepelyak K, Gailloud P, Jordan LC. Thrombosis of a developmental venous anomaly with hemorrhagic venous infarction. Arch Neurol. 2010 Aug;67(8):1028. doi: 10.1001/archneurol.2010.176. PubMed PMID: 20697060; PubMed Central PMCID: PMC2920504.
10)

Abarca-Olivas J, Botella-Asunción C, Concepción-Aramendía LA, Cortés-Vela JJ, Gallego-León JI, Ballenilla-Marco F. [Two cases of brain haemorrhage secondary to developmental venous anomaly thrombosis. Bibliographic review]. Neurocirugia (Astur). 2009 Jun;20(3):265-71. Spanish. PubMed PMID: 19575131.
11)

Ballarin R, Di Benedetto F, De Ruvo N, Masetti M, Montalti R, Spaggiari M, Longo C, Gerunda GE. Thrombosis of developmental venous anomalies of the brain after liver transplantation. Transplantation. 2009 Feb 27;87(4):615-6. doi: 10.1097/TP.0b013e3181963beb. PubMed PMID: 19307801.
12)

Walsh M, Parmar H, Mukherji SK, Mamourian A. Developmental venous anomaly with symptomatic thrombosis of the draining vein. J Neurosurg. 2008 Dec;109(6):1119-22. doi: 10.3171/JNS.2008.109.12.1119. PubMed PMID: 19035729.