



Developmental Venous Anomaly with Adjacent Parenchymal Abnormality Masquerading as Glioma-A Unique Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

ABSTRACT

Developmental venous anomaly (DVA)/ Cerebral Venous Angioma is the most common type of cerebral vascular malformations, mostly an incidental benign finding. But there are documented associated complications like parenchymal signal abnormalities, thrombosis, cavernous malformations and parenchymal atrophy.

In this report, we present a unique case of cerebral DVA with cortical changes mimicking the Glioma. Altered haemodynamics in DVA is the underlying pathophysiology for these changes. Correct MRI interpretation, by an expert neuroradiologist, can establish the diagnosis of DVA and its related changes. Therefore, it can reduce the morbidity and mortality by preventing the unnecessary invasive procedures like biopsy for diagnosis.

Keywords: Cerebral venous angioma; glioma.

1. INTRODUCTION

Among the different types of cerebral vascular malformations, developmental venous anomaly (DVA) is the most common type incidentally encountered on enhanced CT or MRI of the brain [1,2,3]. Most of the times DVA is an incidental benign finding with no other associated abnormality. However, there are documented related complications including, in Magnetic Resonance Imaging (MRI), parenchymal signal abnormalities, thrombosis with venous infarction, cavernous malformations and parenchymal atrophy [4].

In this report, we present a unique case of cerebral DVA with cortical changes mimicking a Glioma. Parenchymal hyperintense signal abnormalities on T2 and FLAIR sequences are usually seen in the deep white matter in the drainage territory of DVA's. There are a few reported cases of cortical involvement with associated parenchymal atrophy [1,2,5], nevertheless there is no such reported case with marked cortical thickening and intense gyriform lobar enhancement, provided by contrast media uptake, mimicking a neoplastic lesion.

2. CASE PRESENTATION

Was a 25 year old male patient presented in emergency department of Aga Khan university hospital on January 7th, 2017 with the complains of seizures, vomiting and headache for the last 3 days. On examination he was lying in bed comfortably, alert and oriented to surroundings. The parameters registered were blood pressure of 106/64 mmHg, heart rate 78 /min, temperature 36.6 C, respiratory rate 16 /min and oxygen saturation was 94%. No significant malfunction was found on systemic examination of cardiovascular, respiratory, gastrointestinal and central nervous systems. Emergency treatment was given.

His initial laboratory workup showed increased total leucocyte count of 14.3×10^9 cells per liter with raised neutrophils i-e 86.7%. The renal function tests and blood coagulation profiles were normal. Blood glucose levels were in normal range. No additional laboratory workup was done. The CT Head without contrast showed marked vasogenic oedema in right frontal lobe with a suspected thrombosed vessel in it. There was a linear hyperdense structure arising from the centre of the area of oedema and extending up to the superior sagittal sinus, likely

representing thrombosed vessel. Left side of the brain showed normal grey and white matter differentiation.No acute intracranial haemorrhage was found. There was no midline shift or hydrocephalus.Ventricles, basal cisterns and extra-axial CSF spaces were preserved. (Fig. 1) Patient was then admitted in ward under neurosurgery care.

2.1 Further Radiological Work up

CT angiography was applied later on same day that showed multiple abnormal bunch of small vessels in right frontal lobe with a large vein draining into the superior sagittal sinus, these vessels were giving typical caput medusae appearance consistent with developmental venous anomaly. Anterior circulation showed both internal carotids, anterior and middle cerebral arteries which are patent and normal in calibre. Posterior circulation showed vertebral, basilar and posterior cerebral arteries which were also patent. No definite large vessel occlusion/stenosis or aneurysmal dilatation seen on either side (Fig. 2).

Two days later MRI Brain with contrast and MR Spectroscopy were done on which the initial diagnosis of DVA was confirmed and additional abnormalities were found in the right frontal cortex. Large developmental venous anomaly with umbrella-like Medusa head of enlarged medullary (deep white matter) veins converging on a dilated transcortical collector vein, was draining into the superior sagittal sinus.Cortical thickening with intense gyriform enhancement noted in the right frontal lobe. T2/FLAIR hyperintense area was noted in the right frontal lobe white matter representing oedema. Overall imaging appearances were suggestive of a large developmental venous anomaly with altered haemodynamics. (Figs. 3 and 4)

Spectroscopy of the frontal lobe thickened enhancing cortical area was done that was not suggestive of glioma (Fig. 5).

2.2 Diagnosis

Developmental venous anomaly (DVA) with parenchymal changes secondary to altered haemodynamics.

2.3 Treatment

The patient was managed conservatively. Occupational physiotherapy was done. He was

ambulated with the help of physiotherapist. With this treatment, patient got better. Remained stable, seizureless and afebrile, therefore was discharged home after 3 days on 10th of January 2017.

2.4 Follow Up

Follow up visits were in VIR and neurosurgery clinics.

Digital Subtraction Angiography (DSA) was done on 05 May, 17.

On DSA there was a bunch of small fine veins in right frontal lobe draining in a centripetal pattern into a large venous trunk draining into superficial vein which was connecting with superior sagittal sinus. Findings were consistent with DVA. Left internal carotid and vertebral angiograms were unremarkable. No aneurysm identified and there was no evidence of spasm in the major vessels (Figs. 6 and 7).

Follow Up MRI, MRA & MRV BRAIN was done after 6 months on 24th of July, 17.

There was redemonstration of DVA converging on a dilated transcortical collector vein, which drains into the superior sagittal sinus. In comparison to the previous examination there was slight reduction in the diameter of the DVA with interval development of gliosis in the right frontal lobe. There was also improvement in vasogenic oedema. There was no cerebral venous sinus thrombosis. (Fig. 8 and 9)

Last Follow up was on Aug 10, 2017 in **VIR CLINIC**. He remained stable, seizureless and afebrile during this interval.

3. DISCUSSION

DVA, also known as venous angioma but as this term gives the nuance of a neoplasm so, Huang et al suggested the term medullary venous malformation for this condition in 1984 [6]. Lasjaunias et al coined the term developmental venous anomaly (DVA) in 1986. They regarded DVA as variant of the medullary venous system [7].

These are actually dilated medullary veins draining the brain parenchyma into enlarged transcortical or subependymal veins and are classified as superficial and deep drainage types

according to the drainage of central medullary vein into dural sinus or Galenic system respectively [8].

The possible mechanism of formation of DVA is the activation of compensatory mechanism secondary to venous occlusion of the superficial or deep venous drainage system during intrauterine life or after birth giving an alternative route for venous drainage [9,10].

These can be seen anywhere, however the most common locations are frontoparietal region (36-64%) and cerebellar hemisphere (14-27%) DVA is solitary in 75% of the cases but in approximately 20% of cases there can be mixed vascular malformation due to the presence of cavernous malformations [11].

On unenhanced CT the draining vein if large/thrombosed can be seen. However confirm diagnosis is made on contrast CT on which it appears as enhancing linear or curvilinear structure [12]. On MRI, they may show flow void on T1- and T2-weighted images. When small it might not be detected on MRI. At contrast MRI, DVA is often visible with post contrast T1 and susceptibility weighted imaging (SWI) as the main tools for evaluation [13]. At angiography (venous phase) bunch of dilated medullary veins converging into transcerebral or subependymal collector vein is seen, giving the pathognomonic medusa like appearance. Arterial phase is usually normal with occasional late capillary blush [14].

DVA is usually benign and are unlikely to become symptomatic. But there are reported complications like parenchymal signal abnormalities, thrombosis with venous infarction, cavernous malformations, intracerebral hemorrhage and parenchymal atrophy. Hemorrhage occurs in association with cavernous malformations which are prone to bleeding. It can also result from the thrombosis with increased pressure of the draining vein. [15].

Increased signal-intensity on T2-FLAIR images in the areas drained by DVA is not a rare finding on brain imaging studies with an incidence of 7.8%–54.1% on MRI with an increasing prevalence with advancing age [8].

Such abnormal SI has been explained as edema, ischemia, demyelination, gliosis, leukoariosis, or a combination of these conditions [15,16].

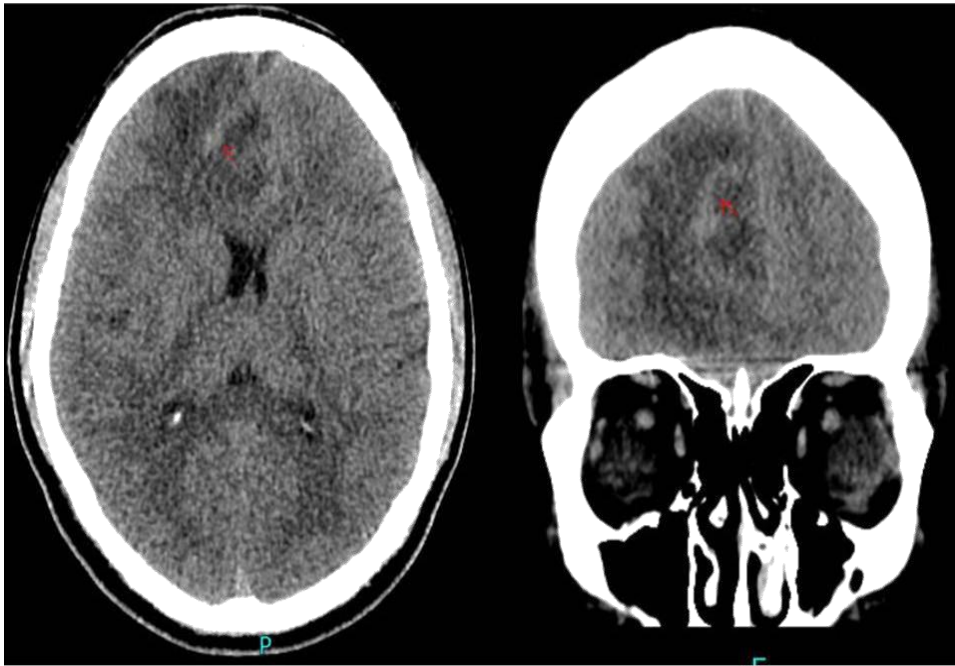


Fig. 1. Unenhanced CT brain axial sections

Red arrow shows a dense linear structure with surrounding hypo dense area in right frontal lobe. Appearance are likely of an abnormal large vessel with surrounding edema

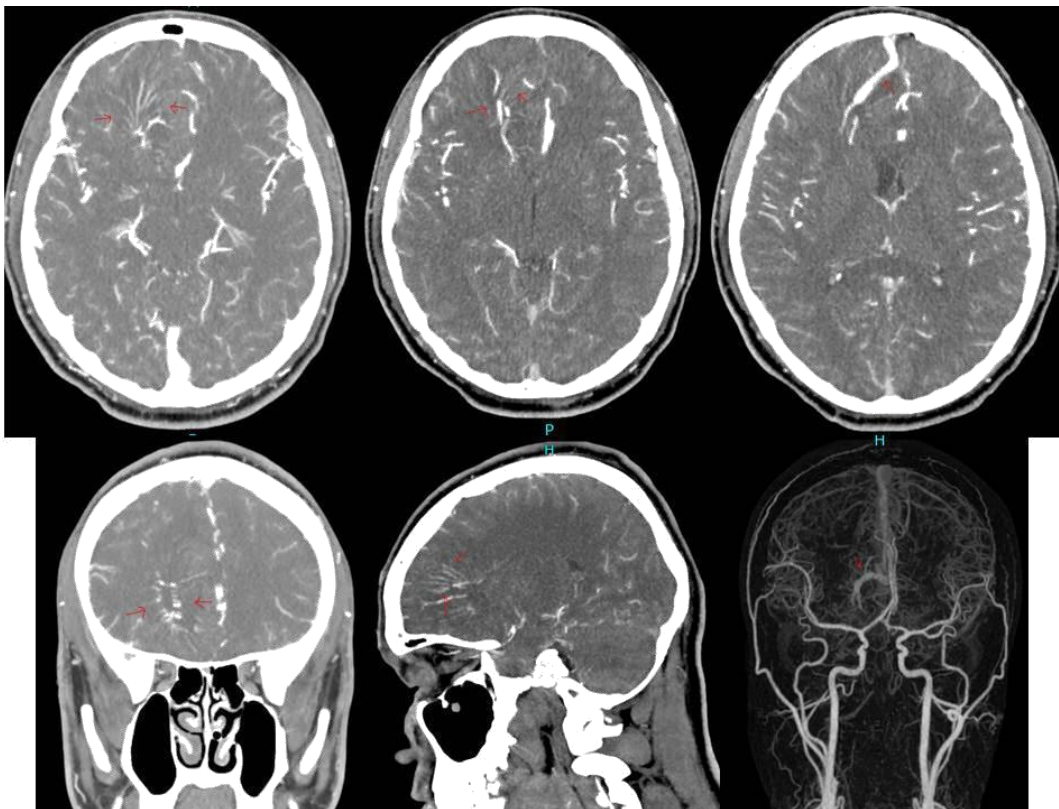


Fig. 2. CT Angiography axial, coronal and sagittal images

Red arrows are pointing to multiple medullary vessels draining into a large vessel in right frontal lobe giving typical medusa head appearance consistent with DVA

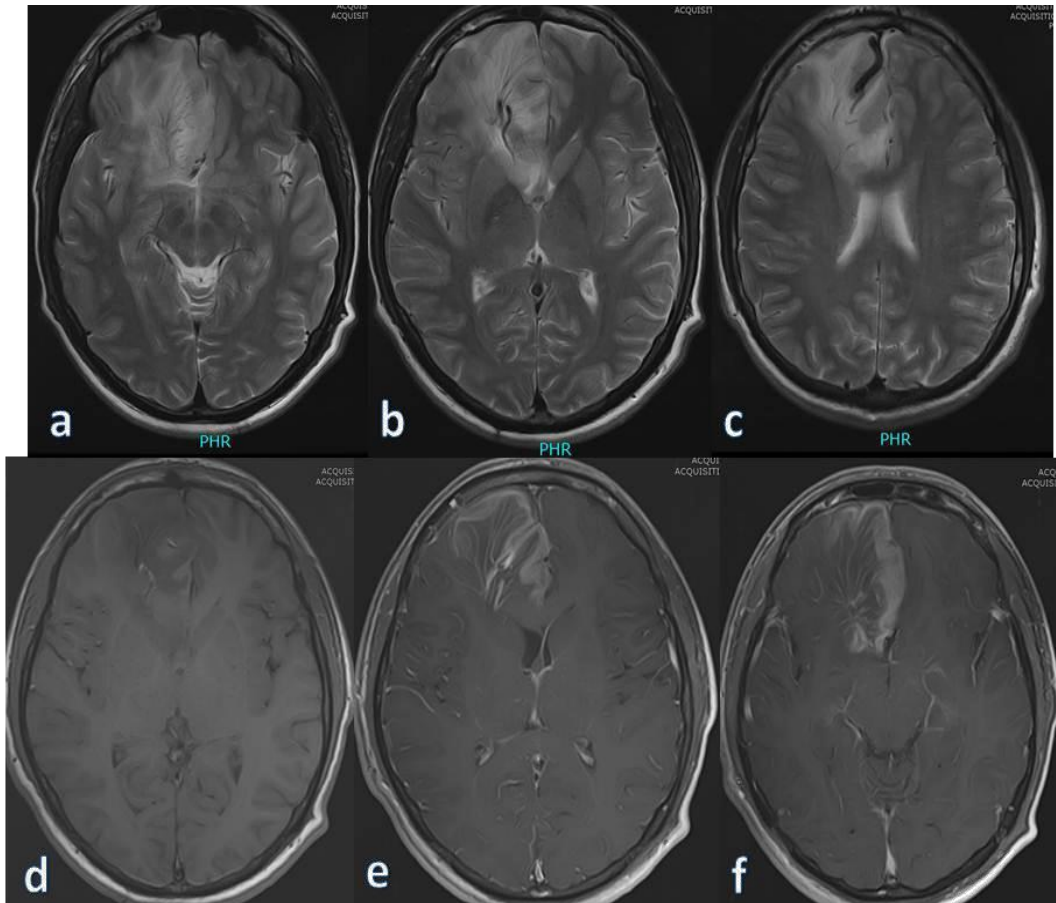


Fig. 3. a,b,c= Axial T2WI, d=Pre contrast T1WI, e,f=Post contrast T1WI

A large DVA with umbrella-like Medusa head of enlarged medullary veins converging on a dilated transcortical collector vein, which drains into the superior sagittal sinus. There is cortical thickening with intense gyriform enhancement in the right frontal lobe. T2/FLAIR hyperintense area is noted in the right frontal lobe white matter representing oedema. Overall appearances are suggestive of a large developmental venous anomaly with altered haemodynamics

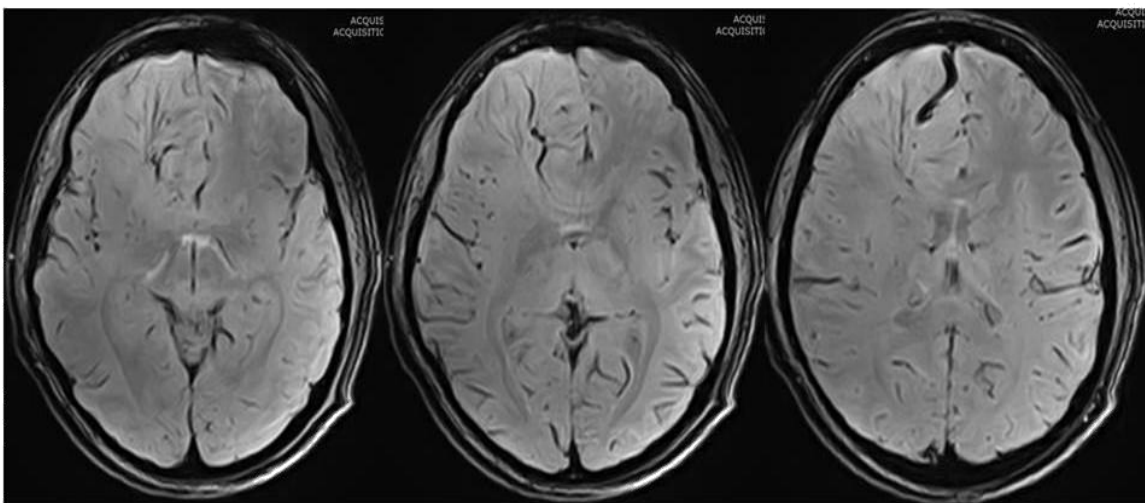


Fig. 4. Susceptibility weighted images (SWI) show signal drop out in DVA and its draining medullary veins

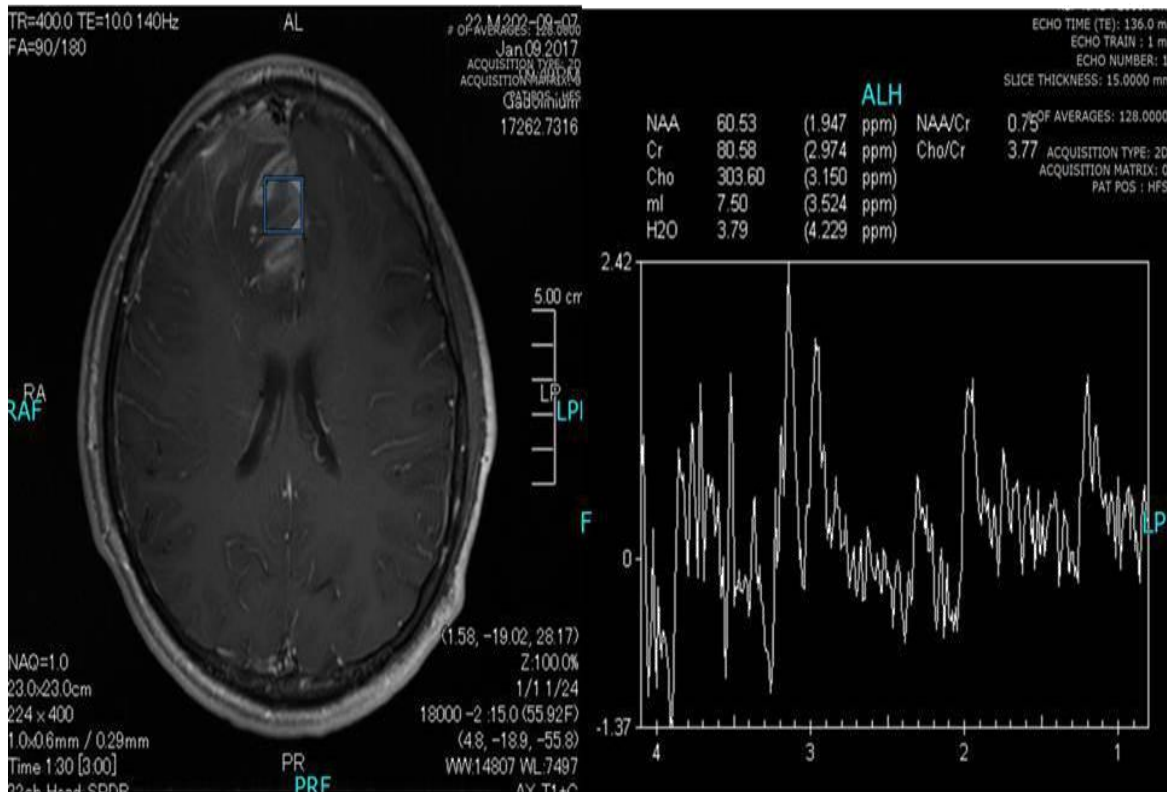


Fig. 5. Magnetic resonance spectroscopy (MRS)

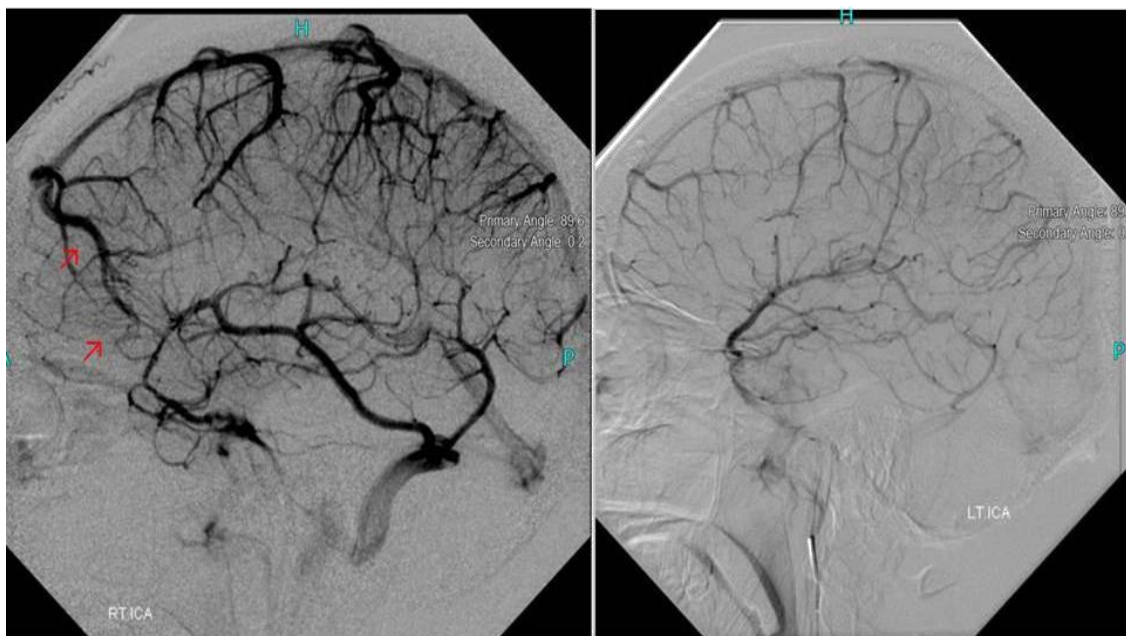


Fig. 6. Right and left internal carotid artery (ICA) digital subtraction angiography (DSA)
*A bunch of small fine veins in right frontal lobe draining in a centripetal pattern into a large venous trunk draining into superficial vein which is connecting with superior sagittal sinus . Findings representing DVA. Red arrows show right sided DVA.
 Left internal carotid and vertebral angiograms are normal. No aneurysm or spasm in the major vessels*

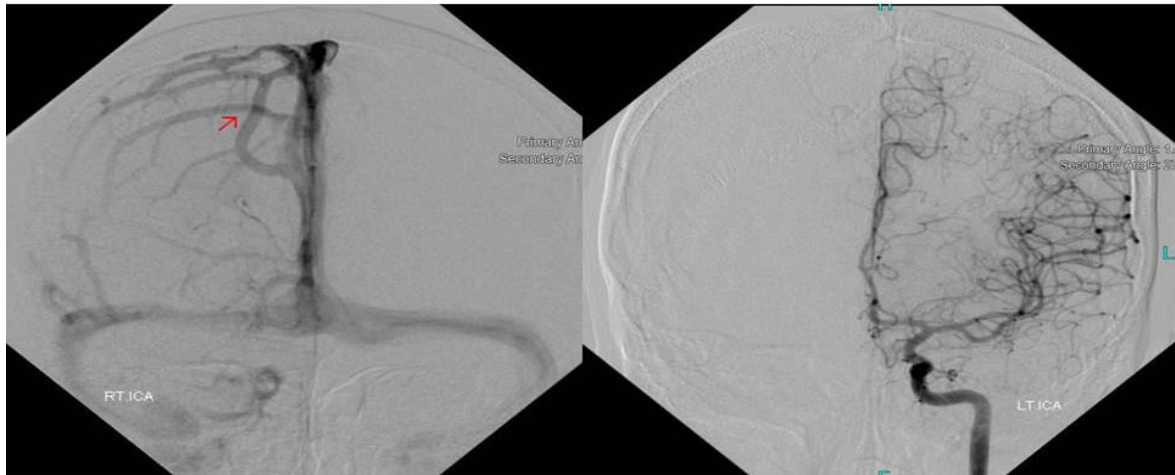


Fig. 7. Right and left internal carotid artery (ICA) digital subtraction angiography (DSA)
Red arrows show right sided DVA

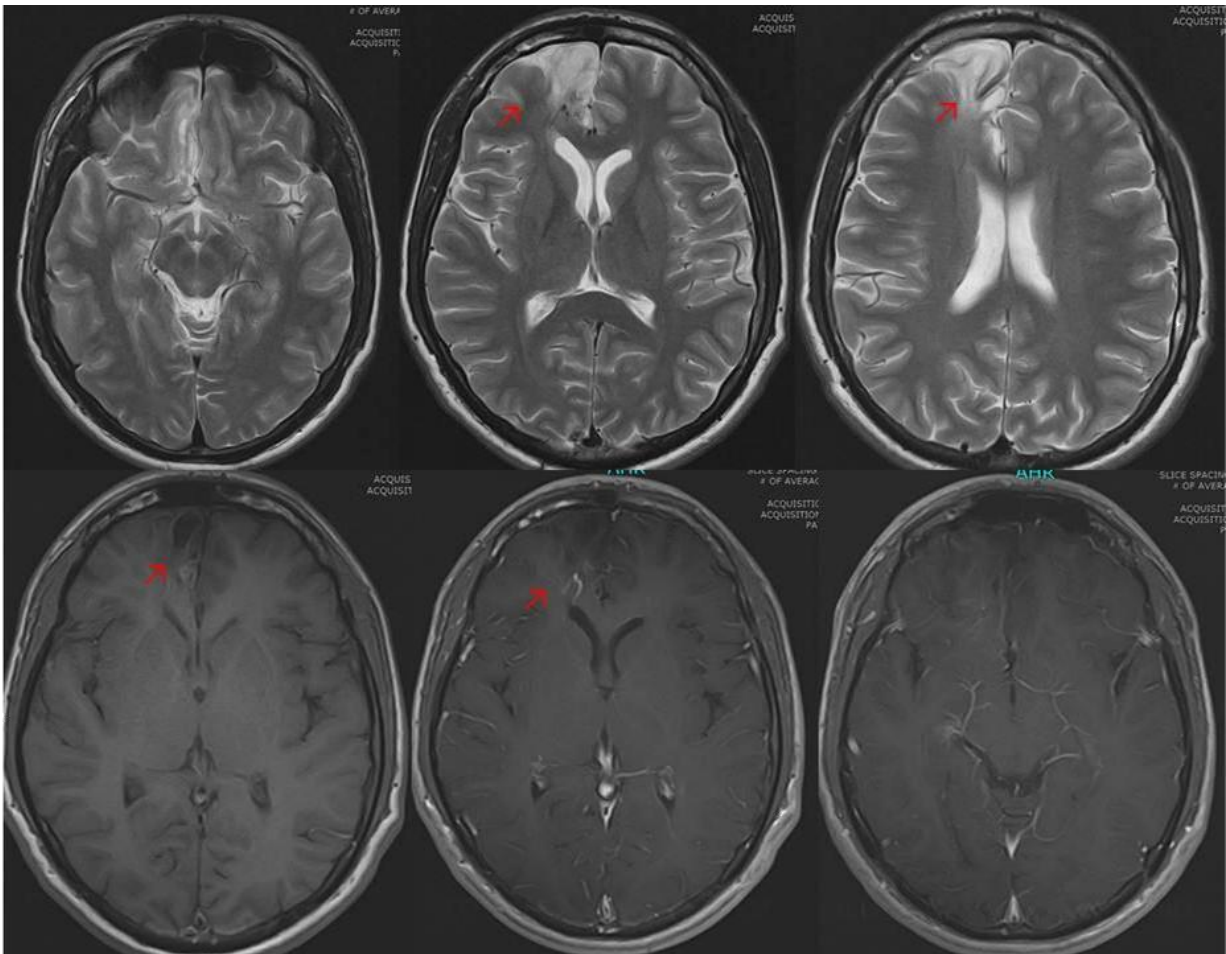


Fig. 8. Axial T2WI, pre contrast T1WI and post contrast T2WI. Red arrows pointing the right sided DVA

DVA with veins converging on a dilated transcortical collector vein, which drains into the superior sagittal sinus. In comparison to the previous examination there is reduction in the diameter of the DVA with interval development of gliosis in the right frontal lobe. Improvement in edema, mass effect and midline shift



Fig. 9. Susceptibility weighted images (SWI) show signal drop out in DVA and its draining veins

On diffusion and perfusion MRI, it gives high apparent diffusion coefficient (ADC), increased regional cerebral blood volume, and a perfusion delay is seen in the drainage area of DVA representing venous congestion [8,17,18].

The pathogenesis of parenchymal signal abnormalities associated with DVA is incompletely understood.

The most common hypothesis is chronic venous hypertension/insufficiency. Changes such as thickening and hyalinization of vessel walls [19] or focal stenosis of the draining vein at the site of its entry into the dural sinus cause alteration in haemodynamics with impaired cerebral blood flow, volume overload, venous congestion and chronic cerebral edema or ischemia. [20]. So, alteration of hemodynamic factors influence parenchymal manifestations of DVA. And this hypothesis clearly explains the abnormal parenchymal signals that lead to the onset of seizures in our patient.

4. CONCLUSION

Diagnosis of DVA and its associated complications can be made confidently on non invasive imaging- MRI especially post contrast T1WI and SWI sequences are the key tools.

In DVA parenchymal signal abnormality is common in white matter secondary to altered haemodynamics but it can involve the grey matter as well with cortical thickening and enhancement masquerading a glioma as in our case. Clinical presentation i.e seizures, headache and vomiting etc can also mislead to the diagnosis of glioma. So, correct MRI interpretation by an expert neuroradiologist can

prevent unnecessary invasive procedures like biopsy to establish the diagnosis.

CONSENT

As per international standard informed and written participant consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard written ethical permission has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Ruíz DS, Delavelle J, Yilmaz H, Gailloud P, Piovan E, Bertramello A, Pizzini F, Rüfenacht DA. Parenchymal abnormalities associated with developmental venous anomalies. *Neuroradiology*. 2007;49(12):987-95.
2. Linscott LL, Leach JL, Zhang B, Jones BV. Brain parenchymal signal abnormalities associated with developmental venous anomalies in children and young adults. *American Journal of Neuroradiology*. 2014;35(8):1600-7.
3. Umino M, Maeda M, Matsushima N, Matsuura K, Yamada T, Sakuma H. High-signal-intensity abnormalities evaluated by 3D fluid-attenuated inversion recovery imaging within the drainage territory of developmental venous anomalies

- identified by susceptibility-weighted imaging at 3 T. Japanese journal of radiology. 2014;32(7):397-404.
4. Takasugi M, Fujii S, Shinohara Y, et al. Parenchymal hypointense foci associated with developmental venous anomalies: evaluation by phase-sensitive MR imaging at 3T. *AJNR Am J Neuroradiol.* 2013;34:1940–44.
 5. Santucci GM, Leach JL, Ying J, et al. Brain parenchymal signal abnormalities associated with developmental venous anomalies: detailed MR imaging assessment. *AJNR Am J Neuroradiol.* 2008;29:1317–23.
 6. Huang Y, Robbins A, Patel S, Chaudhary M. Cerebral venous malformations and new classification of cerebral vascular malformations. New York, NY: Grune & Stratton; 1984.
 7. Lasjaunias P, Burrows P, Planet C. Developmental venous anomalies (DVA): the so-called venous angioma. *Neurosurg Rev.* 1986;9(3):233–242.
 8. Jung HN, Kim ST, Cha J, Kim HJ, Byun HS, Jeon P, Kim KH, Kim BJ, Kim HJ. Diffusion and perfusion MRI findings of the signal-intensity abnormalities of brain associated with developmental venous anomaly. *American Journal of Neuroradiology.* 2014;35(8):1539-42.
 9. Taoka T, Fukusumi A, Miyasaka T, Kawai H, Nakane T, Kichikawa K, Naganawa S. Structure of the medullary veins of the cerebral hemisphere and related disorders. *Radiographics.* 2017;37(1):281-97.
 10. Okudera T, Huang YP, Fukusumi A, Nakamura Y, Hatazawa J, Uemura K. Micro-angiographical studies of the medullary venous system of the cerebral hemisphere. *Neuropathology.* 1999;19:93–111.
 11. Lee C, Pennington MA, Kenney CM. MR evaluation of developmental venous anomalies: medullary venous anatomy of venous angiomas. *AJNR Am J Neuroradiol.* 1996;17 (1):61-70.
 12. Dehkharghani S, Dillon WP, Bryant SO, Fischbein NJ. Unilateral calcification of the caudate and putamen: Association with Underlying Developmental Venous Anomaly. *American Journal of Neuroradiology.* 2010;31(10):1848.
 13. Hammoud D, Beauchamp N, Wityk R, Yousem D. Ischemic complication of a cerebral developmental venous anomaly: case report and review of the literature. *Journal of computer assisted tomography.* 2002;26(4):633-6.
 14. Hon JM, Bhattacharya JJ, Counsell CE et al. The presentation and clinical course of intracranial developmental venous anomalies in adults: a systematic review and prospective, population-based study. *Stroke.* 2009;40 (6):1980-5.
 15. Hong YJ, Chung TS, Suh SH, et al. The angioarchitectural factors of the cerebral developmental venous anomaly: can they be the causes of concurrent sporadic cavernous malformation? *Neuroradiology.* 2010;52:883–91
 16. Rammos SK, Maina R, Lanzino G. Developmental venous anomalies: current concepts and implications for management. *Neurosurgery* 2009;65:20-29, discussion 29–30.
 17. Kroll H, Soares BP, Saloner D, et al. Perfusion-CT of developmental venous anomalies: typical and atypical hemodynamic patterns. *J Neuroradiol.* 2010;37:239–42.
 18. Sharma A, Zipfel GJ, Hildebolt C, et al. Hemodynamic effects of developmental venous anomalies with and without cavernous malformations. *AJNR Am J Neuroradiol.* 2013;34:1746–51.
 19. Galambos C, Nodit L. Identification of lymphatic endothelium in pediatric vascular tumors and malformations. *Pediatr Dev Pathol.* 2005;8:181–89.
 20. Pereira VM, Geibprasert S, Krings T, et al. Pathomechanisms of symptomatic developmental venous anomalies. *Stroke* 2008;39:3201–15.

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