Case report

Cerebellar developmental venous anomaly with associated cavernoma causing a hemorrhage – a rare occurrence

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A B S T R A C T

Developmental venous anomalies (DVAs) are variations in the transmedullary veins required for drainage of the brain. Normally, when occurring in isolation, DVAs are not clinically significant and are therefore usually a benign diagnosis. Thus, they are most often an incidental finding unless associated with an adjacent pathology. However, intracranial haemorrhage induced by a DVA alone can rarely occur and has been scarcely reported. In this case report we discuss a 58-year-old woman who presented with signs and symptoms of a cerebellar syndrome. Following a non-contrast CT, a CT angiogram and MRI contrast scan of the brain, she was found to have a cerebellar DVA and an intracranial haemorrhage. Subsequent imaging 3 months later with CT and MRI redemonstrated additional evidence of a cavernoma. The patient was managed conservatively.

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Introduction

Developmental venous anomalies (DVA) have been referred to by a variety of names including venous angiomas and cerebral venous malformations. DVAs are congenital malforma-

tions that consist of dilated medullary veins that join to form a single channel, draining into the superficial and deep drainage systems.

They are the most common type of vascular malformation occurring in the brain and present in up to 3% of the population [1]. They are widely considered to be congenital in origin.

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and there are many theories regarding the underlying pathology including the idea that DVAs are an embryological alteration of venous drainage rather than a true vascular abnormality [2].

DVAs are often encountered incidentally as they tend to be asymptomatic and follow a benign course. Neurological symptoms in patients seen are therefore often due to the coexistence of an adjacent pathology, most commonly a cavernoma [3], fistula or aneurysm when the underlying cause is intracranial hemorrhage (ICH). DVAs can also be symptomatic due to mechanical or flow related mechanisms. However, an ICH can be found in close proximity to a DVA and accompanying pathology is not readily identifiable.

In this report, we discuss the case of a 58-year-old woman, who presented with signs of cerebellar syndrome, including the clinical presentation and imaging findings.

Clinical presentation

A 58-year-old woman presented to a district general hospital with a 1-week history of an acute cerebellar syndrome. Her symptoms were diplopia, dizziness, tinnitus, headache, and vomiting. On examination she exhibited past pointing to the right side, dysdiadochokinesia, nystagmus, and ataxia.

She is an ex-smoker with a past medical history of fibromyalgia, hypertension, and depression.

As part of her work-up she was investigated with a non-contrast Computed tomography (CT) of the brain, CT angiogram and Magnetic Resonance Imaging (MRI) pre and post-contrast during her admission.

Unenhanced CT (Fig. 1) demonstrated a hyperdense focus of acute parenchymal hemorrhage within the right cerebellar hemisphere adjacent to the middle cerebellar peduncle. No obvious evidence of an underlying lesion was found. Reviewing the bone window raised suspicion of a high density within, which may reflect a focus of calcification. The case was discussed further with the tertiary Neurosurgery center and further investigations, with CT intracranial angiogram and contrast enhanced MRI, were advised due to the atypical location of the hemorrhage raising suspicion of an underlying lesion.
A CT intracranial angiogram (Figs. 2, 3 and 4) was subsequently performed with the standard protocol at 120 KVP and with Omnipaque 300 intravenous contrast. This revealed evidence of a developmental venous anomaly with no evidence of an associated cavernoma.

The patient went on to have a contrast enhanced MRI of the brain (GE 1.5T Artist and 10 mL of Clariscan intravenous contrast). The standard brain sequences were acquired as per the local protocol: T2w, T2 FLAIR, pre- and post-contrast volume T1w (Figs. 5 and 6), susceptibility weighting images (SWI) (Fig. 7) and diffusion weighted images. The study confirmed the findings of a cerebellar hemorrhage with an adjacent DVA. This was best demonstrated on the T1w post contrast images. It appeared as a flow void on unenhanced T1w and T2w images. It was visible on SWI images with blooming artefact also seen to arise from the nearby acute hemorrhage. No definite associated cavernoma or any other cavernomas were found on initial imaging. No evidence of any other intracranial lesion or territorial infarct was found. The patient’s symptoms resolved, and she managed conservatively. She was given safety netting advice to re-present if symptoms recur.

The patient re-presented 3 months later due to continuing intermittent headache and went to have further imaging. She was initially investigated with an unenhanced cranial CT which showed interval resolution of the previous bleed. The focus of calcification seen at the site of the previous hemorrhage was redemonstrated, raising suspicion of a cavernoma (Fig. 8).

Further imaging with MRI was performed with T2w, T2 FLAIR, volume T1w, DWI/ADC and SWI sequences. A signal void was seen at the site of the previous hemorrhage with an eccentric signal focus on T1w images (Fig. 9). No T1w hyperintensity to suggest residual subacute hemorrhage contributing to the signal was seen. Blooming artefact was demonstrated on SWI images (Fig. 10). Features were in keeping with an associated cavernoma. The final diagnosis was DVA associated with a cavernoma which was managed conservatively.
Fig. 7 – SWI sequence: The DVA is again well seen. Extensive blooming artefact is seen associated with the recent hemorrhage.

Fig 8 – Repeat axial CT head: resolution of the previous hemorrhage with a focus of calcification in the middle cerebellar.

Fig. 9 – Repeat T1w volume: The DVA is again demonstrated with a nearby low signal focus. No areas of high signal to indicate.

Fig. 10 – Repeat SWI: Site of cavernoma indicated by green arrow. A blooming artefact with hypointense limb demonstrating.
Discussion

Developmental venous anomalies can be well imaged with cross sectional imaging. We discuss the imaging findings below.

Developmental venous anomalies are not well demonstrated on unenhanced CT unless they are large. It is of note that interpretation in the posterior fossa can be limited by beam hardening artefact arising from the skull base [4]. DVAs and cavernomas have been known to occur together in patients with an ICH [5-7]. For example, the presence of vascular calcifications in an unusual location within an accompanying cavernoma can provide a clue as to their presence [4].

DVAs are usually well demonstrated on CT angiograms, with the primary venous collector of the DVA appearing as a linear or arcuate enhancement with multiple branches, giving rise to the classic “caput medusae” or “palm tree” appearance. This anomaly will usually start in the white matter and connect to a venous sinus, deep or cortical vein as demonstrated on these images.

Typical appearances of a DVA on unenhanced T1w and T2w sequences are flow voids in the white matter traversing towards the cortex with a possible connection to the cerebral venous system. They are typically linear and/or curvilinear or round depending on the imaging plane. It is important to note that small DVAs may not be well seen on standard unenhanced sequences [4].

DVAs are characterized by low flow and low resistance hemodynamics and consequently exhibit strong post contrast T1 enhancement. The primary collector vein is shown as an arcuate enhancement from white matter to a connection with the dural or subependymal venous system. The constituent veins in the DVA give rise to a spoke-wheel appearance with peripheral veins gradually enlarging as they approach the collector vein, typically referred to as a “caput medusae” appearance [8].

In cases with an associated cavernoma susceptibility weighted sequences are most useful in detecting this lesion due to blooming artefacts and can exhibit punctate or popcorn calcifications. SWI has been shown to have greater sensitivity than T2*-weighted imaging [9]. SWI signal is not compromised by low-velocity venous flow and therefore is well demonstrated [9].

Typically, DVAs occur as individual lesions [3] and, occur most commonly in the frontoparietal region, with a reported range of 36%-64%, whereas in the cerebellum this has been reported to be 14%-27% [1].

Appearances of a developmental venous anomaly are generally characteristic. In cases confounded by intracranial haemorrhage, special attention should be paid to an accompanying pathology such as cavernoma or associated arteriovenous malformation [3,7].

DVAs are most often encountered incidentally in brain scans when there is a nearby pathology, such as a cavernous malformation (which may result in a haemorrhage) [10], or a thrombosis obstructing normal flow in the venous drainage system [4].

DVAs generally tend to follow a benign course unless occurring with an adjacent pathology, except in very rare circumstances where patients may present with symptoms such as headache, dizziness or epilepsy [11,12]. They may also present with a neurological deficit due to ICH. One study of 63 patients, of which 2 had an ICH attributed to a DVA, demonstrated an annual haemorrhage risk of 0.15% per DVA [11]. Whereas, another study of 100 patients demonstrated bleeding due to a DVA in one patient – resulting in an annual haemorrhage risk of 0.22% per year [10].

Cerebellar DVAs have rarely been reported to be associated with haemorrhage [13-21]. In one study, of 32 patients with cerebellar DVAs, none resulted in intracranial haemorrhage [20]. Whereas, another study demonstrated that of the 10 patients, with a DVA induced ICH (out of 52 patients), 6 occurred in the cerebellum [21]. Interestingly, in the latter study the authors postulated that ICH caused by a DVA alone may result in smaller ICH and less severe symptoms compared to ICH associated with cavernomas. However, sample size was small.

In this case the radiological findings clearly identified the characteristic appearance of a DVA, later also demonstrating evidence of a co-existing cavernoma. The presence of a cerebellar DVA with haemorrhage from an associated cavernoma is a rarely reported entity and maybe of interest to the reader.

Patient consent

Written informed consent was obtained from the patient for publication of this case report, including accompanying images.

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