

## CASE REPORT

# Hemispheric venous dysgenesis in a woman presenting with seizures, weakness and parkinsonism

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## SUMMARY

This report describes an unusual case of a woman who developed progressive hemiparesis, seizures and hemiparkinsonism associated with MRI and angiographic evidence of chronic venous hypertension in the contralateral cerebrum and cerebellum. In the absence of inflammatory or veno-occlusive disorders, the patient's clinical and neuroradiological findings point to a developmental disorder, such as a hemispheric venous dysgenesis, as the underlying lesion.

## BACKGROUND

This middle-aged woman with an adult-onset progressive neurovascular syndrome involving almost the entire right hemisphere had a unique and bizarre combination of extensive right hemispheric and cerebellar hemosiderosis, with cavernomas and absence of superficial veins restricted to the same regions.

## CASE PRESENTATION

A 55-year-old right-handed woman presented with left-sided focal motor seizures with secondary generalisation for 3 months, on a background of progressive left-sided weakness and gradual loss of dexterity for 2 years. She had had a single generalised seizure 2 days after an uncomplicated pregnancy 19 years prior. There was no subsequent pregnancy after that. Details of the seizure were unknown, but her obstetric records did not suggest eclampsia or cerebral venous thrombosis. She was otherwise well without history of febrile seizures, headache, trauma, fever or other constitutional symptoms. She had not taken antipsychotic or antiemetic agents. The family history was non-contributory.

Her vital signs and general examination were normal. Other than dysarthric, hypophonic speech, mental status and cognitive function appeared normal for her age and educational status, without inattention or neglect. In addition to left facial weakness and mild spastic hemiparesis, cogwheeling and bradykinesia were noted in the left limbs, without postural instability or resting tremor. Sensory and cerebellar functions were normal.

## INVESTIGATIONS

Complete blood count and routine chemistries, including glucose, were normal, as were iron studies and screening tests for rheumatological disorders. Non-contrast head CT (figure 1) and MRI revealed right hemispheric cortical and subcortical calcifications. The T1-weighted images without gadolinium

showed diffuse cortical thickening, reduced white matter volume (figure 2A) and hyperintense nodular areas resembling cavernomas in right centrum semiovale (figure 2B) and cerebellum. Susceptibility-weighted images (figure 3) demonstrated extensive mineral deposition throughout the right hemisphere (figure 3A,B). MR tractography (figure 4) demonstrated severe loss of right hemisphere white matter fibre tracts. MR angiogram showed an attenuated appearance of the right middle cerebral artery without obvious stenosis or occlusion (figure 5). Cerebral digital subtraction angiogram (figure 6A) revealed absence of right cerebral and cerebellar cortical veins, with dilated deep medullary veins, and confirmed a diminutive, but patent middle cerebral artery without vascular beading.

## DIFFERENTIAL DIAGNOSIS

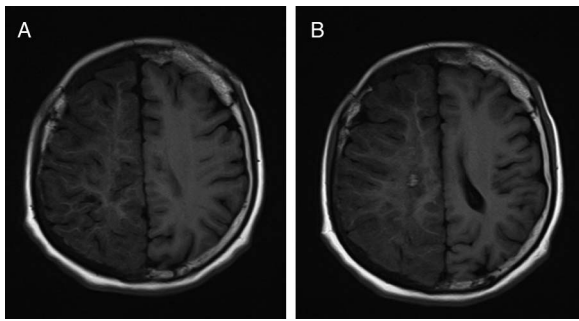
Chronic venous ischaemia has been hypothesised as causing the focal atrophy and extensive unilateral calcification occasionally seen adjacent to a developmental venous anomaly (DVA).<sup>1</sup> We believe a similar process due to impaired venous drainage from the right hemisphere resulted in the extensive mineralisation seen in our patient's head CT and MRI.

Cavernomas usually arise de novo, but familial forms have also been reported. Our patient did not have typical risk factors for cavernoma such as head injury, cranial surgery, radiation and DVA.<sup>2</sup> We hypothesise that chronic venous hypertension, and venous ischaemia in the right hemisphere triggered release of angiogenic factors, resulting in



**Figure 1** Non-contrast head CT showing multiple patchy calcification along the right posterior parietal subcortical region and in centrum semiovale.

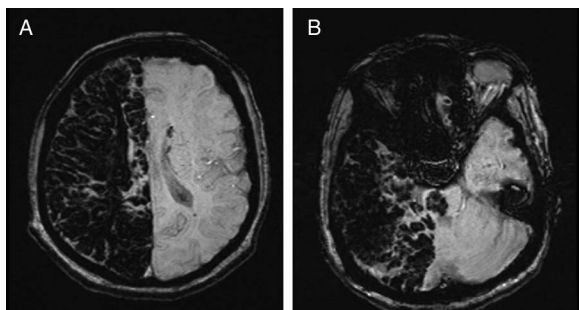
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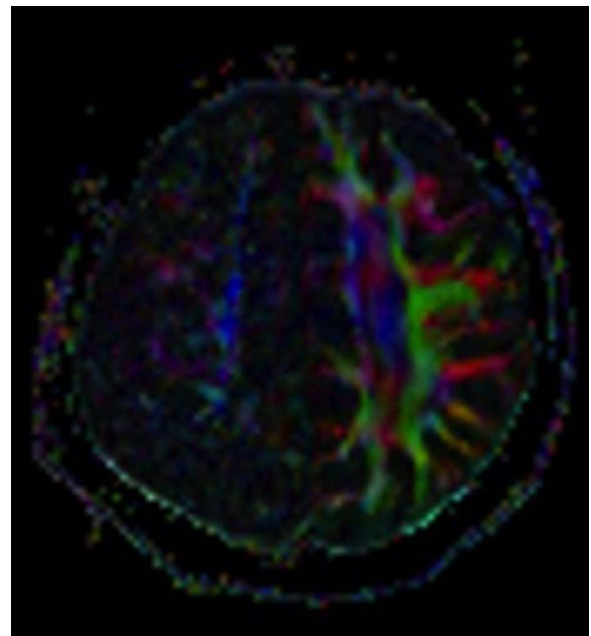
**Figure 2** T1-weighted brain MRI images without gadolinium showing diffuse signal alteration and thickening of cortex with moderate white matter atrophy in the right entire hemisphere (A) and subcortical hyperintense lesion with a popcorn appearance (B), suggesting cavernous angioma.

cavernoma formation ipsilateral to mineralisation. A teenager with focal seizure due to impaired venous drainage from a holo-hemispheric venous angioma with cerebral hemiatrophy has recently been reported,<sup>3</sup> although our patient had extensive supratentorial and infratentorial hemosiderin deposition ipsilateral to her absent cerebral and cortical cerebellar veins, suggesting congenital venous dysgenesis without any evidence of venous angioma.

Though the pathogenesis of the Sturge-Weber Syndrome (SWS) is typically seen by child neurologists, it occasionally presents during adulthood and may be relevant to our patient.<sup>4</sup> Seizure is the most common manifestation of SWS with brain involvement, but hemiparesis and cognitive deficits can be acquired over time as the result of stroke-like episodes associated with seizures or migraines. Aggression and self-abusive behaviour are more common in cognitively impaired adults with SWS. It is believed that impaired venous drainage accounts for the progression of cerebral disease in SWS, but the course of SWS during pregnancy is not well known. It is possible that our patient had undiagnosed SWS that became symptomatic after pregnancy and delivery of her first child resulting in focal seizure, slowly progressive mild hemiparesis and parkinsonian features. No port wine birthmark was noted on general examination. The typical leptomeningeal vascular malformation could not be confirmed as the patient did not undergo contrast-enhanced MRI scan. The angiographic characteristics of SWS were not seen in our patient. The absence of pial capillary blood pooling in our patient distinguishes her from individuals with typical SWS and the absence of cerebral atrophy indicates that the dilated deep medullary veins allow marginal,



**Figure 3** Susceptibility-weighted sequences showing extensive right hemispheric (A, B) and infratentorial (B) signal alteration suggesting diffuse unilateral hemosiderosis.



**Figure 4** MR tractography showing extensive loss of right hemispheric white matter tracts.

but not really adequate, venous drainage. The lack of atrophy in this patient's right hemisphere and cerebellum combined with absent veins from these areas suggests a disorder in which venous drainage, while deficient, is still occurring. The reduced white matter volume suggests a chronic process, perhaps developing after infancy, since there was no hemicranial atrophy or calvarial thickening.

#### OUTCOME AND FOLLOW-UP

Her seizures stopped with phenytoin 5 mg/kg/day and motor abnormalities improved with low-dose levodopa and baclofen, along with aggressive physiotherapy.

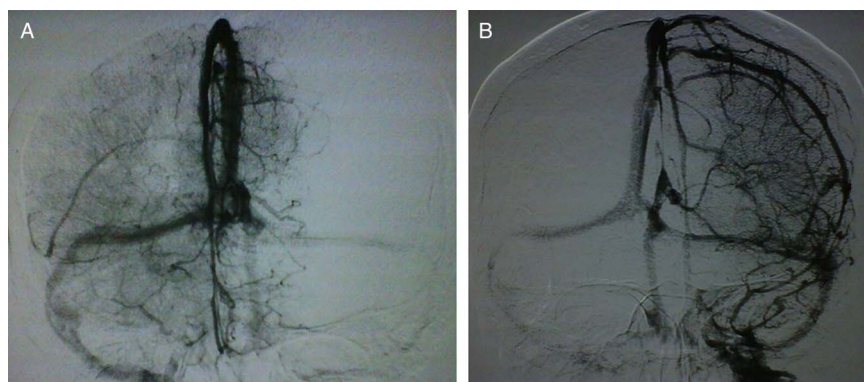
#### DISCUSSION

An earlier report of unilateral cerebral venous dysgenesis in a woman presenting with focal seizures<sup>5</sup> was confirmed by an angiogram showing complete absence of internal cerebral, basal and cortical veins. The absence of deep cerebral veins bilaterally



**Figure 5** MRA revealing attenuated right middle cerebral artery.

**Figure 6** (A) The venous phase of right internal carotid angiography showing prominent medullary veins and complete absence of superficial cortical and cerebellar veins in the right hemisphere. (B) The venous phase study of left ICA injection.



is the key diagnostic feature of cerebral venous dysgenesis, a rare disorder of childhood typically presenting with mental retardation, seizures and spasticity.<sup>6</sup> Our patient was asymptomatic until the second postpartum day after delivering her first and only child, which may suggest subclinical cerebral venous thrombosis or repeated and protracted Valsalva manoeuvres during delivery overwhelming an already marginal venous system. After that initial seizure, she remained healthy and neurologically intact for nearly two decades, when seizures recurred being preceded by progressive hemiparesis and hemiparkinsonism for 2 years. Unlike the typical bilateral involvement of deep venous structures in children, our patient's abnormalities appeared confined to the superficial veins of one side of her brain, possibly accounting for initially mild symptoms, protracted clinical course and definitive diagnosis in middle age.

### Learning points

- ▶ An asymptomatic developmental venous abnormality can be triggered by changes during pregnancy and childbirth in a woman.
- ▶ A rather benign clinical course despite the presence of extensive cranial venous abnormality should point to a developmental aetiology.
- ▶ If all other causes of venous drainage impairment have been systematically ruled out, a rare case of late occurrence of congenital venous dysgenesis may be considered.

**Contributors** DD, MT and VS participated in study concept and design; DD and BKR participated in acquisition of data; DD, BKR, MT and VS participated in analysis and interpretation of data; DD and VS participated in drafting of the manuscript; DD, MT and VS participated in critical revision of the manuscript for important intellectual content and VS participated in study supervision.

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

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