CASE REPORT

Hemiparesis post angiography in a patient with Sturge-Weber syndrome

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Introduction

We report the complications of cerebral angiography in a 19-year-old woman with a longstanding history of hemiplegic migraine and port wine stain in the distribution of the trigeminal nerve. Cerebral angiography performed to exclude the presence of an arteriovenous malformation demonstrated a developmental venous anomaly. The woman subsequently experienced prolonged hemiplegia and dysphasia, resolving over a period of 3 weeks. Possible explanations for the neurological deterioration are discussed.

Case report

A 19-year-old right-handed nursery nurse complained of intermittent mild generalized headaches since early childhood. At about the age of 16 years she began to experience more intense right-sided headaches lasting for several hours. At the onset of the headache she would feel light-headed, going on to develop right hemiparaesthesia and a moderate global aphasia. The hemisensory disturbance would resolve within 1 h or so; the language deficit persisted for the duration of the headache. On occasion, these symptoms would be accompanied by a visual aura of flashing coloured lights all over the visual field, lasting for 10–15 min. There was no history of seizures. She had a port wine birthmark in the distribution of the first and second divisions of the trigeminal nerve on the left and a similar lesion on the left lower limb. She had developed normally as an infant and child. Oral treatment with sumatriptan shortened headache duration, and headache frequency had decreased following the introduction of pizotifen.

On examination she had a port wine stain as described. General physical and neurological examinations were otherwise normal.

A diagnosis of migraine was made. In view of the accompanying focal neurological symptoms and the presence of the birthmarks, a CT scan of the brain was performed. This showed enhancing serpiginous vessels coursing around the body and splenium of the corpus callosum and left lateral ventricle, extending inferiorly towards the pineal cistern (Fig. 1). The choroid plexus on the left was slightly larger than on the right. Of note was the absence of ectopic calcification within either brain or leptomeninges. There was no abnormal cortical or leptomeningeal enhancement. It was proposed that this lesion represented an arteriovenous malformation, and to delineate the lesion further formal catheter cerebral angiography was performed. Common carotid injection on the left showed relative lack of superficial cortical veins. During the normal venous phase there were numerous radially orientated medullary veins draining into an enlarged deep venous system (Fig. 2). There was no early venous filling to suggest an arteriovenous malformation. Selective angiography of the right common carotid artery was normal.

A severe generalized headache developed 24 h after the angiogram and she became drowsy and vomited. She was febrile and went on to develop a right hemiparesis associated with expressive dysphasia and right sensory and visual inattention. There were signs of meningeal irritation. CT scanning performed within 3 h of the onset of her hemiplegia (Fig. 3) showed subtle left frontal...
convexity sulcal effacement with the previously noted serpiginous vessels. There was no haemorrhage. Her pyrexia persisted, and at 48 h after angiography a diagnostic lumbar puncture was performed. The CSF opening pressure was elevated at 40 cm H₂O but CSF constituents were normal and the fluid was sterile. The elevated CSF pressure raised suspicions that she might have sustained a cerebral venous sinus thrombosis. MRI was undertaken and demonstrated a normal signal void within the cerebral venous sinuses. There was evidence of left posterior frontal gyral hyperintensity (Fig. 4). An EEG showed slowing over the left cerebral hemisphere.

In view of persistent pyrexia and neurological signs there were concerns that she could have an encephalitis. She embarked on courses of acyclovir and steroids. Oral sumatriptan was given and had no effect. Repeat CT 7 days after the angiogram showed mild left convexity sulcal effacement. There was no evidence of infarction or haemorrhage.

She made a complete recovery over the course of 1 month. One year following the angiogram, she was still receiving pizotifen 0.5 mg at night. She was experiencing infrequent mild generalized headaches but had no focal neurological symptoms.

Discussion

According to the classification of Mulliken and Glowacki¹ facial port wine stains are termed capillary malformations. Unlike infantile haemangiomas (e.g. capillary and strawberry haemangiomas) they do not involute spontaneously with time. They are usually isolated but approximately 1-2% are associated with Sturge-Weber syndrome (SWS). SWS is one of the neurophakomatoses, exhibiting cutaneous and intracranial abnormalities. It is diagnosed clinically by the presence of a typical facial port wine stain in the cutaneous distribution of the trigeminal nerve with accompanying atrophy and calcification of the ipsilateral cerebral hemisphere, predominantly the occipital lobe.² The underlying mechanism is a relative lack of superficial cortical venous drainage which occurs early in fetal development, either as a result of under-development of the venous structures or venous thrombosis. As a consequence blood is redirected into the leptomeninges, producing abnormal vascular channels. The resulting leptomeningeal angiomatosis is the hallmark of SWS.³ Blood is also
redirected centripetally into the deep venous system, causing persistence of more primitive drainage structures and increasing the risk of developmental venous anomalies.\(^3\)

Not infrequently the radiological features of ipsilateral cerebral atrophy and parenchymal calcification may be absent in SWS,\(^4,5\) as in our patient. In these cases MRI with gadolinium to demonstrate enhancement of the pial angioma should be considered.\(^4\) Venous hypertension may cause engorgement within the choroid plexus with subsequent enlargement of the plexus itself. The degree of enlargement may correlate with the extent of leptomeningeal involvement.\(^5\)

There is a recognized association between SWS and other vascular abnormalities such as developmental venous anomalies, dural arteriovenous fistulas and, infrequently, arteriovenous malformations (AVMs).\(^6\) Occasionally, as in our case, enlarged vessels can be seen in a periventricular distribution on CT and MRI. This appearance is likely to represent enlargement of the vessels of the deep venous system of the brain.\(^7\) It may not be possible to distinguish between an AVM and hypertrophied venous drainage on CT or MRI. Occasionally the demonstration of enlarged feeding arteries may allow the distinction to be made.

Although our patient did not show the classical constellation of clinical and neuroradiological signs, the presence of a capillary facial angioma (port wine naevus), enlargement of the choroids plexus, lack of superficial cortical veins and anomalous deep venous system are suggestive of the diagnosis.\(^5\)

It is thought that venous stasis and recurrent thrombotic episodes are the main factors responsible for the neurological deterioration and brain atrophy seen in SWS.\(^8\) Hosokawa et al.\(^9\) serially tracked the cerebral blood flow of an infant with SWS from 6 months to 10 months old, using cerebral perfusion single photon emission CT (SPECT) with \(^99m\)Tc-ethyl cysteinate dimer (\(^99m\)Tc-ECD). Initially SPECT demonstrated an increase in regional cerebral blood flow (rCBF) over the affected area. Simultaneous CT showed faint lobar hyperdensity coupled with leptomeningeal enhancement on MRI but no volume loss. Subsequent imaging showed cortical and subcortical calcification on CT with volume loss and accompanying reduction in rCBF on \(^99m\)Tc-ECD. This supports the view that congestion results in a chronic hypoxic state with eventual loss of neuronal function. There is consequently a reduced demand for oxygen leading to a decrease in perfusion.

The acute neurological deterioration of our patient was preceded by a rise in intracranial

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**Figure 3** Unenhanced CT brain. There is subtle effacement of the left frontoparietal convexity sulci. No intracranial haemorrhage.

**Figure 4** T2 Weighted axial brain. Subtle increased cortical signal of left cerebral hemisphere, consistent with infarction.
pressure of up to 400 mm H₂O. A report by Kiley et al. documented a case of Klippel-Trenaunay-Weber syndrome (KTWS), which has intracranial features similar to SWS. Their patient presented with an acute history of spontaneous headache, vomiting and profound visual disturbance. The opening CSF pressure was 650 mm H₂O. An angiographic pattern of paucity of cortical veins with abnormal central venous drainage was seen similar to that in our patient. They postulated a mechanism of intracranial hypertension secondary to compromised cerebral venous outflow. It is plausible that in our case a venous thrombotic episode triggered by cerebral angiography further compromised already compromised venous drainage. This manifested as left posterior frontal gyral hyperintensity likely to be oedema secondary to ischaemia or a small infarct in this region. Diffusion-weighted MRI might have assisted in the diagnosis of cortical ischaemia in this region but was not available. Masson et al. reported two cases of cerebral venous angioma presenting as venous infarction, one in the left parietal lobe and the other in the left frontal lobe. Cerebral imaging demonstrated thrombotic occlusion of the draining vein. Both patients were free of coagulopathy and had to receive anticoagulants.

Alternative explanations would include an acute migrainous episode triggered by cerebral angiography or an embolic event. The association between migrainous events and cerebral angiography has been previously reported. A complication rate of 1% has been quoted for cerebral angiography in the general population. There has not been any reported study on the complication rate of cerebral angiography in patients with SWS, and there has been mixed opinion on this complication rate in patients with migraine. For example, Patterson et al. quote a higher complication rate in this subgroup, whereas in a more recent study Shuaib et al. conclude that with the use of more modern angiographic technique and intravenous contrast medium there is no significant difference between the two groups. The presence of focal areas of high signal intensity in the white matter on T2-weighted MR in persons with uncomplicated migraine is well recognized. In addition, cortical abnormalities, similar to infarcts, in persons with complicated or hemiplegic migraine have been described. These cortical abnormalities, unlike the white matter lesions, tend to be associated with neurological deficits. Reversible cerebral oedema, presenting as high signal cortical lesions on T2-weighted MR, has been described and was presumably due to ischaemia as opposed to infarction. The exact cause of our patient’s pyrexia following the procedure is unclear. On the basis of the neurological signs this could be attributable to herpes encephalitis, but pyrexia and even CSF leucocytosis has been described in hemiplegic migraine and migraine coma.

In conclusion, we postulate that the acute deterioration in our patient was due to the cerebral angiogram. This may have caused a subtle cortical venous thrombosis or precipitated a migrainous episode, both of which could have the CT and MRI features shown above. The lesson from our case is that non-invasive techniques should be the first imaging step in the assessment of persons with SWS, venous drainage abnormalities or a history of hemiplegic migraine, and that formal cerebral angiography should be undertaken with caution.

References


