SHORT REPORT

New onset migraine with a brain stem cavernous angioma

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There is increasing evidence that migraine is primarily a brain disorder, probably involving subcortical sensory modulating areas with a secondary neurovascular response. It is also well recognised from experimental work that activity in nociceptive pathways is modulated by structures in the brain stem. Cavernous angiomas are reported to occur in 0.1–0.5% of the general population, with about 10–30% of these involving the brain stem or cerebellum.

We describe a case of new onset migraine in a patient who had a pontine bleed from a cavernous angioma.

CASE REPORT

A 22 year old woman presented in June 2000 with symptoms of ataxia, dysarthria, dysphagia, and left facial numbness that had developed over two weeks. She was admitted to hospital and went on to develop a sudden onset severe, generalised headache. On examination she was found to have horizontal nystagmus on left gaze, hyperreflexia of right upper and lower limbs, an extensor plantar response on the right, and right ankle clonus. Her gait was markedly ataxic.

The headache persisted for two weeks. Magnetic resonance imaging of the brain showed a bleed from a left pontine cavernous haemangioma as well as a Dandy–Walker variant malformation (fig 1).

Following this episode her neurological symptoms improved but she was left with episodes of severe head pain three to four times a month. She describes attacks that start as a left sided retro-orbital and occipital sharp pain, eventually becoming bilateral, and associated with photophobia and worsening of pain on movement. The headache is also associated with transient ataxia and paraesthesiae in the right hand, as well as fuzziness of vision. These symptoms last less than an hour and occur during the headache. The headaches last 24 to 48 hours. Stress was the only trigger she had noted. Before June 2000 there was no history of headaches.

Figure 1  Panels A & B: Magnetic resonance imaging showing left pontine cavernous angioma and Dandy–Walker malformation.
In her past medical history she underwent cranial irradiation and had intrathecal methotrexate for acute lymphoblastic leukaemia in 1987; full remission was achieved. She is a non-smoker and does not drink alcohol. There is a family history of migraine affecting her father, and her uncle had a Dandy–Walker malformation.

Her drug treatment was paracetamol (acetaminophen), tramadol, or morphine for her severe headaches. She was on the oral contraceptive pill.

On examination there were no abnormal neurological findings.

**DISCUSSION**

This patient provides further evidence for a role of the brain stem in the pathophysiolo gy of migraine. The patient describes symptoms that fulfill the International Headache Society (IHS) criteria for migraine with aura following from an episode during which she sustained a haemorrhage from a previously undiagnosed left pontine cavernous angioma. Before that event she had no history of headaches.

Functional evidence of a role for the brain stem in migraine has been provided by positron emission tomography (PET) studies showing increases in regional blood flow that persist after the headache is relieved. Areas activated are slightly contralateral to the side of pain and include the dorsal midbrain and dorsolateral pontine tegmentum. These areas have not been found to be active in other primary headache disorders, such as cluster headache and short lasting neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), nor in experimentally induced facial pain. Structural evidence of brain stem involvement in primary headache is provided by observations of excess iron deposition in the periaqueductal grey matter (PAG) in both episodic and chronic migraine, although no grey or white matter changes could be seen using the sensitive automated method of voxel based morphometry.

It is of interest that her headaches always start on the left side and then may become bilateral. These findings are consistent with those observed in a seminal observation of headache contralateral to the site of the pain is ipsilateral to the lesion. This case provides further evidence for the involvement of the brain stem in the initiation of migraine and adds to the debate over the lateralisation of the lesion.

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**REFERENCES**

14. **Knight YE, Goodson PJ. The periaqueductal grey matter modulates trigeminovascular input: a role in migraine? Neuroscience 2001;106:793–800.**
A 26 year old man with cirrhosis and subacute cognitive decline was admitted with a decrease in his activities of daily living. Hyperpigmentation of his lower extremities, asterixis, and Kayser-Fleischer rings were observed. Neurologic examination revealed psychomotor retardation, upper extremity weakness, hypertonia, a left upper extremity tremor, and extensor plantar responses. Serum ceruloplasmin and urine copper studies confirmed the diagnosis of Wilson’s disease. He subsequently developed stereotypical episodes of fever, diaphoresis, tonic decorticate posturing, and autonomic instability followed by somnolence. Infectious investigation and serial EEG were unrevealing. Progressive neurological decline ensued, despite D-penicillamine, trientine, and zinc sulphate, leading to akimetic mutism and rigidity with profound autonomic dysfunction.

Serial MRI revealed symmetric bilateral areas of T2-hyperintensity involving subcortical white matter, basal ganglia, external capsules, thalami, midbrain, and pons. Progressive involvement of the midbrain demonstrated the characteristic MRI evolution of the “face of the giant panda” (Figure 1, A, C, and E). Contemporaneous evolution of dorsal pontine signal abnormalities (Figure 1, B, D, and F) resembled the face of a cub, with eyes formed from the central tegmental tracts. Encircling signal abnormality was possibly because of involvement of the superior cerebellar peduncles, pedunculopontine tegmental nuclei, rubrospinal tracts, or lateral lemnisci. Selective vulnerability and progressive involvement of midbrain and dorsal pontine structures may be chronicled on MRI by the faces of the giant panda and her cub, respectively.

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References

Figure 1 Serial T2-weighted MRI of Wilson’s disease over a six month period illustrating evolution of the characteristic “face of the giant panda” in the midbrain (A, C, and E) and face of her cub in the dorsal pons (B, D, and F).