Simultaneous Moyamoya disease and cervical spinal cord low-grade astrocytoma in a child with neurofibromatosis type 1

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DESCRIPTION

A 17-year-old girl with a clinical diagnosis of neurofibromatosis type 1 (NF-1) presented for a routine follow-up with worsening headaches, left-sided hemiparesis and hyper-reflexia. Her medical history was significant for prematurity, speech delay and mild left hemiparesis since the age of 10 years initially thought to be related to her prematurity. Physical examination revealed multiple café au lait macules and axillary freckling in addition to her hemiparesis. MRI brain showed a small right internal carotid and middle cerebral artery. Magnetic resonance angiogram demonstrated retrograde flow from the external carotid artery filling the middle and anterior cerebral arteries, consistent with Moyamoya disease. However, on sagittal and coronal MRI brain sequences an expanded cervical

cord was appreciated and MRI spine revealed a non-contrast enhancing cervical cord tumour (figure 1). The patient underwent simultaneous encephalodurangiomylar repair of the Moyamoya disease and subtotal resection of the cervical tumour without complication (figure 2). Pathology was consistent with a diagnosis of juvenile pilocytic astrocytoma. Moyamoya disease and low-grade spinal cord astrocytomas are known associations of NF-1. Moyamoya disease can be a presenting sign of weakness in patients with NF-1 and often show improvement following revascularisation procedures.¹ Similarly, spinal cord low-grade gliomas associated with NF-1 can be asymptomatic.² This case is unique in the simultaneous presentation and repair of comorbid conditions of NF-1 associated with weakness.

Figure 1 (A) Coronal fluid attenuated inversion recovery MRI brain demonstrates prominent R sylvian fissure consistent with atrophy, a stable optic glioma and cervical hyperintensity (arrow). (B) Magnetic resonance angiogram reveals narrowing of the right internal carotid and middle cerebral arteries consistent with Moyamoya disease. (C–E) MRI cervical spine demonstrates a non-contrast enhancing intramedullary juvenile pilocytic astrocytoma (arrow).

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Figure 2 (A) Magnetic resonance angiogram of 6 months post encephalodurangiomyangioma repair reveals evidence of early revascularisation (arrow). (B) Sagittal T2 MRI spine postoperatively reveals subtotal resection of intramedullary juvenile pilocytic astrocytoma.

Learning points

- ▶ Moyamoya disease can be a complication of neurofibromatosis type 1 (NF-1); identification is important because patients are likely to improve with revascularisation therapy.
- ▶ Spinal cord tumours are common in NF-1; patients with NF-1 and symptoms of weakness should undergo spinal imaging.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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