CASE REPORT

Unilateral movement disorder as a presenting sign of paediatric post-varicella angiopathy

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SUMMARY

Diagnosing ischaemic stroke in children is often difficult. Post-varicella angiopathy (PVA) is a well-recognised and frequent cause of childhood ischaemic stroke, particularly affecting the basal ganglia. When a previously healthy child presents with unilateral abnormal involuntary movements, cerebral infarction should be included in the differential diagnosis and PVA should be considered, even when there is no recent history of rash and cerebrospinal fluid is normal. Medical history and intracranial vascular imaging are important for early diagnosis and treatment.

BACKGROUND

The diagnosis of arterial ischaemic stroke (AIS) in children is often initially missed, owing to the many stroke mimics and a large variety of presenting symptoms, among which (haemi)paresis, dysphasia, loss of consciousness, headache and seizures are the most frequent.¹ Symptoms are often non-abrupt in onset and vary according to age and arterial territory. Movement disorders in children with stroke have been reported, and occur most often in the chronic stage.^{2–4} We report three children with a transient movement disorder as the presenting symptom of lenticulostriate infarction owing to post-varicella angiopathy (PVA).

CASE PRESENTATION

Patient 1

The parents of a 3-year-old boy had noticed an unstable gait, causing him to fall frequently, as well as clumsiness of his left arm. Initially, they attributed these signs to fatigue and sleep deprivation during their summer holidays, but when they noted involuntary arm and leg movements after a week, they sought medical help. He had chickenpox 3 months earlier. The history was unremarkable. On admission he showed dystonic movements of his left arm, more clearly during walking, and a subtle paresis of his left leg with a Babinski's sign. MRI showed infarction in the right caudate nucleus and adjacent frontal white matter. MR angiography revealed mild irregular stenosis of the right distal internal carotid artery (dICA), proximal anterior (ACA) and middle cerebral artery (MCA) (figure 1A). Cerebrospinal fluid (CSF) examination revealed 35 leucocytes/ μ L, normal protein (0.20 g/L) and a positive PCR for varicella zoster virus (VZV). Further work-up for stroke aetiology was normal. He was treated with aspirin, and acyclovir for 2 weeks. Abnormal involuntary movements disappeared within 1 month. Repeated magnetic

resonance angiography after 3 months was unchanged and there were no recurring symptoms during the first year of follow-up.

Patient 2

A 2-year-old boy was admitted for a second opinion after he was diagnosed with a lenticulostriate infarction. Parents had noticed fluctuating subtle motor problems of his right leg 4 weeks earlier, and abnormal involuntary movements of his right arm, leg and facial muscles 2 days before admission. He had chickenpox 2 months earlier. On neurological examination there was right-sided haemichorea without other abnormalities. MRI showed a left caudate nucleus and putamen infarct with severe stenosis of the left dICA, proximal ACA and M1–M3 branches of the MCA (figure 1B). CSF examination showed a normal leucocyte count and protein. PCR for VZV was negative. Anti-VZV IgG antibodies were not tested. Further work-up for stroke aetiology was normal. He was treated with aspirin. At 1 month follow-up there was complete clinical recovery.

Patient 3

The mother of a 3-year-old girl noticed involuntary movements of her daughter's left arm and leg shortly after awakening. She had chickenpox 7 months earlier. On neurological examination choreatic movements of the left arm and leg were seen, without pyramidal signs. MRI showed infarction in the right caudate nucleus and putamen, and a stenosed proximal MCA and ACA (figure 1C). Work-up for stroke aetiology was normal. Lumbar puncture was not performed. She was treated with aspirin. The haemichorea disappeared over the course of 3 weeks. On follow-up after 1 year, she had not experienced recurrent episodes and MR angiography showed normalisation of arteriopathy.

DISCUSSION

We report three children with a unilateral movement disorder as presenting symptom of lenticulostriate infarction and a non-progressive unilateral intracranial arteriopathy. They had chickenpox within 12 months preceding stroke onset, and were therefore diagnosed with PVA.⁵

An arteriopathy was found in 53% of children who underwent vascular imaging in a large international study on childhood AIS.⁶ In the majority of children with AIS of arteriopathic origin, the arterial abnormalities are intracranial, unilateral and non-progressive on follow-up vascular imaging. This condition is classified as 'transient cerebral arteriopathy' (TCA).^{5 7 8} The vast majority of these

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Figure 1 MRI (T2 sequence) and 3D time-of-flight MR angiographies of all three patients at baseline (A–C) showing basal ganglia infarcts and a large artery arteriopathy in all three patients.

children have basal ganglia infarcts.^{7–8} Many arguments favour post-infectious inflammation of the arterial wall as the cause of TCA.⁷ In 31–44% of these children a primary VZV infection preceded stroke by less than 12 months.^{7–9} The diagnosis of PVA is based on the combination of these clinical and radiological findings and is a well-recognised entity in childhood ischaemic stroke.^{3–5–9–10} Recent rash or CSF pleocytosis is not required for the diagnosis of PVA, neither is the detection of VZV DNA or anti-VZV IgG antibodies in CSF.^{5–10}

The incidence of post-stroke movement disorders in children is unknown, but is assumed to be higher than in adults.² Most publications are single case reports or small case series.^{2–11} Haemidystonia as a consequence of stroke occurs more often in children than in adults, but particularly in the chronic stage.⁴ Two studies have showed that haemichorea is the most common stroke-associated movement disorder in the acute phase of adult stroke.^{12–13} Reports on chorea in children with acute stroke, however, are limited to a few cases, in whom other focal signs accompanied the chorea.³

Movement disorders in the acute stage of adult stroke are generally associated with spontaneous recovery within weeks,¹³ similar to the time course of symptoms in our three children. Haemidystonia is attributed to lesions that interrupt the corticostriatopallidothalamocortical loop, most often in the striatopallidal complex.⁴ Similar lesions have been described in patients with haemichorea.¹⁴ Accepted models of basal ganglia circuitry often do not correspond to MRI lesion localisation and the type of movement disorder,^{12–13} possibly because MRI does not visualise the full extent of pathology and the distant functional effects of the lesion.¹²

Competing interests None.

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Learning points

- ▶ Post-varicella angiopathy (PVA) is a well-recognised and frequent cause of childhood ischaemic stroke.
- ▶ When a child presents with haemichorea or haemidystonia, basal ganglia infarction should be included in the differential diagnosis. Transient cerebral arteriopathy and its subset PVA should then be considered, even if there is no recent history of rash and cerebrospinal fluid is normal.
- ▶ Brain MRI and vascular imaging of the intracranial arteries in children with an acquired unilateral movement disorder is important for early diagnosis and treatment of PVA.

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