Ischaemic Stroke In Children Secondary to Post Varicella Angiopathy

Introduction

Varicella in childhood is a self-limiting disease, which usually follows a benign course. However, complications, although rare, may have serious consequences. Ischaemic stroke secondary to post varicella angiopathy is a well-described complication and is estimated to account for up to a third of all strokes in infants. We present three previously healthy children who presented to our centre with ischaemic cerebrovascular infarction due to varicella angiopathy. All three children first presented within six weeks after onset of varicella infection and had MRI changes characteristic of ischaemic stroke secondary to post varicella angiopathy. While one child made an excellent recovery being left with only a minor deficit, the remaining two children were left with considerable morbidity severely affecting quality of life. The varicella vaccine has been proven to be well tolerated, safe and effective. We conclude that varicella vaccination should be considered for inclusion in the vaccination schedule to prevent serious complications which while rare may have devastating consequences.

Case 1

A five-year-old girl was referred for investigation of her gait abnormality. She had a ten-month history of left sided facial twitching, clumsiness of the left foot and frequent falls. She had clinical varicella infection one month prior to onset of symptoms, which had required admission to hospital. Examination revealed dystonia of the left upper and lower limb. MRI of brain showed increased signal intensity in the right insular cortex, also involving the caudate and lentiform nucleus on the same side, consistent with infarction (figure 3a). Repeat MRI four months later showed no evidence of progression. Extensive work up was negative. Her dystonia showed limited response to medical treatment. She subsequently progressed to develop complex partial seizures. Given the typical MRI findings of basal ganglia infarction with the history of varicella infection and all other common causes for childhood infarction being excluded, postvaricella angiopathy was felt to be the most likely diagnosis. Now five years following the acute episode she has severe medically refractory epilepsy, which has led to considerable morbidity and limitation in her everyday life.

Case 2

A six-year-old boy presented with a two-day history of left facial drooping, left sided clumsiness with dystonic posturing of left foot and dysarthric speech. He had a ten-month history of clinical varicella infection two weeks prior to presentation. Crusting varicella lesions were evident at the time of presentation. MRI of brain showed a well-defined area of increased T2 signal in the right internal capsular region that extended upwards to involve the body of the caudate nucleus in keeping with infarction (figure 3a). All other investigations were within normal limits apart from SMA (smooth muscle antibody), which was slightly elevated at a value of 40. This was felt to be secondary to recent viral infection. He was treated with a three-day course of intravenous methylprednisolone followed by a six-week course of oral steroids. He also received a ten-day course of intravenous acyclovir and was commenced on aspirin 75mgs daily. With intensive physiotherapy he made slow but steady improvement over the next few months. Nine months post initial presentation he had greatly improved function and had very mild blunting of left nasolabial fold. He was back playing sport. At follow up two years later he still has minor residual tightness of left Achilles tendon, that does not interfere with function. He has had no further recurrences.

Case 3

A seven-year-old girl was referred for investigation of her gait abnormality with total left sided paresis and slurred speech. This was one-month following onset of clinical varicella infection. An MR angiogram showed decreased flow in right middle cerebral artery with occlusion of the anterior cerebral artery (figure 3b). MRI of brain showed infarction in right middle cerebral artery territory (figure 3a). She was treated with three days of intravenous methylprednisolone followed by oral steroids. A four-week treatment course of intravenous acyclovir was commenced. Repeat MRI seven days later showed no extension of the lesion.

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Abstract:

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Discussion

Varicella in childhood, while usually a self-limiting disease, with a benign course, may have serious consequences. Cerebrovascular infarction following varicella angiopathy is a well-described complication. Given that it is a probable cause in one third of acute strokes in childhood one should always ask whether there is a history of varicella infection. Ischaemic cerebrovascular disease typically occurs approximately six weeks after primary varicella zoster virus infection. Magnetic resonance imaging is the most sensitive neuroimaging tool in these patients. The proximal portion of the main cerebral arteries are frequently affected, typically the middle and anterior cerebral arteries. Infarction of the basal ganglia is also frequently associated. The prognosis is good, with recurrence being uncommon once other causes have been excluded. Physiopathology and treatment remain under discussion. It is generally initially treated with a short course of intravenous methylprednisolone followed by oral steroids. Acyclovir treatment is justified as PCR analysis and antibody testing of cerebrospinal fluid confirm the presence of varicella virus and are strong presumptive evidence of viral infection rather than solely immunemediated mechanisms as previously thought. Detection of antibodies to varicella virus in the cerebrospinal fluid further supports the diagnosis of infection of the nervous system. Debate still exists as to optimal duration of treatment. The intravenous route is preferred as oral acyclovir has poor penetration into CSF spaces. It is recommended to test cerebrospinal fluid for both varicella DNA using PCR and varicella antibody analysis.

We conclude from our experience of these three cases that there is a need for varicella vaccination to prevent serious complications, which, while rare, may have devastating consequences.

References