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CASE REPORT

Postvaricella Basal Ganglia Infarction — Early and Late Computed Tomography and Magnetic Resonance Imaging Findings

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ABSTRACT

The case of a 9-year-old boy who developed acute neurological complications soon after varicella infection is reported. Computed tomography and magnetic resonance imaging showed bilateral basal ganglia lesions, which almost completely resolved on follow-up scans. There has been little previous discussion in the literature of the morphological changes detected on imaging.

***Key Words**: Basal ganglia infarction, Computed tomography, Magnetic resonance imaging, Varicella infection*

CLINICAL DETAILS

A 9-year-old boy initially presented with pyrexia of unknown origin and bilateral cervical masses. Investigations including viral titre and immune status were normal. Subsequent excisional biopsy of the masses showed histiocytic necrotising lymphadenitis, and the diagnosis of Kikuchi disease was made. He was given a short course of steroids and was discharged soon after. A brief contact with a child suffering from chickenpox occurred but no prophylaxis was given. Five days after corticosteroid therapy was stopped, the child presented with eruption of typical varicella maculopapular skin lesions. He was treated with a course of intravenous acyclovir. However, during the middle of the vesiculation period, his condition deteriorated and he became lethargic with a fluctuating level of consciousness. He also complained of illusions and was noticed to have abnormal behaviour, continuous flickering of the eyelids, and intermittent uprolling of the eyeballs. Cranial nerves as well as peripheral motor and sensory functions were normal. The serum and cerebrospinal fluid (CSF) analysis including ammonia level and coagulation profile were within normal limits. The first CSF viral titre, as well as blood culture and viral titre, were negative. The electroencephalogram was unremarkable.

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On admission to hospital, a computed tomography (CT) scan demonstrated hypodensity in bilateral external capsules and basal ganglia (Figure 1). Magnetic resonance imaging (MRI) of the brain was obtained a few days later. There was no abnormal signal in the external capsules, but bilateral and symmetric abnormal signal intensities were noted in the caudate nuclei, globus pallidi, anterior portions of putamina and internal capsules,

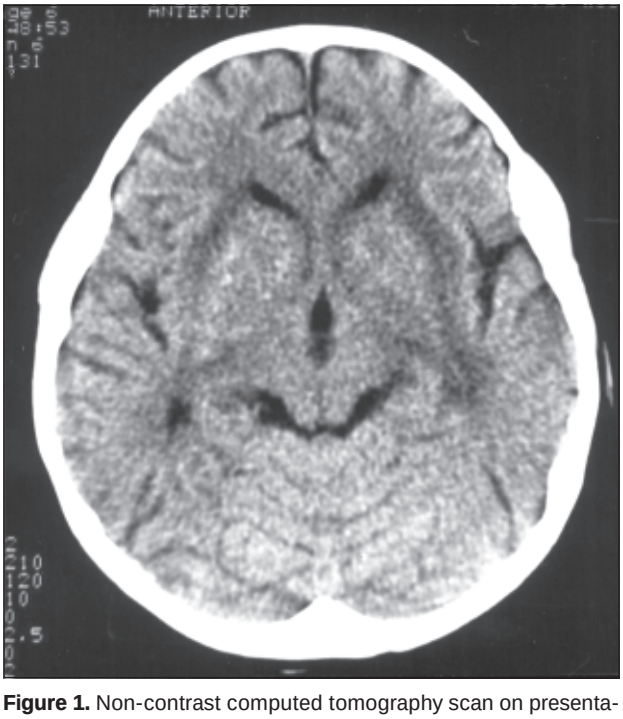

Figure 1. Non-contrast computed tomography scan on presentation showed hypodense areas in external capsules and both basal ganglia.

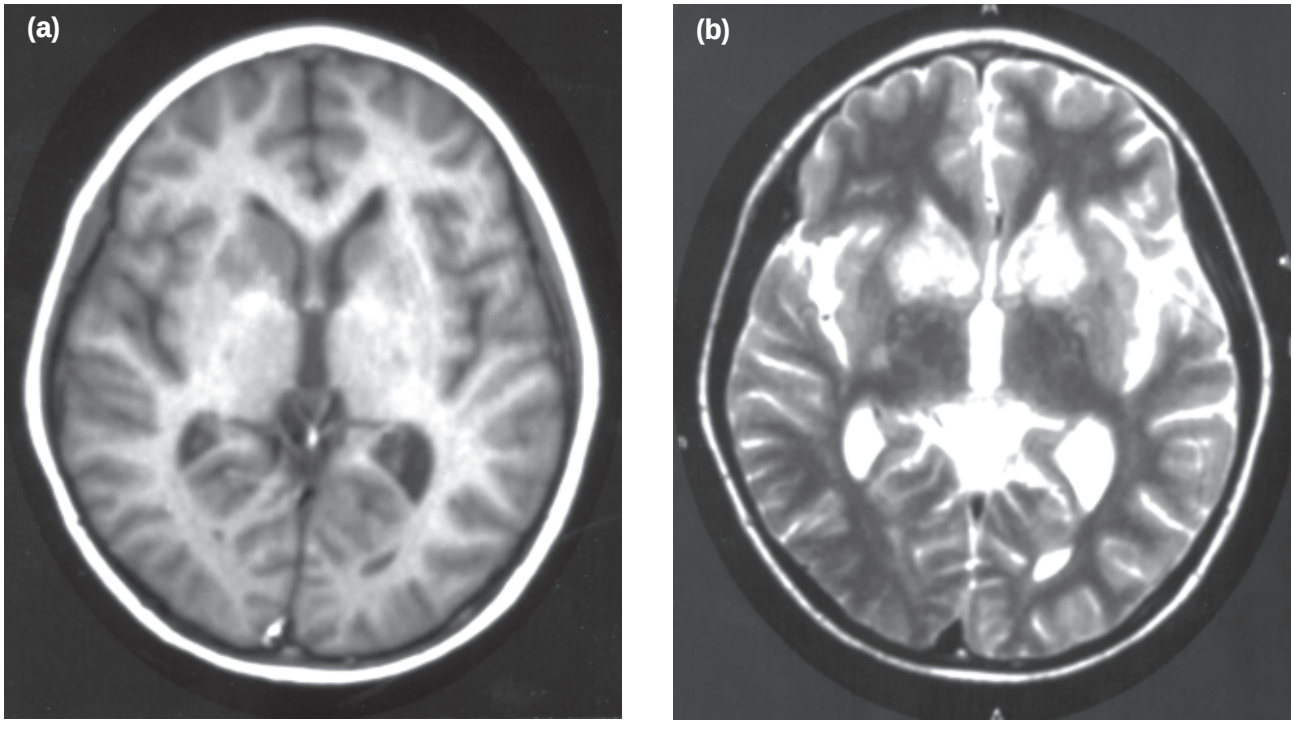

Figure 2. Axial T1-weighted (a) and T2-weighted (b) magnetic resonance imaging of the brain showed symmetrical abnormal signals in basal ganglia and internal capsules.

which appeared hypointense on T1-weighted images and hyperintense on T2-weighted images (Figure 2).

A second lumbar puncture was performed and a raised CSF titre for varicella was documented. The neurological symptoms persisted for 2 weeks and gradually returned to normal during the following month. Follow-up MRI studies of the brain were performed 4 months after initial presentation. The original signal aberrations in the basal ganglia and internal capsules had nearly completely disappeared and were replaced by subtle increased signal intensity on T1-weighted images, thought to be due to calcification (Figure 3). The basal ganglia had also diminished in size. No recurrence of abnormal signals in the basal ganglia was noted. A CT scan at 2 years confirmed the presence of minimal calcifications in both basal ganglia and mild dilatation of the frontal horns, suggestive of adjacent brain substance loss (Figure 4).

DISCUSSION

Varicella infection is a common childhood infectious disease. Neurological complications occur in less than 1% of patients, mostly affecting children younger than 15 years.[1-4] Complete recovery of neurological functions without sequelae occurs in most patients.[5] Immunocompromised patients are known to be more prone to develop neurological and other complications. Kikuchi's disease (histiocytic necrotising lymphadenitis) is a self-limiting disease of unknown cause. It often presents with intermittent fever and persistently enlarged cervical lymph nodes. The disease is not known to have neurological sequelae and its association with postvaricella complications is not documented in the literature. In this patient, the brief history of previous steroid therapy might have altered his immune status and accentuated the subsequent development of postvaricella cerebral vasculopathy.

Among the neurological sequelae of varicella infection, encephalitis is the most common complication.[1] Other complications include meningitis, Reye's syndrome, transverse myelitis, polyradiculoneuropathy and optic neuritis. Encephalitis can solely affect the cerebellum but diffuse cerebral involvement, although being much less common, can also occur. To date, the pathogenesis of encephalitis remains unclear. Direct viral invasion and the immunologically-mediated process of neurological injury are the two commonest hypotheses mentioned in the literature.[2,3,6,7] During the past 2 decades, basal ganglia lesions with imaging and clinical features compatible with infarcts have increasingly been recognised as a distinct complication of varicella infection.[8-13] Most investigators have proposed that the infarcts resulted from vasculopathy causing damage to the vessel wall media by direct viral invasion, immune-complex

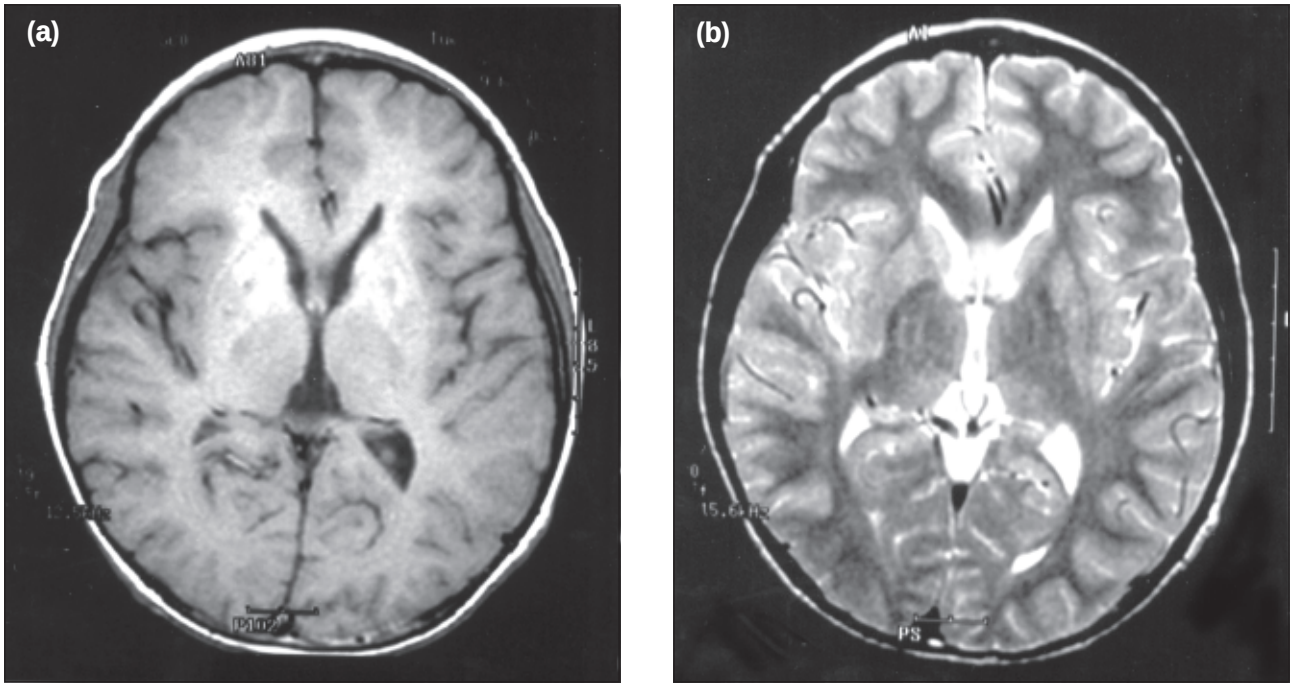

Figure 3. Axial T1-weighted (a) and T2-weighted (b) magnetic resonance imaging 4 months after presentation. The abnormal signals in the bilateral basal ganglia had nearly completely disappeared. Note the subtle T1-hyperintense areas in the caudate nuclei which were thought to be foci of early calcification.

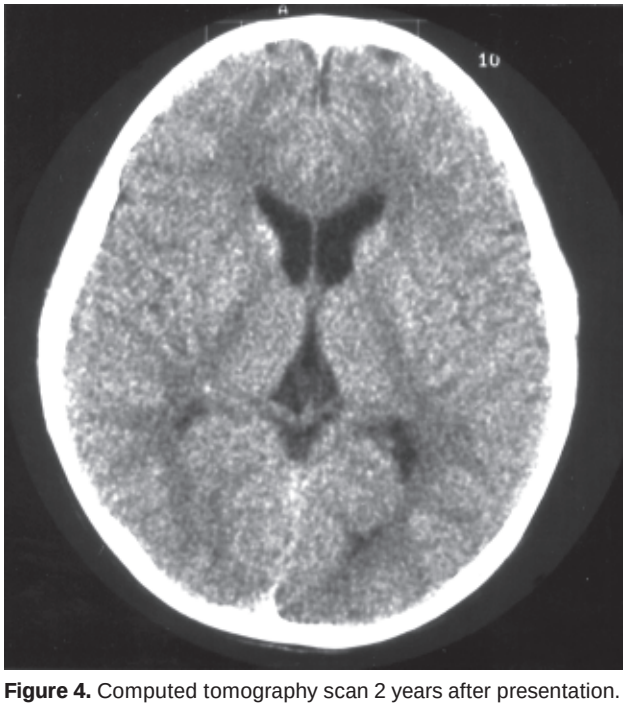

Figure 4. Computed tomography scan 2 years after presentation. Both basal ganglia were hyperdense due to calcification, particularly prominent in both caudate nuclei. Note mild dilatation of frontal horns of the lateral ventricles.

reactions, or a combination of the two. The cerebral angiogram is abnormal in some of the affected patients, providing evidence of vasculitis-based infarction. The common angiographic findings are luminal narrowing and intimal irregularities of the distal internal carotid arteries, the A1 segment of the anterior cerebral arteries, and the M1 segment of the middle cerebral arteries.[1,10,11,13]

The CT and MRI findings in this patient is compatible with postvaricella basal ganglia infarcts. Periarterial distribution of the lesions involving deep gray matter without involving white matter differentiates them from varicella encephalitis, which is typically perivenous and mainly affects the cortex and superficial white matter.

Complete recovery of neurological functions in this patient is in keeping with the excellent clinical outcome documented in most cases.[1,13] However, to our knowledge, there are no reports of longitudinal follow-up of postvaricella basal ganglia infarction by imaging in the literature. In this patient, although mild atrophy of basal ganglia was noted in follow-up studies, most of the initial abnormal attenuation of signal intensities, notably those in the external capsules and basal ganglia, showed remarkable temporal resolution on CT scan and MRI. This suggests that a substantial portion of the changes in the acute phase of the disease is reversible. In a review of five patients suffering from postvaricella infarction, Bodensteiner et al observed that all patients with postvaricella infarction had full recovery of neurological function despite extensive MRI abnormalities.[11] The authors proposed that some of the

imaging findings were likely to be caused by oedema. Since biopsy of the brain is rarely performed for varicella-related complications, this suggestion has not yet been confirmed. Here, indirect supportive evidence of the presence of significant oedema during the initial period of postvaricella infarction is provided by the morphological evolution in this patient as depicted by CT and MRI.

Most cases of postvaricella infarction occur 1 to 4 months after the exanthem, but a latent interval as short as days, as in this case, is possible.[1,11,13,14] Rarely, the onset of the condition is prior to the eruption of skin lesions.[15] In paediatric patients presenting with neurological deficits and abnormalities of basal ganglia on imaging, varicella infection should be considered along with other causes of cerebral infarcts such as heart disease, mitochondrial angiopathy, coagulopathy, collagen vascular disease and moya-moya disease. Obtaining a detailed history of recent viral illnesses is the key to establishing the correct diagnosis.

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