Clinics in Diagnostic Imaging (75)

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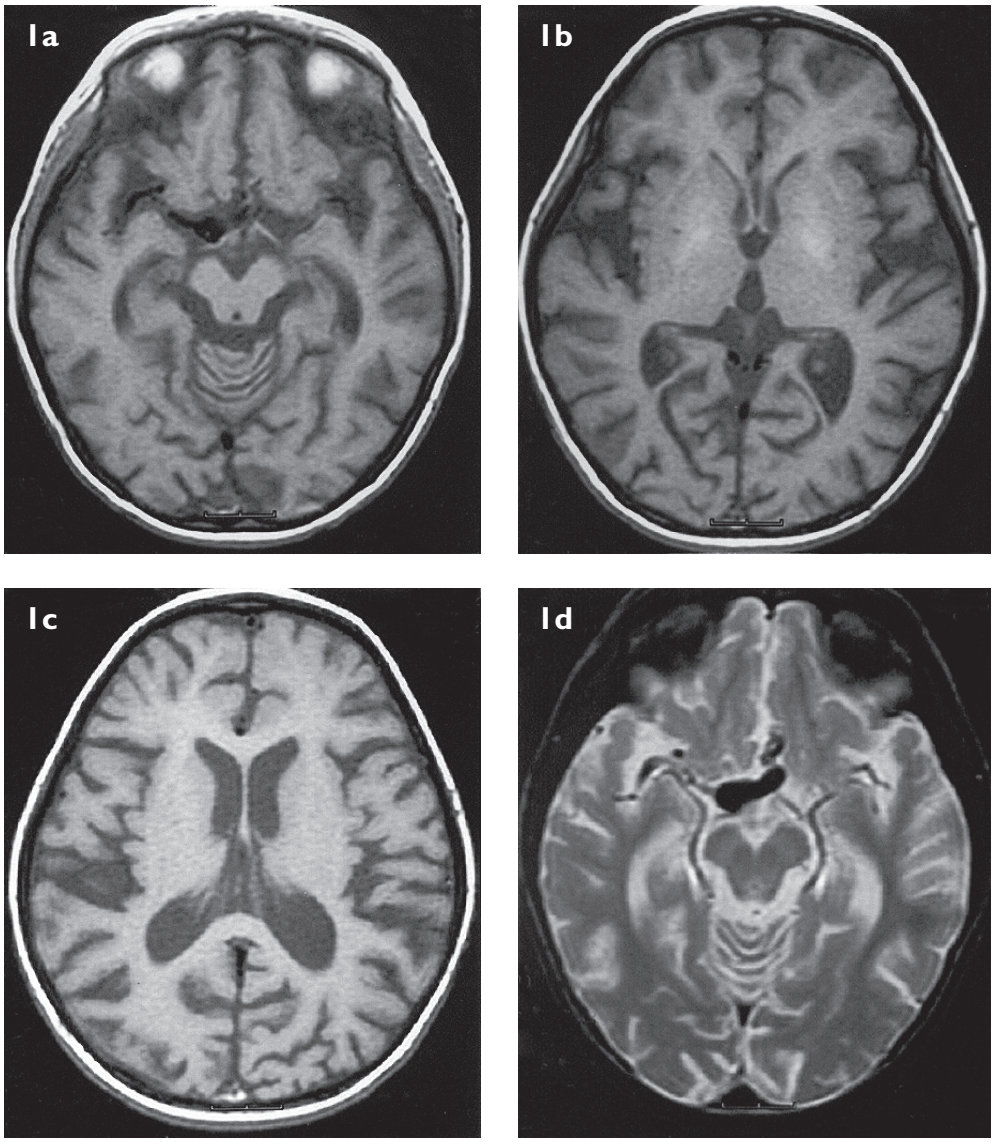

Fig. 1 Axial MR images. T1-weighted scans taken at the level of the (a) midbrain, (b) third ventricle and (c) body of the lateral ventricles. (d) T2-weighted scan taken through the level of the midbrain.

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CASE PRESENTATION

A six-year-old girl presented with a high-grade fever, which she had for a few days. She had been diagnosed as having chronic bilateral otitis media and she also had poor weight gain. Physical examination revealed oral thrush, hepatosplenomegaly, sustained clonus, and hyperreflexia. What do the magnetic resonance (MR) images show (Figs. 1a-d)? What further MR imaging technique is useful? What is the diagnosis?

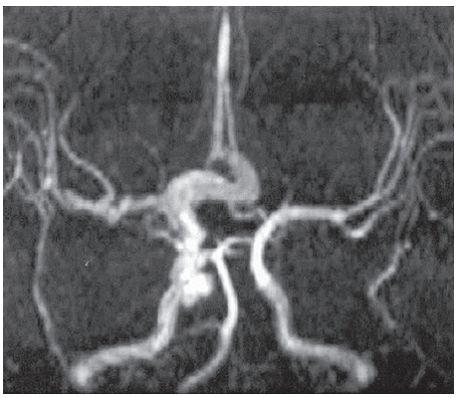

Fig. 2 MR angiography shows fusiform dilatation of the cavernous and supraclinoid segments of the right internal carotid artery and the horizontal segment of the right anterior cerebral artery.

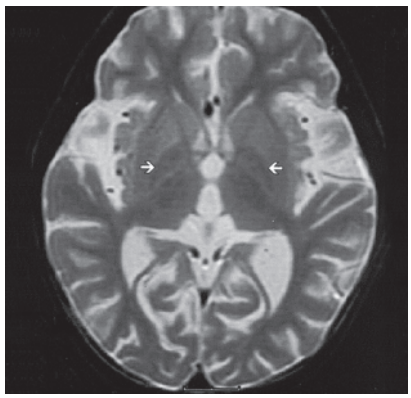

Fig. 3 Axial T2-weighted MR image shows hypointensity of the globi pallidus (arrows) caused by minimal calcification.

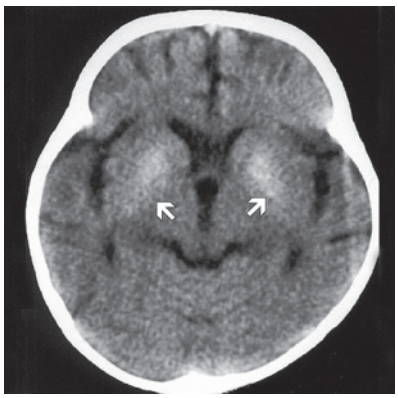

Fig. 4 Unenhanced axial CT scan of a one-year-old girl shows bilateral basal ganglia calcifications (arrows).

IMAGE INTERPRETATION

Axial T1-weighted MR images (Figs. 1a-c) showed diffuse prominence of the subarachnoid spaces and ventricles secondary to atrophy. There is fusiform dilatation of the supraclinoid segment of the right internal carotid artery (Figs. 1a, 1d). Magnetic resonance angiography (MRA) (Fig. 2) confirmed fusiform dilatation of the supraclinoid segment of the right internal carotid artery, and showed fusiform dilatation of the cavernous segment of the right internal carotid artery and the horizontal segment of the right anterior cerebral artery. The globi pallidus are hyperintense on a T1-weighted MR image (Fig. 1b) and hypointense on a T2-weighted image (Fig. 3) because of minimal calcification.

DIAGNOSIS

HIV encephalopathy and cerebral aneurysmal arteriopathy

CLINICAL COURSE

The patient and her parents were found to be human immunodeficiency virus (HIV) positive. Her blood cultures yielded Salmonella choleraesuis. She responded to antibiotics and was discharged from the hospital. Four months later, she was readmitted because of dyspnoea and cough. She died five hours after readmission.

DISCUSSION

The central nervous system (CNS) is often involved in acquired immunodeficiency syndrome (AIDS), resulting in a variety of lesions and diseases. They can be divided into the primary effects of HIV, opportunistic infections, tumours, and vascular disease[1]. The CNS manifestations of AIDS in children differ markedly from those in adults. These differences have been attributed to an immature immune system[2]. In addition, their brains are still developing and they lack previous exposure to most opportunistic pathogens[1].

The most common CNS abnormality in HIV-positive children is HIV encephalopathy, a broad term for the clinical deterioration of higher functions, with associated white matter disease and cerebral atrophy[3]. Clinical manifestations of HIV encephalopathy include developmental delay, loss of previously-acquired milestones, cognitive impairment, microcephaly, weakness, spastic paresis, and hyperreflexia[4]. The imaging findings of HIV encephalopathy are prominence of the subarachnoid spaces and ventricles secondary to atrophy, calcifications of the basal ganglia and subcortical white matter, and white matter lesions. Brain atrophy is predominantly central, with ventricular enlargement[1,3]. The degree of atrophy observed on CT or MR imaging correlates well with the severity of encephalopathy, and may decrease with anti-retroviral agents or correction of contributing factors such as metabolic or nutritional deficiency[1,5].

Basal ganglia calcifications are usually bilateral and symmetrical, involving the globus pallidus and putamen (Fig. 4). Subcortical calcification is most common in the frontal lobes but can occur in other areas of the cerebrum[6]. The extent and progression of calcification correlates with the presence of encephalopathy and its progression[3]. CT is much more sensitive than MR imaging and ultrasonography in detection of these calcifications. White matter lesions are of low attenuation on unenhanced CT, low signal intensity on T1-weighted MR images, and increased signal intensity on T2-weighted MR images. They have no mass effect and are not enhanced by the administration of intravenous contrast media[3]. MR imaging is better than CT in detecting white matter lesions.

Children with HIV infection have a clinical incidence of cerebrovascular disease of 1.3% per year but at autopsy, cerebrovascular disease is found in 24% of children with HIV infection[7]. The mechanisms

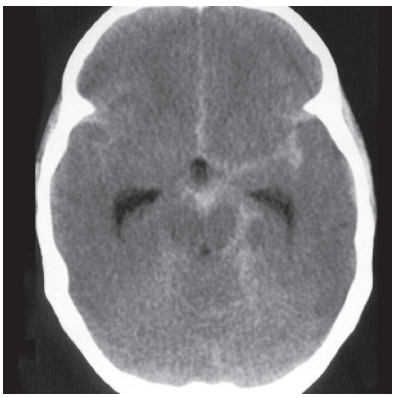

Fig. 5a Unenhanced axial CT scan of a four-year-old boy shows subarachnoid haemorrhage in the anterior interhemispheric fissure, suprasellar cistern, ambient cistern, and along the tentorium cerebelli.

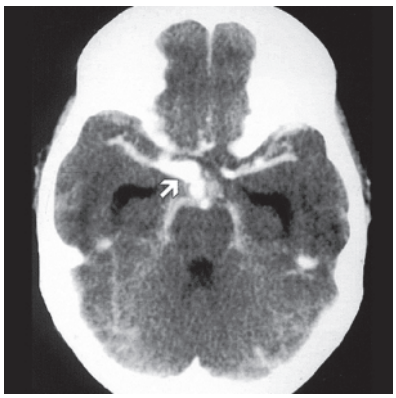

Fig. 5b Enhanced axial CT scan shows fusiform dilatation of the right internal carotid artery (arrow).

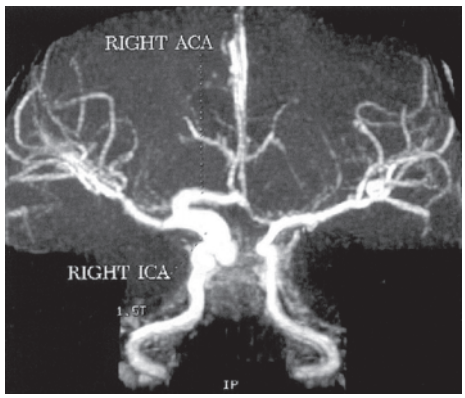

Fig. 5c MR angiogram shows the dilated artery and fusiform dilatation of the horizontal segment of the right anterior cerebral artery.

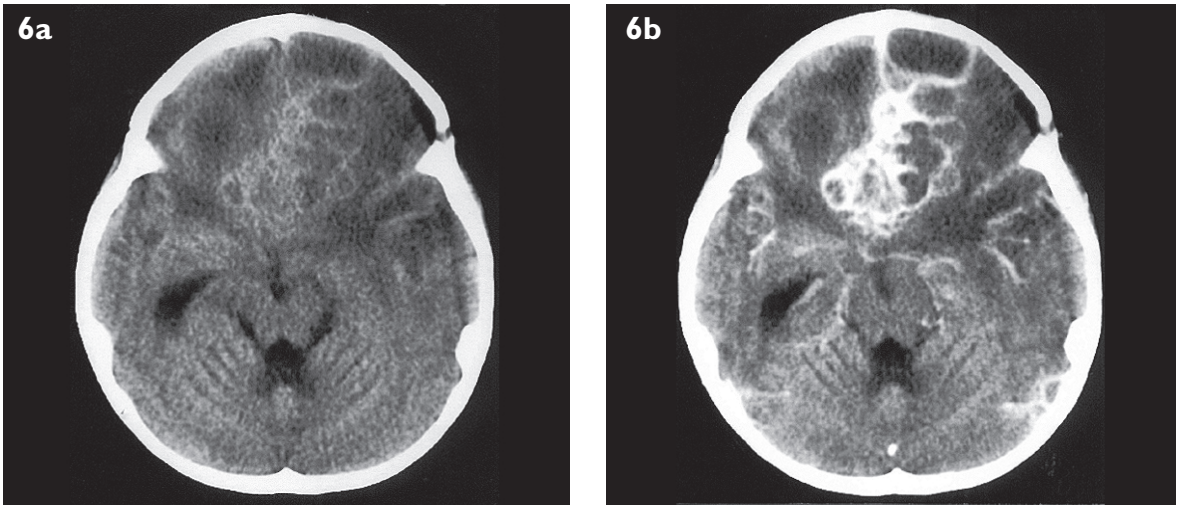

Fig. 6 Four-year-old girl with cerebral nocardiosis. (a) Unenhanced axial CT scan shows a large hetergeneously dense mass in the frontal lobes. (b) Corresponding enhanced axial CT scan shows multiseptated ring enhancement.

include hypoperfusion; septic or thrombotic emboli from endocarditis or cardiomyopathy; thrombocytopenia; and infectious vasculitis of intracranial vessels caused by cytomegalovirus, varicella zoster virus, and mycobacterial or fungal infections[8].

Two uncommon but important cerebrovascular manifestations of HIV virus infection in children are arteritis with formation of fusiform aneurysms, and arterial sclerosis with vascular occlusion[2]. This aneurysmal arteriopathy is confined to the large arteries of the circle of Willis and has been reported as an incidental finding, as in this case, in patients with infarcts and intracerebral haemorrhage[1] (Figs. 5a-c). This aneurysmal arteriopathy may be due to the primary effect of HIV or secondary to opportunistic infections. Medial fibrosis and loss of muscularis with destruction of the internal elastic lamina and intimal hyperplasia are common[8]. It has been reported that once a diagnosis of arterial ectasia has been made, the patient deteriorates rapidly, with death occurring in less than six months[8]. CT or MR imaging is the primary imaging modality for cerebrovascular disease. MR or fluoroscopic angiography best demonstrates either stenosis or aneurysmal dilatation.

Opportunistic infections occur less frequently in children than in adults. HIV-positive children are at risk of fungal, mycobacterial, and nocardial infection, in addition to infection by the pathogens that are the usual causes of meningitis in children[3]. In AIDS patients, the most commonly-reported manifestations of CNS nocardiosis are brain abscesses. However, nocardia also causes meningitis[9]. The CT findings of HIV patients with cerebral nocardiosis have included solitary or multiple low attenuation on unenhanced CT, ring enhancement on enhanced CT, multiloculated ring-enhancing lesions (Figs. 6a, b), hydrocephalus in patients with associated meningitis, and focal small subependymal nodules caused by ventriculitis/ependymitis[9]. The MR appearances of nocardial abscesses are similar to those of other pyogenic brain abscesses. Cerebral nocardiosis is usually associated with pulmonary disease.

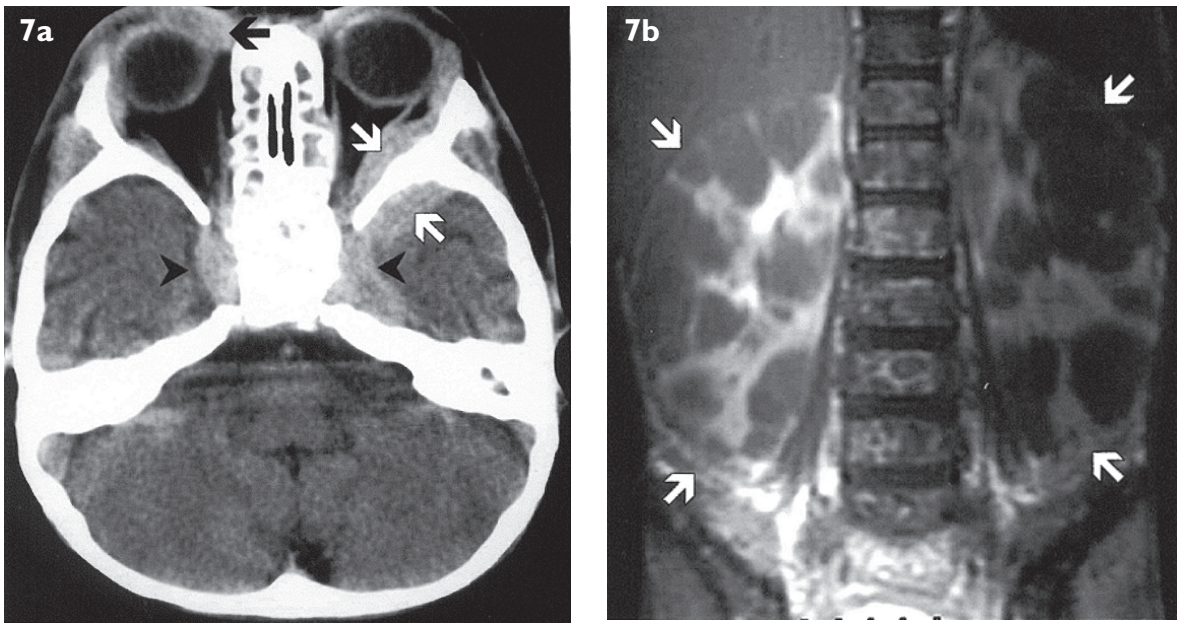

Fig. 7 Eight-year-old boy with non-Hodgkin lymphoma. (a) Enhanced axial CT scan shows extra-cerebral hyperdense masses in the middle cranial fossae (black arrowheads). A soft tissue mass surrounding the greater wing of the left sphenoid bone (white arrows) and a soft tissue mass at the antero-medial portion of the right orbit (black arrow) are seen. (b) Coronal T1-weighted MR image of abdomen of the same patient shows bilateral enlarged kidneys (arrows) with multiple hypointense masses. Note diffuse heterogeneous intensity of the bone marrow.

In children with AIDS, non-Hodgkin lymphoma is the most common cancer[10], and is the most common malignancy associated with HIV infection of the CNS. This lymphoma is associated with Epstein-Barr virus infection, and is either primary or metastatic[3] (Figs. 7a, b). Primary lymphoma is hyperdense on unenhanced CT[2,5] and enhances uniformly. It usually arises in the periventricular white matter, basal ganglia, or corpus callosum. In AIDS, a peripheral location, haemorrhage, and ring enhancement are also common.

ABSTRACT

A six-year-old girl with vertically-transmitted HIV infection had hyperreflexia. MR imaging showed diffuse prominence of the subarachnoid spaces and ventricles caused by HIV encephalopathy. Fusiform dilatation of the supraclinoid segment of the right internal carotid artery was also noted. MR angiography confirmed cerebral aneurysmal arteriopathy. Imaging findings of the various cerebral manifestations and complications found in children with AIDS coming from the primary effects of HIV, opportunistic infections, tumours, and vascular disease are discussed.

Keywords: HIV infections, Acquired immunodeficiency syndrome, Arteritis, AIDS dementia complex, AIDS-related lymphoma.

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