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Unilateral Calcification and Contrast Enhancement of the Basal Ganglia in a Child with AIDS Encephalopathy

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For the majority of adults and children with AIDS, the disease is associated with neurologic manifestations [1, 2]. In children, a progressive encephalopathy has been described that often has characteristic findings on CT scans, including diffuse cerebral atrophy with secondary enlargement of the ventricular system [2–4] and symmetrical bilateral calcifications of basal ganglia and white matter adjacent to the frontal horns [3–4]. In addition, contrast enhancement of the basal ganglia has been noted in association with basal ganglia calcification [4]. This report describes a child who presented with unilateral calcification and contrast enhancement of the basal ganglia, signs of a progressive encephalopathy, and evidence of infection with the AIDS retrovirus HTLV-III/LAV (human T-lymphotropic virus type III/lymphadenopathy associated virus) [5, 6]. The child did not have clinical AIDS. The presentation of HTLV-III/LAV infection in children with neurologic signs and symptoms associated with a focal finding on CT scan makes recognition of the characteristic radiographic appearance imperative for correct diagnosis.

Case Report

A 5-year-old boy presented at age 14 months with idiopathic thrombocytopenic purpura (ITP). His mother was an intravenous drug abuser. The child was subsequently found to be seropositive for HTLV-III/LAV, although his immunologic parameters showed only minimal abnormalities and he did not develop opportunistic infections. A splenectomy was performed at age 2½ years as part of the management of thrombocytopenia. HTLV-III/LAV was grown from resected splenic tissue. The child's perinatal history was unremarkable and his neurodevelopment was normal until age 5, when he presented with two brief generalized seizures that occurred 10 days apart. Additional history indicated that he had become withdrawn, his speech had become dysarthric, and his gait had been mildly unsteady in the 6 to 8 weeks preceding his admission.

Neurologic examination revealed a shy and easily distracted boy

with mildly dysarthric speech. His cognitive abilities were slightly below age expectation. Cranial nerve examination including fundoscopy was normal. His overall strength and tone were within normal limits. There was a generalized awkwardness about all his movements, although specific testing of cerebellar functions showed no abnormality. No adventitious movements were noted. Deep tendon reflexes were abnormally brisk, particularly in the lower extremities. Plantar responses were equivocal bilaterally. His gait was unsteady. He was able to walk on his heels and toes but was not able to balance on either leg or tandem walk. Sensory examination was normal.

CT scan exhibited generalized cerebral atrophy and a discrete calcification in the left basal ganglia (Fig. 1A). After injection of intravenous contrast medium (iothalamate meglumine 1.33 g iodine/kg body weight), an exquisitely circumscribed enhancement of the left corpus striatum was noted, anatomically defining the putamen and the head of the caudate nucleus (Fig. 1B). Neither mass effect nor edema was noted. Digital cerebral angiography performed several days later was normal; specifically, no enlarged or abnormal vessels were seen in the area of the left basal ganglia.

Examination of CSF on two occasions showed acellular fluid with normal glucose and protein. Cultures for bacteria, viruses, and fungi were negative, as was India ink preparation and cryptococcal antigen. Culture of CSF specifically for HTLV-III/LAV was not possible at this institution at the time the specimens were obtained. Intra-blood-brain-barrier synthesis of LAV/HTLV-III antibodies was demonstrated by using matched specimens of serum and CSF [7]. Other routine laboratory studies performed included a complete blood count, serum electrolytes, calcium, phosphorus, and BUN, all of which were normal. TORCH studies of serum revealed no elevation of titres to *Toxoplasma*, cytomegalovirus, rubella virus, *Herpes simplex virus*, or *T. pallidum*.

The child was subsequently discharged from the hospital on a therapeutic dose of phenytoin and has been closely followed in the pediatric neurologic clinic. Over the following 2 months he developed transient tortopelvis, his speech became more dysarthric, and his gait more unsteady.

A repeat CT scan performed 2 months after the initial study (Fig. 2) shows a new finding of enhancement in the right basal ganglia.

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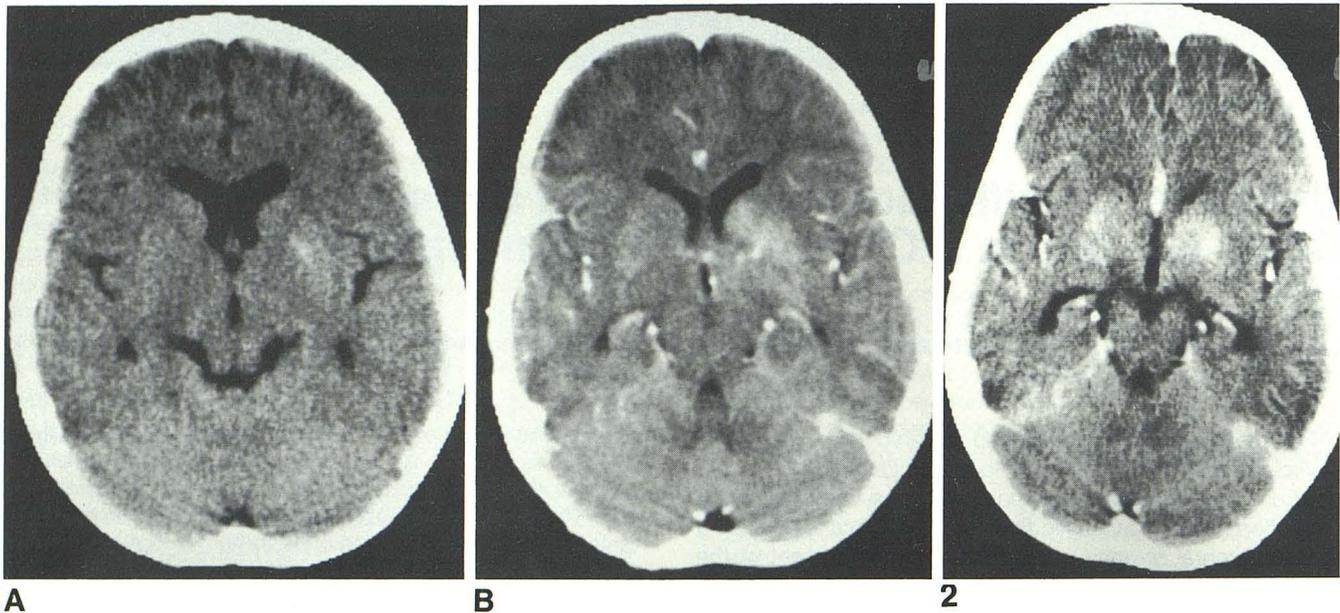


Fig. 1.—A, Noncontrast-enhanced CT scan shows generalized cerebral atrophy with secondary enlargement of ventricular system and a discrete density consistent with calcification in left basal ganglia.

B, CT scan after injection of intravenous contrast medium shows dense enhancement of head of left caudate and putamen.

Fig. 2.—Follow-up CT scan after injection of intravenous contrast medium shows bilateral enhancement of basal ganglia.

Discussion

This child presented with neurologic manifestations of his HTLV-III/LAV infection without the usual manifestations of clinical AIDS. It is anticipated that as the number of drug-abusing mothers infected with the AIDS virus continues to increase, more children with AIDS and with neurologic syndromes associated with HTLV-III/LAV infection will be seen, particularly in urban centers. The variable incubation period from the time of infection until the onset of clinical signs (immunologic or neurologic) in children is often measured in years [8] and L. Epstein, unpublished data). This may obscure the diagnosis for the clinician, particularly when the presentation is neurologic. In this setting, focal findings on CT scan as seen in this case might be particularly difficult to interpret.

When symmetrical bilateral calcifications of the basal ganglia are noted on CT scan in a child, the differential diagnosis should include disorders of calcium metabolism or Fahr's disease. However, in the setting of known HTLV-III/LAV infection, these findings should be interpreted as indicating brain involvement with this virus. Even when the findings are unilateral, certain features point to a diagnosis of HTLV-III/LAV brain infection, including the precise anatomic delineation of the basal ganglia after the administration of intravenous contrast medium and the absence of edema or mass effect. This appearance is distinct from that of lesions of toxoplasmosis commonly seen in adult AIDS patients, which on CT scan are low density, and often multiple, with irregular ring or nodular enhancement [9–12]. The vast majority of cases of

cerebral toxoplasmosis have significant edema surrounding the lesion and clearly evident mass effect, consistent with the necrotic nature of this process [9–12]. Calcifications have not been reported in untreated acquired toxoplasmosis in contrast to those seen in congenital toxoplasmosis [11].

Extensive neuropathologic study of the brains of children who died with AIDS encephalopathy has elucidated characteristic histopathologic findings of inflammatory cell infiltrates, including unique HTLV-III/LAV-infected multinucleated cells, inflammation, and calcification of small- and medium-size vessels [13, 14] (Fig. 3). These changes are consistently found and are most prominent in the basal ganglia. It has been proposed that the vascular inflammation is due to HTLV-III/LAV infection of these structures and that the calcification represents a secondary phenomenon [14]. In children with AIDS, opportunistic or reactivated latent infections of the brain have been infrequent [14] and, in view of the inflammatory changes, it is less likely that metabolic factors would be responsible for the calcification. Last, while vasculitic vessels were not observed at the time of cerebral angiography, such vessels may well have been too small to be visualized. The enhancement demonstrated on CT scan is interpreted as indicating a breakdown of the blood-brain barrier and is consistent with small-vessel inflammation [15]. The repeat CT scan demonstrating bilateral enhancement of the basal ganglia after an interval of 2 months suggests that by serendipity we were able to observe the evolution of a pathologic process that will ultimately show the characteristic symmetrical calcifications of the basal ganglia.

It is concluded that the CT findings described in this child

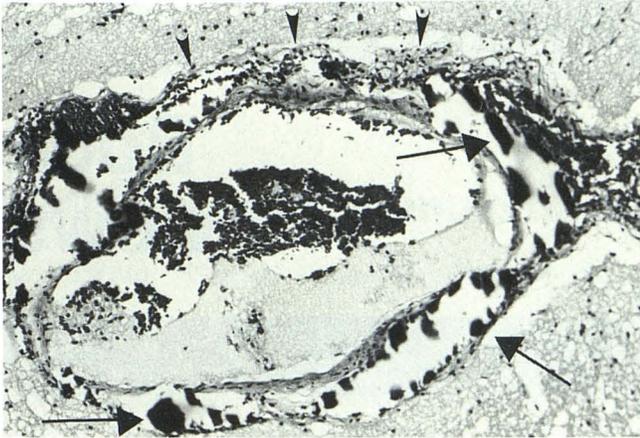


Fig. 3.—Medium-sized artery, putamen, from another child who died at age 5½ months with AIDS encephalopathy. Basophilic calcification (arrows) partly dislodged during tissue handling. Adventitial inflammation (arrowheads) along one edge. (H and E stain, ×98)

are characteristic of HTLV-III/LAV infection of the brain in children and that they correspond to the small-vessel inflammatory changes seen on neuropathologic examination in the basal ganglia. The unilateral appearance on CT scan represents an early manifestation of a process that will ultimately become bilateral. With the availability of serologic tests for the presence of antibody to HTLV-III/LAV, further correlation of clinical and radiologic presentations will be possible.

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