Delayed-Onset Adrenoleukodystrophy after Cerebral Contusion: Progressive Pattern of Demyelination on Serial MR Imaging


We described serial MR findings in a 20-year-old male with adrenoleukodystrophy who presented progressive neurologic deterioration after cerebral contusion. On MR imaging, progressive demyelination was predominant in the white matter of the right temporal lobe as well as in the parietal lobe at the site of prior trauma and extended into the contralateral hemisphere through the anterior commissure.

Index Words: Brain, metabolism
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INTRODUCTION

Adrenoleukodystrophy (ALD) is a metabolic encephalopathy associated with the accumulation of very-long-chain fatty acids (VLCFAs). The disease is inherited as a sex-linked or autosomal recessive (neonatal) trait. Clinically, it commonly begins between the ages of three and ten; symptoms are intellectual impairment, loss of hearing, impaired vision, long tract signs, and adrenal insufficiency.

A few patients who developed clinical ALD after CNS trauma have been reported (1-3). They presented a predominance of intracerebral demyelination at the site of prior cerebral contusion. To our knowledge, a progressive pattern of ALD after CNS trauma has not been well described in the literature. We present a case with ALD who manifested progressive neurological deterioration after cerebral contusion at the age of 20 with emphasis of progressive pattern of demyelination on serial MRI.

CASE REPORT

A 20-year-old man was referred to our hospital from a neurosurgical clinic for the evaluation of recurrent seizure and left hemiparesis that had progressed for 14 months. He had been well until 16 months previously, when he struck the right side of his head in a car accident. Brain CT scan obtained immediately after the injury showed a hemorrhagic contusion in the right parietal cortex (Fig. 1a). At that time, there was no motor or sensory disturbance, but two months later, he developed gait and visual disturbance. On the second CT brain scan, two months after the first, the cerebral hemorrhagic contusion was partially resolved, but newly developed hypodense areas, without enhancement after contrast administration, were present in the white matter adjacent to the site of cerebral contusion. Six months later, the first MR imaging was carried out because of developing progressive left hemiparesis. T2-weighted images showed a wide area of high signal intensity in the white matter of the right temporoparietal areas as well as in a portion of cerebral cortex, including the previous contused region. The lesion appeared predominantly in the temporal lobe and extended into the contralateral temporal lobe through the anterior commissure (Fig. 1b). The lesion included the right side of the optic radiation, the lateral geniculate body, the internal and external capsules, and the corticospinal tract, and these areas were considered to be responsible for the patient’s visual disturbance and
left hemiparesis. T1-weighted images revealed a wide plaque of low signal intensity in the white matter of the right temporo-parietal lobes; contrast-enhanced images after administration of gadopentetate dimeglumine (0.1 mmol/Kg) revealed marked confluent enhancement in the white matter bundles of low signal intensity regions and along the anterior commissure (Fig. 1c). Ten months later, the patient was readmitted because of aggravation of left hemiparesis and development of memory disturbance. The second MRI was performed at that time. T2-weighted images showed widespread diffuse areas of high signal intensity in the right hemisphere, with extension to the left occipital lobe through the splenium of the corpus callosum; T1-weighted images showed more wide plaques of low signal intensity, with atrophy of the white matter of the right temporo-parietal lobe. Contrast-enhanced images revealed thick peripheral enhancement at the margin of the low signal intensity areas. The patient was thought to have post-traumatic organic brain syndrome but had been able to manage without specific medication until that time.

On admission to our hospital 16 months after trauma, the patient was alert, oriented, and fluent. Immediate and recent memory was severely impaired, but remote memory was relatively preserved. Calculation was mildly impaired. There was left hemiparesis that was more severe in the arm than the leg, and walking without support was not possible. Deep tendon reflexes were hyperactive and plantar response was extensor in the left side with ankle clonus. Gait was hemiparetic. Cerebellar dysfunction was absent. Sensory respon-
Adrenoleukodystrophy (ALD) is a genetically determined disorder that is characterized by progressive demyelination of CNS and adrenal insufficiency. It is caused by an enzymatic defect in peroxisomal fatty acid oxidation with resultant accumulation of saturated VLCFAs in plasma, erythrocyte membrane and cultured skin fibroblasts (4,5). As in our case, elevated levels of plasma VLCFAs in plasma is indicative of ALD.

The diagnosis of adult-onset ALD is occasionally difficult because of the variability of initial presentation and symptoms (6). ALD related to CNS trauma is also difficult to recognize, because of its rarity. To our knowledge, five patients with ALD related to CNS trauma have been reported in the literature (1-3). Weller et al. first described MR findings in a patient who developed ALD after cerebral contusion at the age of 57 (1). The case showed a predominance of demyelination in the left temporal lobe of the site of prior cerebral trauma, with extension into the left frontal and occipital lobes. Wilkinson et al. reported two cases with definite features of demyelination at the area of previous trauma on CT scans (2). On the contrary, on the serial MR imaging our case showed that predominant and progressive demyelination occurred in the white matter of the right temporal lobe, as well as at the site of prior trauma in the parietal lobe. In addition to progressive demyelination in the ipsilateral hemisphere, our case showed interhemispheric extension through the anterior comissure. In most cases of ALD, extension through the corpus callosum is the usual pattern presented, with parieto-occipital involvement (6-9). However, interhemispheric extension through the anterior comissure, as in our case has not been previously described. The anterior commissure is a small compact bundle which crosses the midline rostral to the columns of the fornix and has a general shape of bicycle handlebars. This commissure consists of two parts; a small anterior part interconnects olfactory structures on the two sides, while the larger posterior part mainly interconnects regions of the middle and inferior temporal gyri (10). In our case, demyelination of the anterior commissure showed that predominant demyelination had occurred in the right temporal lobe, so that the lesion crossed the midline to the contralateral temporal lobe through a white matter bundle of the anterior commissure. We thought the progressive pattern of our case was based on the anatomical relationship of the association and projection fibers. On serial CT scan and MRI, demyelination started at the site of the prior cerebral trauma of the right parietal lobe and progressed anteriorly to the temporal lobe through the white matter tract including the internal and external capsules, the corona radiate and the visual pathway including the optic radiation, inferiorly into the brain stem through the corticospinal tract, and contralaterally through the anterior commissure and the corpus callosum.

Our case showed marked confluent enhancement in the deep white matter of the temporo-parietal lobe and along the anterior commissure on the first MRI. On the second MRI, the peripheral rim of the demyelination areas was enhanced, and on the third MRI, peripheral enhancement was less pronounced. Contrast enhancement on CT and MR images reflected focal disruption of the blood-brain barrier in active demyelination areas (7-9). Most reported cases revealed peripheral rim enhancement at the leading edge of demyelination areas on contrast enhanced images (1,2,7-9). This contrast enhancement at the edge of demyelination areas showed that the peripheral edge of the lesion was the active demyelinating zone. In our case, a serial change of enhancement pattern suggested that active demyelination had continuously and progressively occurred in a wide bundles of the white matter tract for some time and had been followed by evolution and regression, which had later led to the stage of chronic demyelination. The atrophic change seen in the white matter of the right hemisphere on the second and third MRI also supported this belief.

In summary, we described a patient with rare presentation of ALD which developed after cerebral contusion. Serial CT scan and MRI showed that progressive demyelination started at the site of prior trauma and advanced anteriorly, inferiorly and contralaterally through the white matter tract including the anterior commissure. Serial change in the active zone of demyelination differed according to the time interval after cerebral contusion, and was demonstrated on contrast-enhanced MRI.
REFERENCES


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