

## Multimodal Neuroimages of Adrenoleukodystrophy in Early Stage

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## IV. 5. Multimodal Neuroimages of Adrenoleukodystrophy in Early Stage

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### Introduction

Adrenoleukodystrophy (ALD) is a progressive and genetically determined disorder with an X-linked recessive inheritance. This disorder clinically manifests behavioral disturbances such as bizarre aggressive outbursts, failing memory and poor school performances, and visual impairment at the early stage. We performed the neuroimaging examinations including computed tomography (CT), magnetic resonance imaging (MRI) and positron emission tomography (PET).

The purpose of this study is to evaluate whether neuroimaging methods are useful for early diagnosis of this disorder.

### Subject and Methods

#### Patient

An 8-year-6 month-old boy who had manifested typical symptoms of adrenoleukodystrophy in the course was subjected to this study. The very long chain fatty acid analysis revealed that the ratio of hexacosanoic acid to behenic acid in sphingomyelin fraction of the fasting serum was strikingly high. The diagnosis of adrenoleukodystrophy was made. The CT and MRI were performed at the age of 8 years and 6 months, 3 months after the onset of neurological symptoms. MRI was carried out on a BMT-1000J machine (Bruker, Karlsruhe, West Germany) with a magnetic field of 0.14 tesla. T2 enhanced images were used for evaluation. CT was performed on a new generation machine (Tomoscan 350, Philips, Einhoven, Netherlands).

PET was performed at 8 years and 7 months old. Fluorine-18 was produced at the Cyclotron and Radioisotope Center, Tohoku University and 18F-2-fluoro-deoxyglucose

(<sup>18</sup>F-FDG) was prepared using a fully automated synthesis system.<sup>1)</sup> The patient was already cannulated in the radial artery for serial collection of arterial blood samples, and received 5.3 mCi of <sup>18</sup>F-FDG intravenously from the dorsal vein on the hand. Further detailed explanation of methods for PET procedure was reported elsewhere.<sup>2)</sup> Two PET images at 40 and 55 mm over the orbitomeatal level were scanned for 10 min using an ECAT II (EG & G Ortec, Oak Ridge, TN, USA) with a spatial resolution of 17 mm.

## Results

The CT and MRI are shown in Fig. 1. The CT demonstrated increased density over periventricular zone, and decreased density in the white matter over both posterior regions. No contrast enhancement was observed around the low dense region. The MRI showed normal ventricular size. Marked increase in signal intensity was observed over bilateral posterior white matter on T<sub>2</sub> weighted images. The T<sub>2</sub> relaxation time over the parieto-occipital region was 170 to 250 msec. The lesion was more distinctly and profoundly demonstrated in MRI than in CT.

PET images are shown in Fig. 2. The profoundly lowered CMRglc was clearly demonstrated over both occipital grey matter as well as white matter. The anterior half of the cerebrum showed almost normal CMRglc. The CMRglc in each region of interest is indicated in Fig. 3. Both occipital cortices showed most profound hypometabolism and both posterior temporal cortices were the second. The CMRglc of the occipital and posterior temporal cortex was almost the half of that of the frontal cortex.

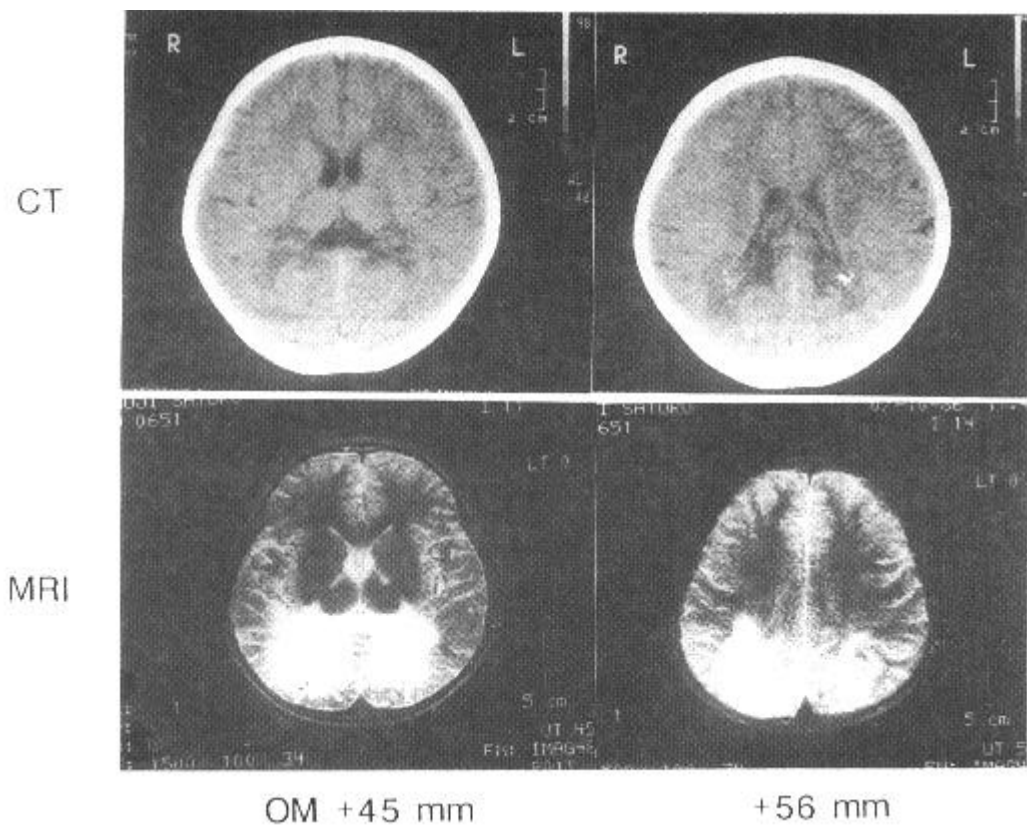
## Discussion

Neuroimaging changes during the course of the progressive disorders such as adrenoleukodystrophy. The initial tests may not show severe abnormalities, however, they give a very important information for the diagnosis. In the typical cases of ALD, the earliest changes of CT findings are low density of the white matter in the parieto-occipital lobes<sup>3)</sup>, and periventricular contrast enhancement.<sup>4)</sup> The CT findings of our patient are thought to be typical as previously reported cases. Recently MRI is suggested to be more sensitive method to distinguish the white and grey matters. Usually MRI of ALD demonstrates demyelination over the posterior cerebral regions at the early stage and widespread demyelination and cerebral atrophy at the advanced stage.<sup>3,5,6)</sup> Our case also showed more extensive lesions over the posterior quadrants in MRI than in CT. The diagnostic value of MRI can be considered to be superior in ALD as suggested by O'Neill<sup>7)</sup> and Kumar et al.<sup>3)</sup>

Volkow et al. reported the PET images for cerebral blood flow and for glucose uptake in a case of ALD of 24-year-old man.<sup>5)</sup> In their report, both cerebral blood flow and glucose metabolism were reduced at the occipital cortex, although their case was tested 10 years after the onset of adrenal insufficiency. These occipitally localized hypometabolism may be sustained for a long period. Their PET image was strikingly similar to ours, although they did not describe the absolute value of the CMRglc of each region. Our results clearly showed the occipital and posterior temporal hypometabolism for glucose and preservation of those in other regions by their absolute values in an early stage. Recently, Chugani et al. reported the normative data of CMRglc in infants and children. According to their report, the CMRglc of frontal cortex and thalamus of control at 8 years old is about 9.8 and 8.9 mg/min/100 g brain (54 and 49  $\mu$ mol/min/100 g brain in their original report), respectively. These values are almost identical to our data. Through our observation, it can be concluded that the glucose metabolism is preserved at the regions other than posterior cerebral area at the early stage of a typical case of ALD. By CT and MR images, the involvement of the parieto-occipital or temporo-occipital white matter is revealed. By PET, however, the occipital grey matter involvement is also suspected.

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ALD 8y 6m (TUCN)

Fig. 1. CT and MRI of the patient. Note more distinct and extensive lesion in MRI than in CT.

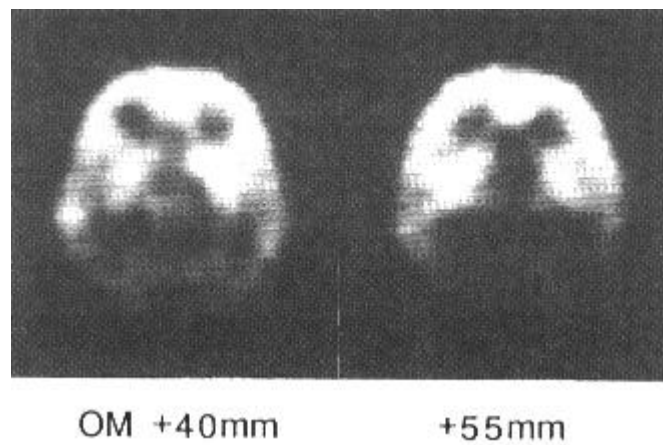


Fig. 2. FDG-PET images of the patient showing very low CMRglc over both white and grey matter in the occipital lobe.

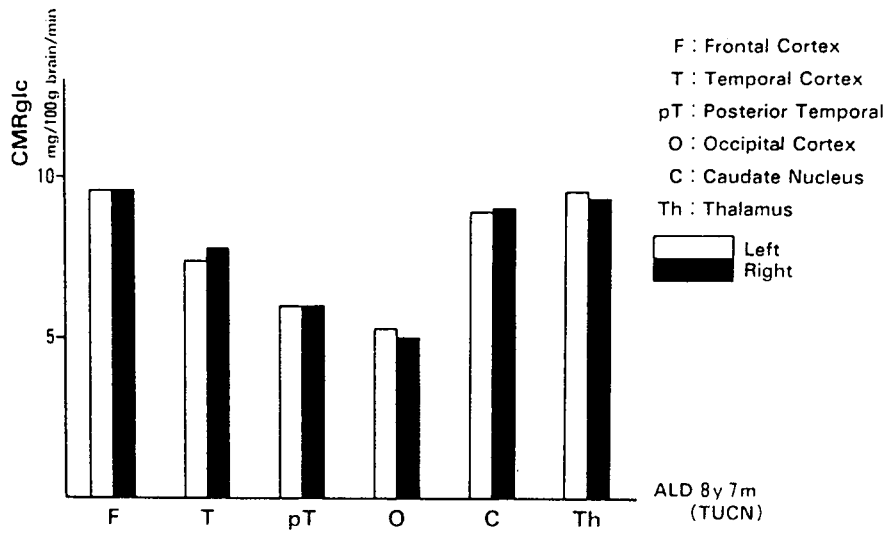


Fig. 3. CMRglc values of each cerebral region.