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# AJNR

## **Dentato-rubral tract involvement in adult-onset adrenoleukodystrophy.**

K Ochi, K Noda, H Kawakami, M Oka, Y Imon, Y Mimori and S Nakamura

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# LETTER

## Dentato-Rubral Tract Involvement in Adult-Onset Adrenoleukodystrophy

Kumar et al investigated the magnetic resonance findings of the brain and spinal cord in a large series of patients with adult-onset adrenoleukodystrophy (ALD), and described tract involvement in the corticospinal, spinothalamic, visual, and auditory pathways (1). Some authors have noticed tract demyelination involving the cerebellar peduncles (2,3). We describe a case of adult-onset ALD with clinical symptoms of spinocerebellar degeneration. Lesions were found mainly in the dentate nuclei of the cerebellum, unilateral superior cerebellar peduncle, and red nucleus.

The patient was a 24-year-old man who had been healthy until age 20, when he first complained of an unstable gait and involuntary movements in the left leg when standing. At age 21 he experienced titubation when descending stairs. Thereafter, he gradually developed dysarthria, writing disturbance, and finger tremor. He was admitted to our hospital for the investigation of these symptoms at age 24. During admission, we tested the patient's verbal and motor aptitude. The patient's verbal, performance, and total scores on a Wechsler Adult Intelligence Scale were 112, 57, and 89 respectively. We noted horizontal nystagmus and faltering speech. There was mild muscle atrophy and bilateral spasticity in his extremities, but no weakness was found. The patient's responses to the finger-to-nose, heel-to-knee, and diadochokinesis tests were most impaired on the right side. He showed tremor and dysmetria predominately in the right arm and leg when he moved. Deep tendon reflexes were exaggerated in all four limbs and the Babinski's sign was positive on the left. He had a staggering and wide gait, and he could not walk in tandem. The results of Romberg's and Mann's tests were positive. Pain and vibratory sensation was mildly elevated in the right lower extremity. The serum cortisol level was normal (10.8 g/dl, 10 min. 18.2 g/dl), suggesting a mild adrenal insufficiency. The very long chain fatty acid ratios (C26/C22, C25/C22, and C24/C22) were markedly elevated in plasma and erythrocyte membranes, a finding compatible with confirmed ALD cases.

The MR image of the brain obtained during admission revealed T2 high-intensity lesions in the right dentate nucleus as well as mild atrophy of the cerebellar vermis, cerebellar hemisphere, and cerebral cortex. Six months after admission, T2 high-signal intensity was observed in both dentate nuclei and in one side of the dentate-rubral tract, extending from the right dentate nucleus through the superior cerebellar peduncle to the left red nucleus (Fig 1). Contrast-enhanced T1 weighted MR imaging revealed lesions in the identical superior cerebellar peduncle and red nucleus, and the cervical cord appeared mildly atrophied.

Tract demyelination in ALD patients involving the corticospinal tract and sensory pathways has been reported (1, 2). Kusaka and Imai reported lesions in the middle and superior cerebellar peduncles revealed by MR imaging of a patient with

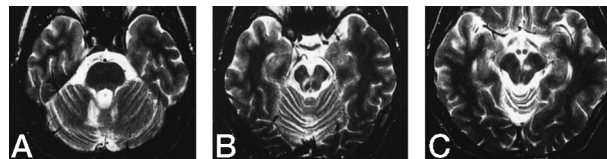


Fig 1. Predominate dentato-rubral tract involvement in 24-year-old man presenting with ALD.

Axial T2-weighted images (3500/112) of the upper pons (A), the inferior colliculus (B), and the superior colliculus (C) show high-intensity signal areas in the bilateral dentate nuclei, the dentato-rubral tract crossing from the right to the left, and the left red nucleus.

ataxic ALD. Their patient's accompanying symptoms, however, were marked atrophy of the cerebellum and pons, and bilateral lesions in the deep white matter of the cerebellum (3). In contrast, the only pronounced lesion we found in our patient was in a dentate-rubral pathway. The findings in our patient may suggest this particular tract involvement may be encountered in other cases of ALD.

Kazuhide Ochi  
Kouichi Noda  
Hideshi Kawakami  
Masaki Oka  
Yukari Imon  
Yasuyo Mimori  
Shigenobu Nakamura  
Hiroshima University  
School of Medicine  
Hiroshima, Japan

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3. Kusaka H, Imai T. **Ataxic variant of adrenoleukodystrophy: MRI and CT findings.** *J Neurol* 1992;239:307-310

## Reply

I have reviewed this case on adult-onset adrenoleukodystrophy. The MR images nicely demonstrate the dentato-rubral tract demyelination which, to our knowledge, has not been previously reported. We have not observed this finding in our large series except that demyelination involving the dentate nucleus was observed in one patient.

A.J. Kumar, MD  
MD Anderson Cancer Center  
Houston, TX