Teaching NeuroImages: Alexander disease with features of both frontal and bulbospinal involvement

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Figure 1 MRI of the supratentorial brain

A 28-year-old woman, who was considered to have Alexander disease (AxD) at 14 months of age, presented with bulbospinocerebellar symptoms from the age of 22 years. Brain MRI showed frontal white matter abnormalities with frontal predominance, and periventricular rim with T2 hypointensity and T1 hyperintensity. Contrast-enhanced axial T1-weighted image (C) shows symmetrical enhancement of periventricular regions. Axial T2-weighted (D) and T1-weighted (E) images show signal changes in basal ganglia and periventricular white matter, with focal target-shaped lesions around the tip of anterior horns of lateral ventricle. Contrast-enhanced axial T1-weighted image (F) shows contrast-enhancing lesions in basal ganglia. These radiologic findings are suggestive of type I Alexander disease.

AxD is classified into 2 subtypes based on onset age and CNS involvement.1 This case suggests AxD can be presented as intermediate form with features of both types I and II.2

Author contributions
T.-S. Nam conceived and designed the study. T.-S. Nam and K.-W. Kang enrolled the subject and collected data. T.-S. Nam and M.-K. Kim analyzed and interpreted the clinical and radiologic data. S.-Y. Choi interpreted the genetic data. M.-K. Kim supervised the study. T.-S. Nam and M.-K. Kim revised the manuscript critically.
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**Disclosure**

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

**References**


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**Figure 2 MRI of the infratentorial brain**

Sagittal T2-weighted image (A) shows mild atrophy and hyperintensities in medulla oblongata, cerebellum, and upper cervical spinal cord. Axial fluid-attenuated inversion recovery (FLAIR) images show pial FLAIR signal changes in midbrain (B), pons (C), and medulla oblongata (D). These radiologic findings are suggestive of type II Alexander disease.
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