

The Cross Sign in the Pons

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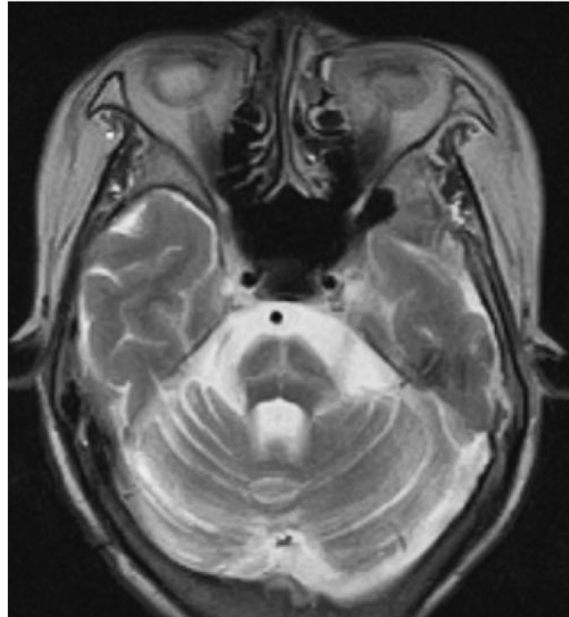


Figure. The cross sign in T2-weighted (TR = 5000ms, TE = 100ms, NEX = 2) imaging: note the cruciform hyperintensity in the pons. Prominent prepontine cistern and cerebellar sulci indicate atrophy of the cerebellum and pons.

Mrs. Yu first noticed clumsiness in the right leg when going upstairs at the age of 56. The symptoms are slowly progressive and she visited our clinics one year later. Physical and neurological examinations revealed mild rigidity in muscle tone (slightly more prominent on the right side). Slowing of rapid-alternating-movement and resting tremor in the right extremities were also noted. There was no muscle weakness or extensor-type plantar response but the muscle stretching reflexes were

slightly increased in the upper extremities. Finger-nose test showed no dysmetria or action tremor. Her gait was slow but steady, with decreased associated movement on the left side. She was treated under the impression of parkinsonism (parkinson's plus). No significant improvement of her motor functions was observed after taking L-dopa at first and then additional dopamine agonist. In addition to deterioration of the foregoing symptoms and signs, limited upward gaze, dysarthria, ataxic

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gait, action tremor in the extremities, hyper-reflexia, and urinary retention all developed in the following two years. However, extensor-type plantar response or orthostatic hypotension was still not observed. A brain magnetic resonance imaging (MRI) study performed at the age of 59 showed atrophy of the cerebellum and the lower part of the pons. In T2-weighted and fluid-attenuation-inversion-recovery (FLAIR) imaging, there were a few small hyperintensities in subcortical white matter, the lateral rim of the left putamen, the pons, and bilateral middle cerebellar peduncles. The hyperintensity in the pons was cruciform in shape (Fig.), as was reported in patients with multiple system atrophy of the cerebellar type (MSA-C)⁽¹⁾.

The term MSA encompasses a variety of degenerative disorders affecting extrapyramidal, pyramidal, cerebellar, and autonomic systems. MSA included disorders previously called olivopontocerebellar atrophy (OPCA, equivalent to MSA-C), Shy-Drager syndrome (SDS), and striatonigral degeneration (SND, equivalent to MSA-P), which have overlapping clinical features. The “cross sign” in the pons (or also known as the “hot cross bun sign”) was first considered to be a specific sign for MSA-C. Burk et al.⁽²⁾ reported that 29 of their 30 patients with MSA-C had cruciform signal abnormalities in the pons on T2- and proton-weighted MRIs. However this sign may also be found in patients with spinocerebellar

atrophy and other diseases.

Although the cross sign is not pathognomonic for MSA, it is informative. Takao et al.⁽³⁾ analyzed the MRI-pathological correlation in a patient with MSA who died of pneumonia. They found that cross-shape T2 signal hyperintensity is primarily ascribable to astrocytosis in the pons rather than to just loss of neurons and myelinated fibers. The horizontal line (of the cross sign) reflects gliosis of the pontocerebellar fibers between the corticospinal tracts and lateral lemniscus, whereas the vertical line reflects gliosis of the middle part of the reticular formation and the crossing part of the pontocerebellar fibers at the basis pontis. Further investigation may disclose the clinical-radiological-pathological correlation of this intriguing sign.

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