Olivopontocerebellar Atrophy: MR Diagnosis and Relationship to Multisystem Atrophy

Clinical diagnosis of olivopontocerebellar atrophy (OPCA) must be confirmed by radiologic demonstration of atrophy in an appropriate distribution. OPCA may be associated with degeneration of other systems in multisystem atrophy (MSA). The authors report 23 cases of OPCA, eight of which were associated with MSA. Atrophy involved the cerebellum, pons, and middle cerebellar peduncles in all cases. On intermediate and T2-weighted magnetic resonance (MR) images, abnormal signal intensity was always observed in the transverse pontine fibers, middle cerebellar peduncles, and cerebellum, structures known from pathologic study to degenerate in OPCA. Pyramidal tracts and superior cerebellar peduncles stood out because of their normal signal intensity. Of the eight patients with MSA, four also had variable abnormal signal intensities in the putamen. The authors believe that the combination of atrophy and abnormal signal intensity in the appropriate distribution strongly supports the diagnosis of OPCA. In some cases, MR imaging may demonstrate involvement of different systems, thus confirming the diagnosis of MSA.

Index terms: Basal ganglia, MR studies, 15.1214 • Brain, atrophy, 15.83 • Brain, diseases, 15.83 • Brain, MR studies, 10.1214 • Brain stem, MR studies, 15.1214

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OLIVOPONTOCEREBELLAR atrophy (OPCA) is a degenerative disease characterized by atrophy of the pons, middle cerebellar peduncles, and cerebellar hemispheres (1). OPCA may present a spectrum of clinical features. It can occur in both familial and sporadic cases. Some nondistinctive enzymatic defects have been reported (2,3). Attempts have been made to organize this nosologic entity by separation into five groups (4). Moreover, transitional and mixed forms exist, in which involvement of the extrapyramidal system and autonomic failure are present in addition to the cerebellar disorder; these forms have been labeled multisystem atrophy (MSA) (1).

Because differences between the clinical variations of OPCA may be subtle and because the clinical symptoms and signs may overlap those of other disorders, most authors now agree that radiologic demonstration of atrophy of the proper structures is essential in establishing the diagnosis of OPCA (4–6).

In addition to gross atrophy, we know from pathologic studies that there are characteristic histologic changes, such as the loss of specific fiber tracts and the presence of gliosis in the pons, middle cerebellar peduncles, and cerebellum. The fibers affected in the pons are the transverse pontine fibers, while the pyramidal tracts and the tegmentum are spared (1); in the cerebellum, a variable loss of Purkinje cells may be seen.

Therefore, we looked for magnetic resonance (MR) signal abnormalities that correspond to these histologic features in a series of patients with the tentative clinical diagnosis of OPCA or other cerebellar degenerative diseases.

PATIENTS AND METHODS

OF 2,200 0.5-T MR studies obtained within a 12-month period at the Istituto Nazionale Neurologico “C. Besta,” in Milan, 18 were characterized by a peculiar pattern of atrophy and abnormal signal intensity in the posterior fossa structures. The clinical diagnosis was OPCA in eight cases, MSA in eight cases, a progressive cerebellar disorder in one case, and a cerebellar and brain stem syndrome in one case. On review of the records, the clinical picture in all cases was consistent with OPCA, although eight patients had additional signs of an extrapyramidal disorder or of autonomic failure, pointing to the diagnosis of MSA.

In the same period, three other patients with the tentative diagnosis of OPCA underwent MR imaging. The first patient had signs of multiple areas of demyelination and was given a final diagnosis of multiple sclerosis. The second patient had cerebellar atrophy, but the pons and middle cerebellar peduncles were normal. The third patient had normal MR findings. No definitive clinical diagnosis was reached in the last two patients.

Of the 18 patients in this series, only two had a familial history of OPCA; the other cases were sporadic. There were 12 male and six female patients, aged 33–64 years (mean, 52.7 years).

The duration of disease was 1–10 years (mean, 4.2 years). The MR studies were obtained on a 0.5-T Gyroscan unit (Philips Medical Systems International, Eindhoven, The Netherlands) and included spin-echo (SE) transverse sections with intermediate and T2-weighted images, generally with a repetition time (TR) of 1,900–2,000 msec and echo times (TEs) of 50 and 100 msec, respectively. Section thickness was usually 6 mm (in rare instances, 8 mm). Sagittal TI-weighted images were always obtained with either SE or, more frequently, fast field echo (FFE) sequences. Coronal SE images with the same TR and TE and/or coronal FFE TI-weighted images were obtained in all but two patients. In eight patients, a study was also obtained with a 1.5-T Magnetom.

Abbreviations: FFE = fast field echo, MSA = multisystem atrophy, OPCA = olivopontocerebellar atrophy, SE = spin echo, TE = echo time, TR = repetition time.
RESULTS

All 23 patients had atrophy of the brain stem and cerebellum. This was more marked than the atrophic changes in the supratentorial brain, which were present in only 12 cases. T1-weighted images best showed the distribution of atrophy; the midline sagittal section showed that the atrophy selectively involved the pons with flattening of its inferior part and with loss, therefore, of the normal bulge of the pons above the profile of the medulla oblongata (Fig 1).

On coronal T1-weighted images, besides the cerebellar atrophy, which was more marked in the hemispheres, atrophy of the middle cerebellar peduncles gave a characteristic pointed appearance to the lateral aspect of the pons. This resulted in a loss of the normal rounded appearance of the middle cerebellar peduncles at their origin from the pons (Fig 2). Atrophy was also well shown, within the same distribution, on the transverse sections. No signal intensity abnormalities were seen on T1-weighted images.

Definite slight hyperintensity was always seen on intermediate images and was seen to a lesser extent on T2-weighted images. Signal abnormality involved the transverse pontine fibers ventral to the tegmentum, on the raphe and on the anterior and anterolateral contours of the pons where the transverse pontine fibers merged with the abnormal signal of the middle cerebellar peduncles. Because of their normal signal intensity, the pyramidal tracts stood out (Fig 3).

Diffuse slight hyperintensity in the cerebellum was sometimes difficult to appreciate on transverse sections where supratentorial brain was not available for direct comparison. However, the abnormal increased signal intensity in the cerebellum appeared obvious when the transverse sections included parts of the temporal or occipital lobes or when coronal sections made the comparison easy (Fig 4).

In the supratentorial compartment, particular attention was paid to changes in the basal ganglia.

Of the eight patients with OPCA, seven also underwent 1.5-T studies. In four of the eight who underwent 0.5-T studies, slightly increased signal intensity was observed in one or both of the putamina on intermediate and T2-weighted images (Fig 5). The findings were normal in three of the seven 1.5-T studies. In two cases signal loss was depicted in the putamina on T2-weighted images; the signal loss was equal to or more marked than that in the pallidum. In one case the signal intensity in the putamina was increased, and in another both decreased and increased signal intensity were present in the same structure. The four abnormal 1.5-T studies were obtained in the patients with abnormal 0.5-T examinations. These patients had the...
most marked extrapyramidal signs. On the 1.5-T studies, a fairly marked decrease in signal intensity on T2-weighted images was occasionally observed in the substantia nigra. Smudging of its borders was present in three cases of OPCA with MSA and in two cases of OPCA without MSA. Ten of the 23 patients had one or more small areas of increased signal intensity in the periventricular or subcortical white matter of the cerebral hemispheres on T2-weighted images. These abnormalities were more frequent in older patients.

**DISCUSSION**

Since 1900, when Déjerine and Thomas first introduced OPCA, the concept of this degenerative disease of the cerebellum and brain stem has become more variegated (1). Subtypes have been created to account for different clinical aspects and inheritance patterns (4). Even the name has been questioned; since degeneration of the inferior olives is secondary to cortical cerebellar lesions, the disease could simply be termed "pontocerebellar atrophy" (1).

Since the clinical features of several cerebellar degenerative and non-degenerative diseases can overlap those of OPCA, and since a unifying biochemical marker is lacking, positive imaging findings have become essential in diagnosing OPCA (4-6). The diagnosis can be made when there is a combination of the appropriate clinical picture and the appropriate brain stem and cerebellar atrophy demonstrated by means of computed tomography or MR imaging (4-8) (Figs 1, 2). A further step toward specificity is the demonstration of abnormal signal intensity in structures that are known to degenerate with this disease. Previous reports did not mention abnormal signal intensities in patients with OPCA (5,6,9,10).

From pathologic studies, we know that in OPCA there is a "dying-back" process of the pontocerebellar fibers, which originate from the pontine nuclei. These fibers have a transverse course in the pons (hence their name, transverse pontine fibers) and run to the cerebellum through the middle...
cerebellar peduncles. A variable degree of degeneration of the Purkinje cells and of their fibers that project to the dentate nuclei is observed. There is also a retrograde cell loss of the inferior olives, secondary to cortical cerebellar lesions (1).

The MR findings in this series of patients match those expected on the basis of OPCA histopathology, in terms of both atrophy and abnormal signal intensity (Figs 3, 4). Not only are the abnormalities observed where they should be, but the structures that should be spared are spared. That is shown in the case of the pyramidal tracts (which become individually demonstrated due to degeneration of the surrounding transverse pontine fibers), the pontine tegmentum, and the superior cerebellar peduncles (1) (Figs 3, 4, 6).

The degeneration of the projections of the Purkinje cells to the dentate nuclei makes these nuclei become gliotic. However, the cells of the dentate nuclei are preserved and so are their projections to the red nucleus and the thalamus through the superior cerebellar peduncles (1). These peduncles are particularly well demonstrated on MR coronal sections, because their normal signal makes them visible against the degenerated background (Fig 4b).

A problem that emerges from this study is the constitution of appropriate imaging support for the diagnosis of MSA. Since MSA is due to the combined degeneration of different systems, the expected MR findings should also involve different structures. Other investigators (11-13) have already reported abnormalities in the region of the substantia nigra, with pathologic hypointensity on T2-weighted images and smudging of its borders toward the red nucleus in Parkinson disease and parkinsonian syndromes. Marked hypointensity in the putamen on T2-weighted images obtained at high field strengths that is equal to or more evident than that in the pallidum has been reported in Shy-Drager syndrome (11,14,15).

Increased signal in the putamen has also been observed on images obtained at 0.5 T (15).

These nonconflicting features may correspond to an increased amount of iron or other paramagnetic elements, as well as to cell loss and gliosis (Fig 5). Both of these features—the presence of neuromelanin and "hematin" pigments and the presence of gliosis and cell loss—are seen in the putamen in striatogniral degeneration (16). Striatogniral degeneration, on the other hand, is an associated or constitutive finding in a high proportion of cases of Shy-Drager syndrome (1). Therefore, what we see in the basal ganglia in patients with Shy-Drager syndrome is perhaps the component of striatogniral degeneration.

In our series, only 13 studies were obtained with a 1.5-T unit. Of the seven studies in cases of OPCA with MSA, four showed increased or decreased signal, or both, in the putamen. Of the six cases of OPCA without MSA studied with the 1.5-T unit, only one showed decreased signal intensity in the putamen. If we compare the 0.5-T studies, available in 18 cases, we find no normal signal in the basal ganglia in 10 cases of OPCA without MSA, while abnormalities are seen in the putamen in four of the eight cases of OPCA with MSA. Therefore, although involvement of the basal ganglia was seen less definitely and constantly than involvement of posterior fossa structures, our data support the hypothesis that MR imaging can contribute to the diagnosis of MSA by demonstrating involvement of the structures of different systems.

References

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