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Thin-Section CT of Midbrain Abnormalities in Progressive Supranuclear Palsy

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Thin-section computed tomographic (CT) scans of 3 and 1.5 mm thickness were obtained using the Philips Tomoscan 310 and General Electric 8800 CT/T scanners in the study of 10 consecutive patients with progressive supranuclear palsy (PSP) and 31 patients with other diseases. Marked midbrain and moderate pontine atrophy, a dilated third ventricle, and enlarged quadrigeminal plate cisterns were observed in all PSP cases. The aqueduct was dilated in several. In six of the PSP cases, there was a striking midbrain abnormality in the form of a low-density area extending from the interpeduncular cistern toward the aqueduct. Thin-section metrizamide-enhanced cisternography of three of the six PSP cases showed that the low-density abnormality was the result of the interpeduncular cistern invaginating the atrophic midbrain.

Progressive supranuclear palsy (PSP) is a disorder characterized by varied gaze palsies together with dementia and cerebellar, extrapyramidal, pyramidal, and pseudobulbar signs. The diagnosis is primarily clinical, although some pneumoencephalographic and computed tomographic (CT) abnormalities have been reported [1-3].

The pathologic changes described by Steele et al. [4] in their original report have been confirmed by others [5-7]. While gross changes were marginally apparent in the cerebrum, there was marked degeneration in the midbrain, including superior colliculi, red nuclei, periaqueductal gray matter, substantia nigra, reticular formation, and tegmentum. The oval part of the tegmentum of the pons was shrunken and the fourth ventricle enlarged. The superior cerebellar peduncles and dentate nuclei were shrunken as well. Except for the corpus Luysii and midline thalamic nuclei, more rostral cerebral structures showed little gross abnormality despite microscopic degenerative changes.

Despite such marked brainstem changes, CT scans with contiguous posterior fossa cuts of 4-8 mm thickness had revealed little of significance in our prior investigation of a number of PSP patients. However, the use of thin-section (1.5 and 3 mm) high-resolution scanning impressed us with the consistently abnormal appearance of the midbrain in 10 cases of PSP. In this article, the technique of thin-section scanning refers to 1.5 and 3 mm thickness scans.

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Subjects and Methods

Computed tomography (CT) was performed using the Philips Tomoscan 310 and the General Electric 8800 CT/T scanners. Eight patients with PSP were scanned with the Philips scanner. Two of these eight patients were later scanned on the GE scanner. Two of our most recent patients were scanned on the GE scanner. The 10 cases with PSP represented all of the patients with PSP admitted to the hospital during the period of the study. The clinical features of the 10 cases are summarized in table 1. Eleven patients with Parkinson disease of 3-12 years' duration and of moderate to marked severity, 10 with presumed Alzheimer disease, and 10 with various other cerebral disorders were scanned using the GE scanner.

Slightly different techniques were used on the two scanners. Patients scanned on the GE

TABLE 1: Clinical Features in Progressive Supranuclear Palsy

	Case No.									
	1	2	3	4	5	6	7	8	9	10
Duration of illness (years) . . .	10	4	5½	4	4	3½	3	6	3	3
Age, gender	61, M	57, M	71, M	72, M	72, M	60, M	67, M	68, F	70, M	77, M
Axial dystonia	+	+	+	+	+	+	+	+	+	+
Limb rigidity	+	+	+	+	+	+	+	+	0	+
Bradykinesia	+	+	+	+	+	+	+	+	0	+
Dementia	+	+	+	+	+	+	0	+	+	+
Facies (stare)	+	+	+	+	+	+	0	+	0	+
Tremor	+	+	+	0	+	0	0	+	0	0
Gait (abnormal)	+	+	+	+	+	+	+	+	+	+
Pyramidal tract signs	+	0	+	+	+	0	0	+	+	0
Cerebellar signs	0	0	+	+	0	+	+	0	+	+
Frequent falls	+	+	+	0	+	0	0	0	+	+
Dysphagia	0	0	0	0	+	0	0	0	0	+
Dysarthria	+	+	+	+	0	+	+	+	0	+
Poor emotional control	+	+	+	0	+	0	0	+	+	0
Eye movements:										
Voluntary vertical, up	0	0	D	D	D	0	D	0	D	D
Voluntary vertical, down	D	0	D	D	D	D	D	0	N	D
Vertical oculocephalic, up	0	N	0	0	0	D	D	N	N	D
Vertical oculocephalic, down	0	N	N	N	N	D	N	N	N	N
Vertical pursuit, up	0	0	D	D	D	0	D	D	D	D
Vertical pursuit, down	D	D	D	D	D	D	D	D	N	D
Voluntary horizontal, right	D	D	D	D	D	D	D	D	N	D
Voluntary horizontal, left	D	D	D	D	D	D	D	D	N	D
Horizontal oculocephalic, right	N	N	N	N	N	N	N	N	N	N
Horizontal oculocephalic, left	N	N	N	N	N	N	N	N	N	N
Bell phenomenon	00	N	00	00	00	00	00	00	N	N

Note.—+ = present; 0 = absent; N = normal; D = diminished.

machine were first studied in a routine manner using 10-mm-thick sections without "targeting." The pons and midbrain areas were identified and 1.5-mm-thick sections at 3 mm intervals were obtained using a 2.1 target factor. The scanning plane was parallel to the canthomeatal line. Those patients studied on the Philips unit had sections through the pons and midbrain at -20° and $+10^\circ$ to the canthomeatal line. These sections were 3 mm thick at 3 mm intervals using varying degrees of targeting. Exposure times were similar for the two scanners, about 10 sec. Milliampereage selection was a compromise between the need for high resolution and the long cooling times associated with the higher milliampereage settings. Intravenous contrast-enhanced CT scans, obtained initially, were discontinued when it was realized that the unenhanced CT scan was sufficient to demonstrate the brainstem abnormalities.

The initial phase of investigation of the CT findings in patients with PSP took place before the installation of a CT scanner at the Washington, DC Veterans Administration Medical Center. Different scanning procedures were used at the two institutions because of the different physical capabilities of the machines used. The gantry of the Philips 310 was capable of tilting 5° more than the GE 8800; the GE 8800 had greater capacity to reformat images. Concern was expressed that there may have been different findings in the PSP patients and the controls because of the different scanning protocols used. Two patients with PSP previously scanned on the Philips 310 scanner were available to be scanned on the GE 8800 at the Veterans Administration Medical Center. The comparison between the two machines was carried out by two processes. First, the two PSP patients were scanned using the protocol for the Philips machine by using a different tilting of the patients' heads so that the angle of

scanning matched. Second, a series of 1.5 mm sections were obtained at 0° . The Arrange program was used to reformat the 0° images to mimic the images obtained on the Philips unit. The angle of sectioning was matched to within a few degrees and the slice thickness was matched within 0.2 mm. CT studies in several control patients were also reformatted. The reformatted GE images of one of the two PSP patients appeared nearly identical to the Philips scans (fig. 1).

The last phase of the investigation consisted of metrizamide-enhanced, thin-section CT of the pontomesencephalic region in three PSP patients in a further attempt to determine the origin of the midbrain abnormality. An intrathecal injection of 10 ml of 170 mg I_2 /ml metrizamide was performed, and the patient was placed in the Trendelenburg position for several minutes. Contiguous 1.5 mm sections parallel to the canthomeatal line were then obtained through the pons and midbrain. Reformating of the images was carried out in oblique planes approximating sections at -20° and $+10^\circ$.

Results

Marked atrophy of the midbrain and moderate atrophy of the pons were present on CT in all 10 cases of PSP (figs. 2–7). In only one instance was there midline atrophy of the cerebellum and in one the fourth ventricle was enlarged. The third ventricle was dilated and the quadrigeminal plate cistern enlarged in all cases. In several instances, the aqueduct was dilated.

In six patients with PSP (cases 1, 2, 5, 8, 9, and 10) (figs.

Fig. 1.—Case 5. Reformatted midbrain CT scans on GE scanner. **A**, Atrophy, dilated aqueduct, and enlarged cisterns. **B**, Low-density midline abnormality. Reformatted images were nearly identical to those obtained with Philips scanner (cf. figs. 6A and 6B).

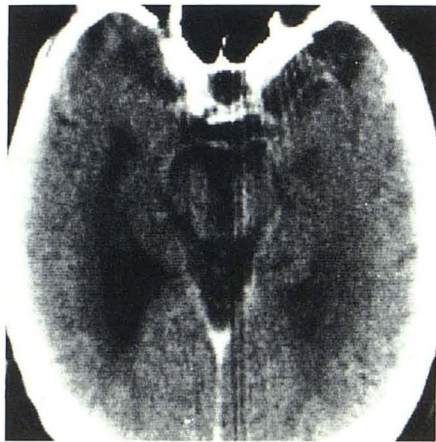
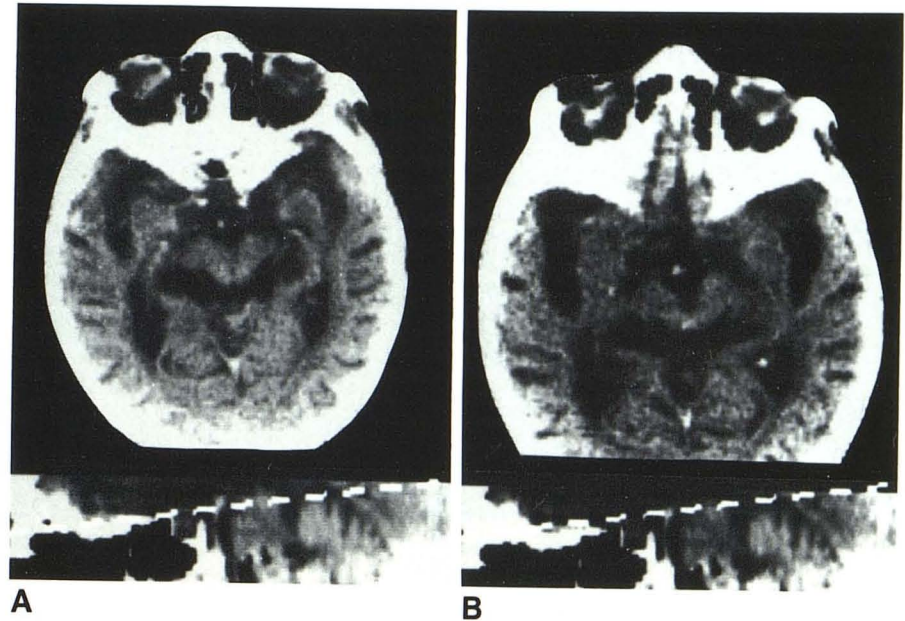


Fig. 2.—Case 1. Midbrain section. Atrophy and striking abnormality in form of low-density area extending from interpeduncular cistern toward aqueduct.

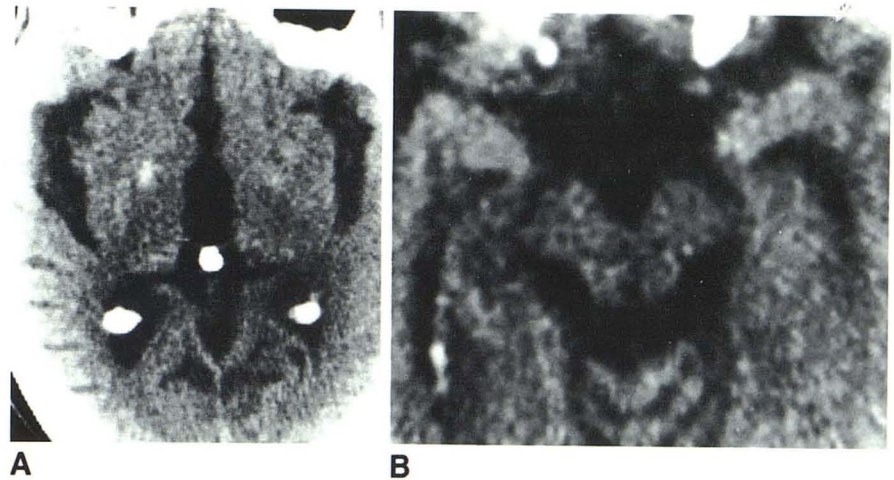


Fig. 3.—Case 2. **A**, CT scan through thalamus. Dilated third ventricle. **B**, Enlarged view. Atrophy of midbrain with dilated aqueduct, enlarged quadrigeminal plate cistern, and widened interpeduncular cistern.

2, 7–9), there was another striking abnormality in the midbrain in the form of a low-density area that extended from the interpeduncular cistern toward the aqueduct. Reformating of sections in two of the six patients with this defect showed that the enlarged third ventricle was invaginating the midbrain in the uppermost sections and that the interpeduncular cistern was causing a similar abnormality in the sections slightly below the level of the pineal. Metrizamide cisternography of three of the six patients with the midline defect showed that the third ventricle was not involved in producing the defect, but that this low-density abnormality was due to an enlarged interpeduncular cistern invaginating the midline midbrain area (figs. 8 and 9).

The midbrain, pons, and cerebellum in all 11 cases of Parkinson disease were normal and the quadrigeminal plate cistern was not enlarged.

The CT scans of another group of 10 patients with presumed Alzheimer disease and 10 with various cerebral disorders including hereditary degenerative cerebellar disease, multiple cerebral and cerebellar infarctions, brainstem infarction with "locked-in" syndrome, hydrocephalus with spastic paraparesis, and Huntington chorea were also studied. The midbrain and pons were normal in all the cases with the exception of Huntington chorea, which showed mild atrophy, and the brainstem infarct, which revealed a low-density area of infarction in the pons and a normal-sized midbrain.

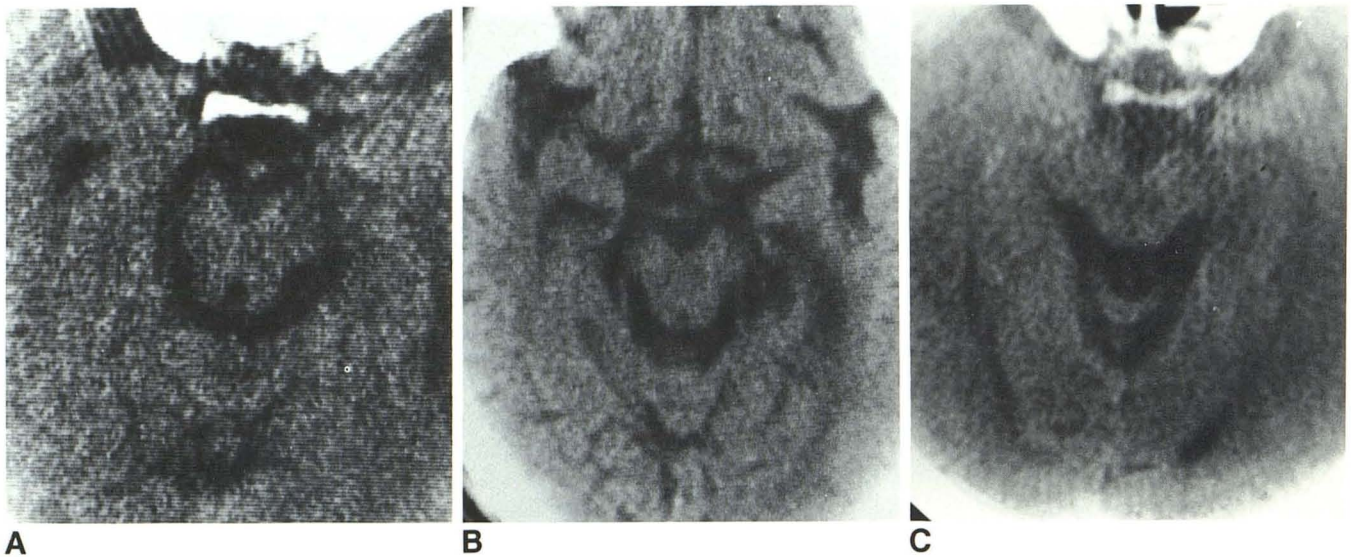


Fig. 4.—Cases 3 (A), 6 (B), and 7 (C). CT sections through midbrain. Atrophy (small midbrain), enlarged quadrigeminal and interpeduncular cisterns, and dilated aqueducts in cases 3 and 6.

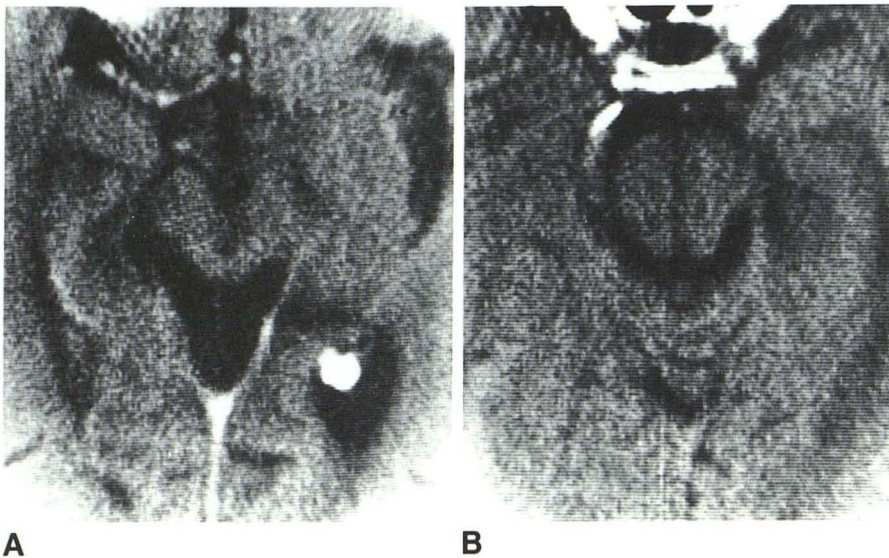


Fig. 5.—Case 4. A, midbrain scan. Atrophy, dilated aqueduct, enlarged quadrigeminal plate cistern, and midline low-density abnormality. B, Upper pons. Generalized atrophy, enlarged cisterns, and midline low-density artifact.

Discussion

Diagnostic procedures in general had been considered to be of little value in the diagnosis of PSP. However, Bentson and Keesey [1] analyzed the mean interpeduncular fossa-aqueduct distance on pneumoencephalograms in six patients with PSP and found severe atrophy of the midbrain tegmentum and atrophy of the superior colliculi [1]. Patients with Parkinson disease and with various cerebellar degenerative disorders showed much less atrophy. Thus, the pneumoencephalogram was considered to be of differential value in the diagnosis of PSP. However, the advent of CT has resulted in the rare use of pneumoencephalography. In 1981, Haldemann et al. [2] reported three patients with PSP who demonstrated

atrophy of the midbrain, pons, cerebellum, and cerebral hemispheres on CT scans through the use of sagittal and coronal reformatting of axial images. In 1981, Ambrosetto and Kim [3] reported a PSP patient with an abnormal routine CT scan showing a very small midbrain with prominent interpeduncular, crural, and ambient cisterns; atrophy of the quadrigeminal plate; and enlargement of the aqueduct.

The thin-section CT brainstem scanning techniques used in our study revealed marked atrophy of the midbrain in all 10 cases of PSP. This degree of midbrain atrophy was not seen in any of the control cases. Unlike Bentson and Keesey's pneumoencephalographic findings of mild atrophy in Parkinson disease, we found no CT evidence of midbrain atrophy in any of the 11 cases of Parkinson disease. It is possible

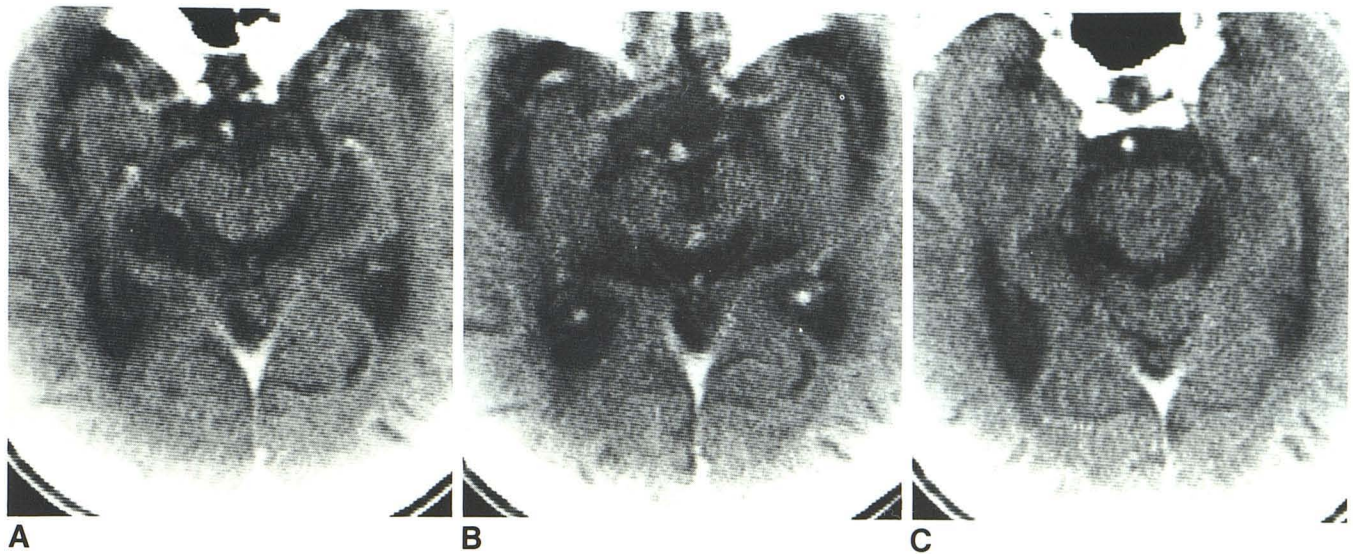


Fig. 6.—Case 5. **A**, Midbrain section. Atrophy and enlarged cisterns including quadrigeminal plate cistern. **B**, Midbrain section. Low-density area extending from interpeduncular cistern toward aqueduct. **C**, Through upper pons. Generalized atrophy.

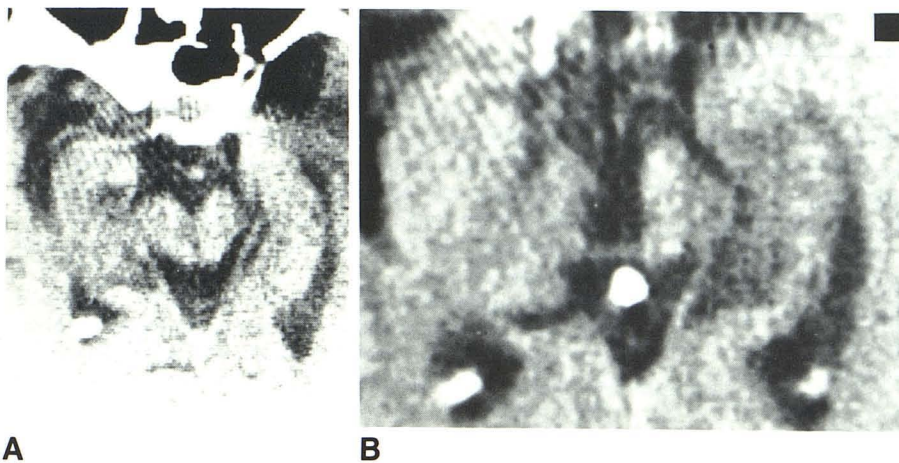


Fig. 7.—Case 8. Midbrain sections. **A**, Atrophy, dilated aqueduct, and enlarged cisterns. **B**, Low-density area extending from interpeduncular cistern toward aqueduct.



Fig. 8.—Case 9. Metrizamide cisternogram. Midbrain section. Atrophy, enlarged cisterns, and enlarged interpeduncular cistern invaginating midline midbrain area.

that CT scanning of a larger number of cases of Parkinson disease might have revealed such atrophy. One case of Huntington chorea showed very mild midbrain atrophy.

The unusual and striking midbrain abnormality observed in six of the 10 cases of PSP, consisting of a density near that of cerebrospinal fluid extending from the interpeduncular cistern toward the aqueduct of Sylvius, had not been described previously. There are several possible explanations for this

low density: (1) atrophy of the midbrain with ex vacuo enlargement of the interpeduncular cistern, (2) degeneration of the midline midbrain structures resulting in decreased attenuation without actual invagination into the midbrain by the interpeduncular cistern, and (3) varying combinations of the above with demonstration of the changes enhanced by the thinness of the sections and the angle of sectioning. No similar invagination of the midline, ventral midbrain has been described at

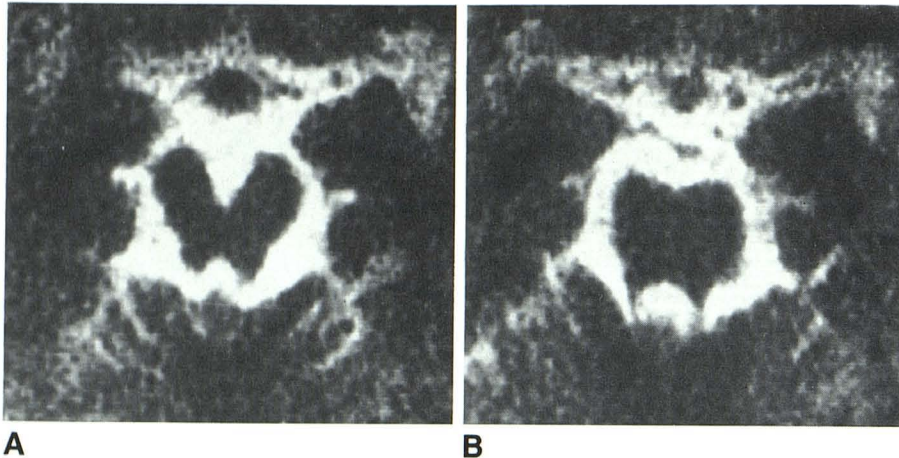


Fig. 9.—Case 10. Metrizamide cisternogram. **A**, Midbrain section. Atrophy, enlarged cisterns, and enlarged interpeduncular cistern invaginating midline midbrain area. **B**, Pontine section. Atrophy of pons; enlarged cisterns.

autopsy. Reformatting of sections in two of the six patients with the midline defect led to the conclusion that the low-density defect in the midbrain was the result of midbrain atrophy with the enlarged third ventricle invaginating the midbrain in the upper midbrain, and the interpeduncular cistern invaginating the atrophic midline area in the sections slightly below the level of the pineal gland. It was postulated that this midbrain defect was due to midline atrophy and partial-volume effect of the third ventricle and interpeduncular cistern. Subsequent metrizamide cisternography of three of the six cases with this midline defect established that the third ventricle was not involved in the defect and that the low-density abnormality was the result of the interpeduncular cistern invaginating the atrophic midline midbrain area.

That the brainstem may be normal at autopsy was shown in one of our PSP patients. This patient was not included in the study because he was hospitalized before CT scanners were available. A normal brainstem at autopsy has been shown in other reports [4, 8, 9], but it is not known how often this occurs. Presumably no abnormality would be shown on CT in such cases.

The clinical differential diagnosis of PSP from Parkinson disease may be difficult. PSP patients originally diagnosed as Parkinson disease have been reported [2, 10, 11]. Some authors doubt the two diseases can be differentiated clinically in many instances [12, 13]. Parkinson disease may present with impaired vertical gaze, including downgaze. Occasionally, even horizontal gaze may be affected.

Our CT findings in PSP and the absence of evidence of midbrain atrophy on CT in Parkinson disease suggest that thin-section CT brainstem scanning methods may be of value in differentiating the early stage of PSP from Parkinson disease. In five of the 10 cases of PSP in our study, the CT scans were obtained within 3 years of the onset of illness. The shortest time from onset of illness to thin-section scanning occurred in case 6, where typical CT abnormalities were present 2 years after onset of symptoms. The striking low-density abnormality of the median ventral midbrain was present in two of our PSP cases (2 and 5) studied by thin-section CT within 3 years of onset of illness.

Though the number of cases of PSP in our study is small, it appears that the midbrain abnormalities observed on thin-

section CT should be helpful in the diagnosis of PSP and in the differentiation from Parkinson disease.

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