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CT in Progressive Supranuclear Palsy

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To determine the usefulness of CT in progressive supranuclear palsy (PSP), the CT scans of 87 patients with extrapyramidal disorders and/or dementia were reviewed. Of these, eight patients whose CT scans showed the findings characteristic of PSP were selected for study. These findings consisted of atrophy of the midbrain and quadrigeminal plate, with prominent interpeduncular, crural, ambient, and quadrigeminal plate cisterns, and dilatation of the aqueduct and posterior third ventricle. All eight patients were found to have PSP while none of the other patients had a clinical diagnosis of PSP. The importance of CT in the diagnosis of PSP, especially in the early phases of the disease, is emphasized.

In 1984 we reported the CT findings of progressive supranuclear palsy (PSP), consisting of atrophy of the midbrain and quadrigeminal plate, with prominent perimesencephalic and quadrigeminal plate cisterns and dilatation of the aqueduct and posterior third ventricle [1]. These data have been confirmed by other authors using CT [2] and MR imaging techniques [3]. In 1985 we reported two additional, less frequent, CT findings of PSP, consisting of a striking midbrain abnormality in the form of a low-density bundle extending from the interpeduncular cistern toward the aqueduct and two bilateral lucent areas in the region of the hippocampus [4]. The aim of this paper is to emphasize the usefulness of CT in the diagnosis of PSP.

Subjects and Methods

The previously described CT findings of PSP [1, 4] were divided into two groups. The first group consisted of major findings, including (1) atrophy of the midbrain and quadrigeminal plate with dilatation of the corresponding cisterns (Fig. 1), (2) dilatation of the aqueduct (Fig. 1), and (3) dilatation of the posterior third ventricle (Fig. 2). The second group consisted of minor findings, including (1) a low-density area in the midbrain from the interpeduncular cistern toward the aqueduct (Fig. 3) and (2) bilateral lucent areas in the region of the hippocampus (Fig. 1).

The CT scans of 87 patients hospitalized for extrapyramidal disorders and/or dementia were personally reviewed. The reviewer knew that the patients were affected by extrapyramidal disorders or dementia but was unaware of the clinical diagnosis. All the examinations were noncontrast-enhanced routine CT scans, with 10-mm-thick sections. The patients whose CT scans showed at least the three major findings were suspected of having PSP; the diagnosis was then checked on the clinical record.

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Results

As shown in Table 1, eight of 87 patients presented at least the three major findings. Two cases also showed the two minor findings. The clinical diagnosis of these eight patients was checked. In seven cases the clinical diagnosis was PSP. In one patient (case 4) the diagnosis was extrapyramidal syndrome. This patient

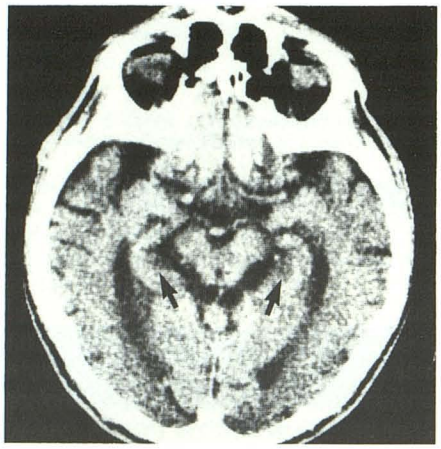

Fig. 1.—CT scan showing atrophic mesencephalon and quadrigeminal plate and prominent perimesencephalic cisterns. Aqueduct is dilated and hypodense areas are present in region of hippocampus bilaterally (*arrows*).

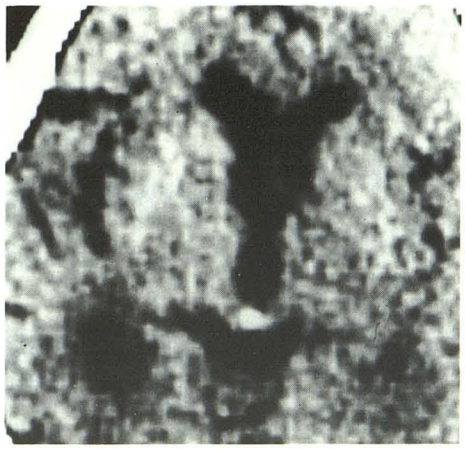

Fig. 2.—CT scan showing dilated posterior third ventricle.

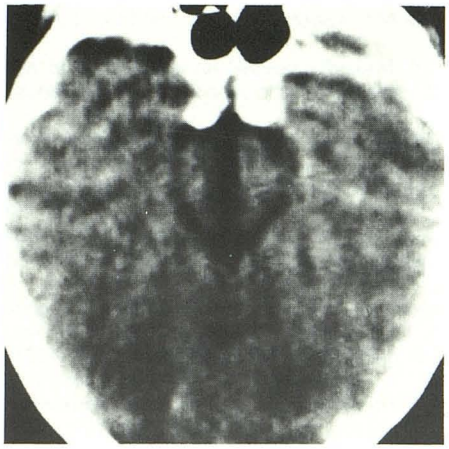

Fig. 3.—CT scan showing hypodense bundle in midbrain from interpeduncular cistern toward aqueduct.

TABLE 1: CT Findings of Patients Affected by PSP

Case No.	Gender	Age	Atrophy of Midbrain and Quadrigeminal Plate	Dilatation of Aqueduct	Dilatation of Posterior Third Ventricle	Hypodense Bundle in Midbrain	Lucent Hippocampal Regions
1	M	72	+	+	+	−	−
2	M	61	+	+	+	+	+
3	F	67	+	+	+	−	−
4	M	58	+	+	+	−	−
5	M	63	+	+	+	−	−
6	M	70	+	+	+	−	−
7	F	65	+	+	+	−	+
8	M	59	+	+	+	−	−

was reviewed clinically to obtain a more precise diagnosis. He had undergone CT 1 year after the onset of the disease and was reviewed for this study 2 years later. At this time he presented two new clinical signs, not detected previously, consisting of axial rigidity and vertical gaze paralysis, thus suggesting the diagnosis of PSP. The patient who also presented the findings of a hypodense bundle in the midbrain (case 2) was reviewed clinically to determine whether he was affected by internuclear ophthalmoplegia (INO) as previously suggested [4]. INO was not found in this patient. Finally, the diagnoses of the 79 patients whose CT scans were not suggestive of PSP were checked. There were no diagnoses of PSP. These 79 patients consisted of 45 men and 34 women, ranging in age from 41 to 79 years (mean, 53 years). The diagnoses included Parkinson's disease (32 cases), Alzheimer's disease (25 cases), normal pressure hydrocephalus (12 cases), Huntington's chorea (6 cases), and Binswanger's disease (4 cases).

Discussion

In the pre-CT era, radiologic diagnostic procedures were considered of little value in the diagnosis of PSP. In contrast, the results of the present paper confirm that CT is of great diagnostic value in this disorder.

As previously emphasized [1], the major CT findings may well correlate with the pathologic features of PSP. In fact, pathologic specimens of the disease show a shrunken mesencephalon with an atrophic quadrigeminal plate, enlarged aqueduct secondary to the periaqueductal gray matter atrophy, and enlarged posterior third ventricle secondary to the atrophy of the posterior medial thalami. In the present paper the characteristic radiologic picture was present in all patients with PSP and was absent in those with different disorders. Of great interest is the fact that the radiologic picture characteristic of PSP was present in case 4 when the clinical diagnosis was uncertain. The axial rigidity and gaze paralysis

were lacking. This case strongly resembles case 2 of our previous paper [1], in which the characteristic CT picture was evident when the clinical hallmarks of PSP were not present. Previous reports of pneumoencephalography [5] and CT [1] in PSP suggest that radiologic features are not related to the duration of the illness and the degree of neurological deficits. However, these two cases seem to suggest that CT findings of PSP could be present when the diagnosis is not yet clinically obvious.

The data are insufficient to allow definitive conclusions, and further studies are advisable to confirm this hypothesis. Patients with extrapyramidal disorders and/or dementia of uncertain diagnosis and with CT scans suggesting PSP should be carefully followed up clinically to identify axial tone alterations and gaze paralysis. The low-density bundle in the midbrain in PSP patients has been reported previously and explained differently [2, 4]. We suggested that this midbrain abnormality could represent the CT expression of a demyelinating process in the medial longitudinal fasciculus on the basis of the patient reported in our previous paper who was also affected by INO [4]. In fact, INO has been reported, even if rarely, in PSP [6, 7] and loss of myelin in the medial longitudinal fasciculus has been pathologically confirmed [8]. On the contrary, Masucci et al. [2], using thin-section metrizamide-enhanced cisternography in three PSP cases, showed that this low-density abnormality was the result of the interpeduncular cistern invaginating the atrophic midbrain. Case 4, showing this abnormality, was not affected by INO. Probably both explanations could be correct, but pathologic correlations are needed to confirm one or both hypotheses.

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