Evolution of central pontine myelinolysis on CT.

S Rosenbloom, D Buchholz, A J Kumar, R A Kaplan, H Moses, 3rd and A E Rosenbaum

*AJNR Am J Neuroradiol* 1984, 5 (1) 110-112

http://www.ajnr.org/content/5/1/110.citation

This information is current as of October 26, 2023.
Evolution of Central Pontine Myelinolysis on CT

Shelley Rosenbloom,1 David Buchholz,2 Ashok J. Kumar,1 Richard A. Kaplan,2 Hamilton Moses III,2 and Arthur E. Rosenbaum1

Central pontine myelinolysis (CPM) was first described by Adams et al. [1] in 1959; since then, well over 100 cases have been reported. The disease has been noted predominantly in alcoholics with electrolyte imbalances, usually hypotension, most especially after electrolyte derangement has been rapidly reversed by vigorous intravenous fluid administration. The characteristic clinical course includes initial neurologic improvement paralleling electrolyte correction and subsequent deterioration, including quadriplegia and pseudobulbar signs. Diagnosis has generally been made on the basis of autopsy material. Before computed tomography (CT), there was no radiologic method for detection. CT findings in CPM have sometimes been reported as negative, but, in a few cases, large pontine hypodense lesions have been uncovered. The following case demonstrates the evolution of these lucencies over the course of hospitalization.

Case Report

A 50-year-old woman with a history of chronic alcohol abuse, diabetes mellitus, and hypertension had the onset of slurred speech and confusion after several days of unexplained diarrhea, nausea, and vomiting. The next day she was found unconscious, presumably after a seizure, and was admitted to a hospital. She was initially unresponsive to pain with hyperreflexia and bilateral Babinski signs. Serum sodium was 108 mEq/L; potassium, 1.6; chloride, 60; and bicarbonate, 41. Depressed serum osmolality and elevated urine osmolality contributed to a clinical diagnosis of inappropriate antidiuretic hormone secretion. During treatment for electrolyte derangement, she improved rapidly and was oriented and cooperative by hospital day 2. One day later she began a progressive decline, and became unresponsive and mute over the next week. In addition, marked hypertension was noted. On hospital day 16 she was transferred to Johns Hopkins Hospital for further evaluation. Physical examination demonstrated a mute patient responsive to pain but not verbal commands. Pseudobulbar signs, marked spastic quadriaparesis, and frontal-lobe release signs were present. CT using an AS&E large-aperture scanner showed normal pontine density, but prominence of the pontine and cerebellopontine angle cisterns and the fourth ventricle compatible with some tissue loss (fig. 1A).

On day 17 of her hospital course (her second day after transfer), she became slightly more alert, still not following commands but occasionally grunting. Another CT scan on that day on the high-resolution Siemens DR3 unit again demonstrated evidence of posterior fossa atrophy with some minimal, questionable mottled lucency in the brainstem (figs. 1B and 1C). Electroencephalography showed bilateral slowing. Cerebrospinal fluid examination was normal except for elevated myelin basic protein at 11.6 ng/ml (normal, 6), suggesting myelin destruction. Brainstem auditory-evoked potentials were indicative of brainstem dysfunction at the pontine-midbrain level. Gradually over the next 2–3 weeks she became more alert, able to talk and follow commands. A follow-up CT scan on the same Siemens DR3 6 weeks after her initial hospitalization demonstrated dramatic change with a large, well margined central lucency now evident throughout the basis pontis and extending into the midbrain (fig. 2).

Discussion

Clinical Features

Our case is typical in its presentation of rapidly evolving flaccid quadriaparesis and mutism in the context of electrolyte disturbance, alcohol abuse, and rapid intravenous hydration. These circumstances have each been noted in more than 70% of the reported cases. Facial weakness and weakness of the tongue with dysphagia may be present also. Level of consciousness may be depressed, and death may follow respiratory arrest. The pathogenesis of the lesion is controversial; a relation between CPM and local edema and electrolyte disturbance is speculated on [2–5].

Pathologic Diagnosis

In the pre-CT era, the diagnosis of CPM was generally established by autopsy findings [1, 6]. A discolored and softened focus in the central pons is described, which is usually well circumscribed, although occasionally it is diffuse and may be surrounded by a zone of gliosis. The lesion may extend rostrally to involve the midbrain, and has been reported in the corpus striatum, thalamus, lateral geniculate bodies, internal capsules, corpus callosum, and gray-white junction in both cerebrum and cerebellum. On histopathologic study, Luxol fast-blue stain shows loss of myelin. The nuclei

Received February 14, 1983; accepted after revision May 27, 1983.

1 Russell H. Morgan Department of Radiology and Radiological Sciences, Johns Hopkins Medical Institutions, Baltimore, MD 21205. Address reprint requests to S. Rosenbloom.

2 Department of Neurology, Johns Hopkins Medical Institutions, Baltimore, MD 21205.

AJNR 5:110–112, January/February 1984 0195–6108/84/0501–0110 $00.00 © American Roentgen Ray Society
Fig. 1.—A, 5 mm cut at level of pons, on day 16. Some prominence of cerebellar folia, pontine cisterns, and fourth ventricle with some mottled density in pons. No definite lucent defect was identified. B and C, 4 mm cuts through pons on day 17. Widening of cerebellar folia and enlargement of pontine cisterns and fourth ventricle, again without evidence of focal pontine lucency.

Fig. 2.—A and B, 4 mm cut through midbrain shows bilateral symmetric parenchymal lucencies. At pontine level a well defined symmetric central lucency is present. These hypodense lesions were demonstrated on hospital day 42. C, Sagittal reconstruction reveals extent of lesion, which includes most of basis pontis. There is also some midbrain extension.

of oligodendroglia are also diminished in number, and myelin debris is present extracellularly and in foamy macrophages. There is relative neuronal sparing.

Differential Diagnosis on CT

Central brainstem lucencies on CT are highly suggestive of CPM in the appropriate clinical context. The differential diagnosis includes multiple sclerosis, brainstem infarction, metastatic disease, brainstem glioma, lymphoma, and brainstem encephalitis. Enlargement of the brainstem is not characteristic of CPM, but may be present in many of these other entities.

Radiographic Features

In the more than 100 cases of CPM that have been described, CT has been reported in only eight, and none of these appeared in the radiologic literature. The first report, that of Messert et al. [2] in 1979, described no positive CT findings in three cases. The scanner used and the relation to the stage of the clinical illness was not specified. Telfer and Miller [7] reported a case with pontine and midbrain lucencies using a GE 8800 CT scanner; these findings had not been detected 26 days earlier when CT was performed on a less advanced CT system. Anderson et al. [8] reported a case in which central brainstem lucency was demonstrated on an EMI scanner (model unspecified), and this lucency showed no change over a 2 week interval. The patient remained essentially normal neurologically during this period. Yufe et al. [9] reported a case of CPM with normal CT findings (EMI Mark I) studied close to the time that the clinical deterioration was beginning; 3 weeks later, on a fourth-generation AS&E scanner, a well defined pontine lucency was demonstrated. These authors suggested that CPM had been missed on their earlier CT scan because of the use of low-resolution technology.

Thompson et al. [10] reported two cases of CPM. In their first case, the CT appearance of the brainstem was initially "normal to gross examination," but 5 weeks later showed pontine lucencies. The authors implied that these lucencies, which were not evident on the earlier axial study, might have been demonstrable on modified coronal sections as they
showed in their second scan. A third CT examination of the patient 4 months later was believed to have shown some resolution of the pontine lucencies. However, artifacts often preclude optimal delineation of the brainstem on CT, and even their published images are somewhat difficult to evaluate. In their second case, CT performed on hospital day 4 was reported as unremarkable except for atrophy. Two weeks later, however, axial-plane CT was suggestive of a pontine lesion (but inconclusive because of artifact). In week 4, a modified coronal-plane CT showed a pontine lucency. The technical aspects of their studies were not published.

Our case documents the evolution of lucencies using the same high-resolution CT system for sequential scans; the change in CT appearance cannot be ascribed to technical differences in the studies, and must be related to progressive demyelination. An initially normal high-quality scan or one demonstrating brainstem or cerebellar atrophy does not exclude the diagnosis of CPM, which may become evident on follow-up studies. Our patient had begun to improve neurologically by the time of the CT study with demonstrated CPM.

REFERENCES