

Neurosyphilis Presenting as a Focal Mass Lesion: A Case Report

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A patient with subacute aphasia and hemiparesis was found to have a low density white matter lesion with mass effect on the computed tomographic (CT) scan. Serological examination and biopsy established the diagnosis of parietic neurosyphilis. This CT appearance has not previously been described in cases of neurosyphilis. Neurosyphilis should be considered as a potentially treatable cause of a cerebral mass lesion. (*Neurosurgery* 14:234-237, 1984)

Key words: Aphasia, Biopsy, Computed tomography, Focal mass lesion, Hemiparesis, Parietic neurosyphilis, Serology

Neurosyphilis, once a major source of neurological morbidity, may still have an incidence as high as 5000 cases annually in the United States alone (8). The computed tomographic (CT) findings in this disease, however, have rarely been reported (3, 4). We report our recent experience with a patient with neurosyphilis whose clinical and radiological presentation was suggestive of a focal cerebral mass lesion.

CASE REPORT

A 59-year-old man was transferred to Saint Vincent Hospital, Worcester, Massachusetts, with increasing confusion. A former nightclub pianist, he had worked for the past several years as a bank guard. He was unmarried and reclusive and had a history of ethanol abuse. One month before admission, his family noted increasing confusion and intermittent difficulty speaking. One week before admission, he developed fever and chills. On the day of admission, he was found by his brother awake and alert, but unable to speak. He was referred to his local hospital, where he was febrile and described as disoriented. A lumbar puncture revealed an opening pressure of 250 mm H₂O, 55 white blood cells (WBCs), 8 red blood cells (RBCs), a protein content of 63 mg/dl, and a glucose content of 69 mg/dl. He received a 5-day course of intravenous nafcillin and chloramphenicol and became afebrile, but remained confused. On the 12th hospital day, he suffered a generalized seizure and was transferred to St. Vincent Hospital.

Physical examination revealed the patient to be awake, alert, and afebrile with stable vital signs. A Grade II/VI systolic murmur was noted. Neurological examination revealed a nonfluent aphasia, with difficulty naming and repeating, poor comprehension, and paraphasic errors. Poorly reactive, irregular pupils and a possible right homonymous field defect were present. He had a mild right hemiparesis, right hyperreflexia, and a right Babinski sign. His gait was markedly unsteady, with a positive Romberg's sign. The sensory examination was limited because of poor cooperation, but sensation was intact to pinprick. Admission serological examination (VDRL) was positive at 1:16. Subsequent FTA-ABS was 4+ positive. Repeat lumbar puncture revealed an opening pressure of 200 mm H₂O, a protein content of 68 mg/dl, 0 RBC, and 3 WBCs. The initial cerebrospinal fluid (CSF) VDRL was reported as nonreactive.

A CT scan with contrast agent demonstrated a large area of decreased attenuation involving the left frontal and tem-

poral white matter. There was slight mass effect on the left frontal horn with frontal and temporal opercular gyral enhancement (Fig. 1). Cerebral angiography revealed a mass effect in the left hemisphere, a small blush in the lateral lenticulostriate region, and early-draining frontal veins.

A biopsy of the left posterior frontal lobe revealed a subacute encephalitis characterized by perivascular cuffing by lymphocytes and plasma cells, microglial proliferation with rod cell formation, focal neuronal necrosis, and fibrillary gliosis (Fig. 2). A Prussian blue stain showed iron deposits in the walls of blood vessels and occasionally in the cytoplasm of the rod-shaped microglial cells. Several attempts to identify *Treponema* organisms using the Dieterle silver method were unsuccessful. The subcortical white matter showed only mild fibrillary gliosis. No tumor or infarction was present. The histological findings were thought to be consistent with, although not diagnostic of, the parietic form of neurosyphilis. Repeat testing on the initial CSF specimen gave a strongly positive VDRL.

The patient received a 10-day course of intravenous penicillin, 18 million units per day. There was resolution of his hemiparesis, but he remained aphasic with a seizure disorder. A repeat CT scan 4 months later revealed complete resolution of the decreased attenuation with ex vacuo dilatation of the left frontal horn and the adjacent cortical sulci (Fig. 3).

DISCUSSION

This 59-year-old man presented with a subacute deterioration in language function with other signs referable to the left hemisphere. Serological testing revealed a positive serum VDRL at 1:16, a 4+ positive serum FTA-ABS, and a positive CSF VDRL. The initial negative CSF VDRL and unusual CT appearance prompted the brain biopsy. Although a specific stain for treponemal organisms was negative, this can be a technically difficult stain to perform and, therefore, the diagnosis of a syphilitic infection was not excluded. Furthermore, the 5-day course of antibiotic therapy that the patient received at the outlying hospital could also have contributed to our inability to demonstrate treponemas in the tissue. We believe that the histopathology is quite characteristic of the parietic form of neurosyphilis (11) and, taken in conjunction with the serological data, it strongly supports this diagnosis.

A variety of CT findings have been described in cases of neurosyphilis. These include irregularly enhancing and nonenhancing low density areas in presumed meningovascu-

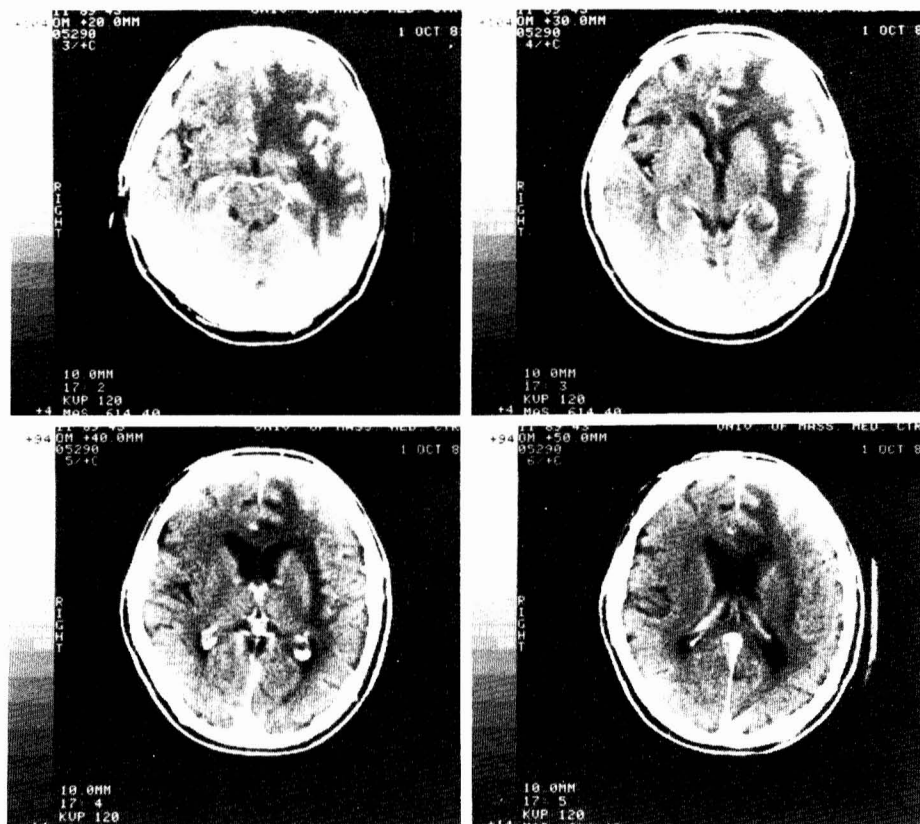


FIG. 1. Initial CT scan after contrast infusion demonstrates decreased attenuation in the left frontal and temporal lobe white matter. Slight mass effect on the left frontal horn and opercular gyral enhancement are identified.

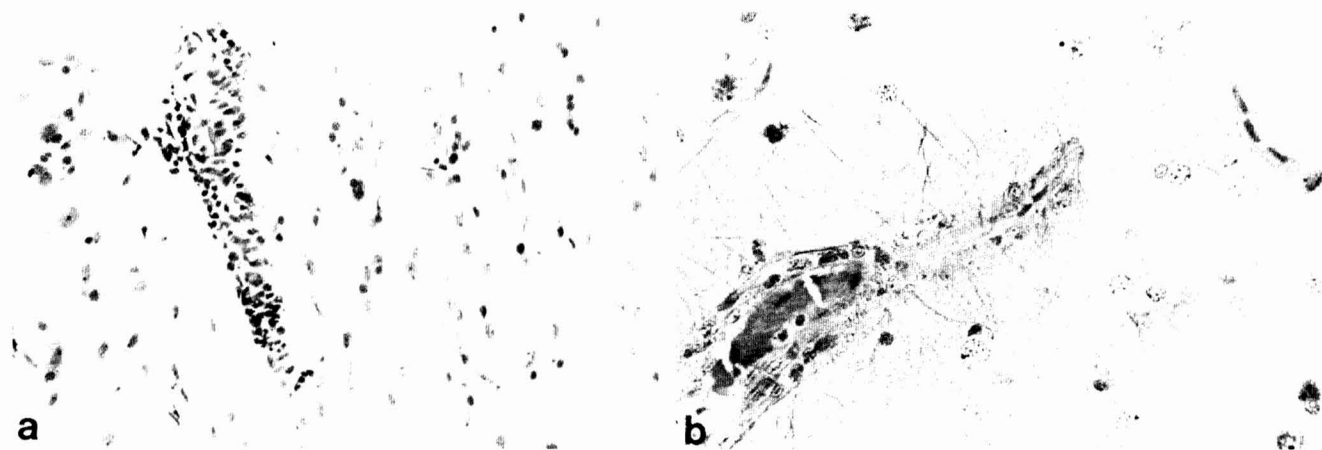


FIG. 2. Photomicrograph of the left frontal cortical biopsy. *a*, perivascular cuffing by mononuclear inflammatory cells and microglial hyperplasia with rod cell formation. (Cresyl violet stain, $\times 180$.) *b*, extensive fibrillary gliosis. (Phosphotungstic acid-hematoxylin stain, $\times 290$.)

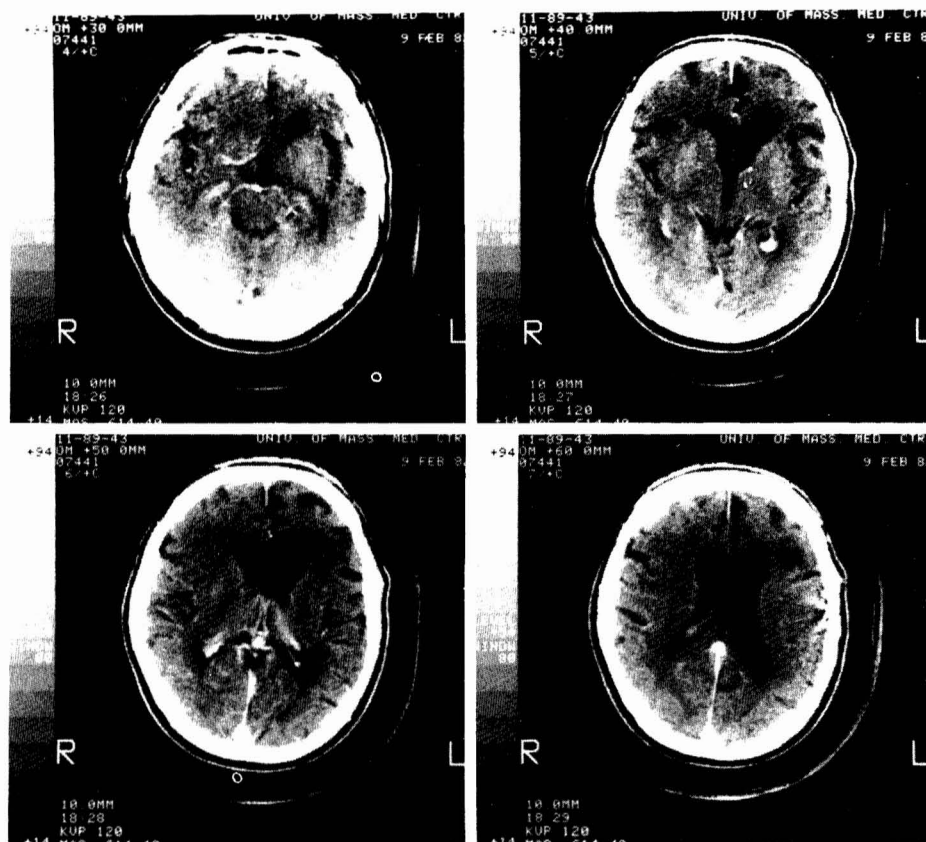
lar syphilis with infarction, a densely enhancing focal lesion in a presumed syphilitic gumma, and both generalized atrophy and bilateral symmetrical areas of white matter hypodensity in presumed general paresis (3, 4). To our knowledge, no one has described the CT appearance of such a widespread hemispheric white matter lesion with mass effect. In addition, neither of the previous reports provides pathological correlation with the CT findings.

The CT-pathological correlation is an intriguing aspect of our case. Initially, the findings of mass effect, white matter edema, and the lack of a discrete mass suggested a low grade infiltrating primary tumor. Subsequently, with the positive serology and the focal neurological abnormalities, meningo-vascular syphilis with infarction was considered, and indeed

the gyral enhancement did suggest a focal ischemic insult. There was, however, no evidence of infarction or vasculitis in the biopsy, and the arteriogram showed neither an occluded major vessel nor the arteritic changes of neurosyphilis (10). The initial CT scan suggested white matter pathway edema, unusual for an infarct (13), and the hypodense area was not confined to one vascular territory. Also, the repeat study seemed more suggestive of a diffuse atrophic process, rather than a discrete infarction. Lastly, the lesions of meningo-vascular syphilis and general paresis are rarely seen concurrently in the same patient (6).

In a rare form of general paresis, Lissauer's dementia paralytica (12), the presentation is epileptic or apoplectic and focal signs occur. Postmortem examinations have demon-

FIG. 3. Four months after therapy, there is ex vacuo dilatation of the frontal horns and left lateral ventricle, as well as the left sylvian fissure and frontal lobe cortical sulci. No discrete parenchymal abnormality other than postoperative change is identified.



strated focal atrophy, often unilateral, involving the temporal and inferior parietal lobes and, to a lesser extent, the frontal and occipital lobes. Microscopic examination of the severely involved cortex has shown neuronal dropout, which varies from pseudolaminar necrosis to frank status spongiosis, and an accompanying loss of myelin in the white matter, which often extends beyond the affected cortical regions. Less severely involved areas have shown the more typical changes of general paresis.

The pathogenesis is unknown, but is thought by Merritt and Springlova to represent a disturbance in the cerebral circulation which, although insufficient to produce infarction, could lead to the microscopic changes described (12). Our patient would best seem to fit this diagnosis clinically and radiographically, particularly when considering the CT scans in sequence. In addition, the gyral enhancement lends support to the notion of an ischemic basis for Lissauer's disease. Although the biopsy findings in our case were compatible with general paresis, the severe changes described in Lissauer's dementia paralytica were not evident. This, however, could be due to both the small size of the biopsy and the focal nature of the abnormalities present in the latter condition.

The serological diagnosis of neurosyphilis at times is difficult. One-third to one-half of the patients will have negative VDRL tests in serum, and as many as one-third will have a nonreactive VDRL in the CSF (1, 5, 7). Treponemal tests, such as the fluorescent treponemal antibody absorption test (FTA-ABS), although more sensitive for syphilis (14), are not specific for neurosyphilis, and the value of the CSF-FTA test remains unproven (2, 9). An increase in CSF lymphocytes and immunoglobulins can support the diagnosis, but these are elevated in a variety of conditions (2). The CT scan abnormalities, although also not specific for neurosyphilis,

can provide additional support for the diagnosis in the appropriate clinical setting. Also, one should consider neurosyphilis in all patients with a focally abnormal examination and atypical localized abnormalities on the CT scan, and one should not exclude that diagnosis until a specific serum treponemal test has been performed.

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