Multiple cerebral gummata

Case report

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The case of a patient with multiple small cerebral gummata presenting with severely raised intracranial pressure is reported. The diagnosis, which was quite unexpected, was based on positive serological tests for syphilis. Computerized tomography (CT) showed small enhancing lesions with intense cerebral edema. The patient was treated successfully with penicillin, and resolution of the lesions was observed on CT scanning over a 1-year period. The importance of “routine” serological testing is noted.

KEY WORDS · cerebral gumma · serology · penicillin · syphilis · neurosyphilis

In the past, 30% to 40% of patients with syphilis have had involvement of the central nervous system. It is probable that the recent resurgence of early infectious syphilis, especially amongst male homosexuals, will produce some increase in cases of late syphilis. Several authors have emphasized that this “fever diluted by time” continues in both classical and atypical forms. This report concerns one of the rarer forms of neurosyphilis.

Case Report

This 32-year-old widow was admitted as an emergency to the Wessex Neurological Centre on the night of November 10, 1978. She gave a 3-week history of increasingly severe headaches associated with vomiting, tinnitus, diplopia, and visual obscurations. In October, 1977, she had developed a pruritic, maculopapular axillary rash, which had resolved spontaneously after 6 weeks only to recur in December, 1977, over a wide area of the trunk and limbs. This eruption was likened to chronic erythema multiforme and a skin biopsy at the time was thought typical of mycosis fungoides. Chlorambucil, 15 mg daily, was prescribed and the lesions disappeared without trace.

Examination. On admission, general examination revealed no abnormality. The patient was alert and oriented, with a mild expressive dysphasia. Her facial expression was taut and hectic; her mood oscillated from truculence to euphoria totally inappropriate to her alarming circumstances; at times she was belligerent and disruptive. She had gross bilateral papilledema, and visual acuity was 6/6 bilaterally with an enlarged blind spot in the visual field of the left eye. The pupils were normal and eye movements were unimpaired. There was a mild pyramidal paresis of the right lower limb. Clinical diagnosis of a left frontal mass was made.

Course. Lumbar puncture was clearly prohibited, so a therapeutic trial of intramuscular aqueous pro-
caine penicillin, 1.2 mega-units daily, was given with dramatic response. Within 3 days the patient lost her tense, wild stare and her mental state assumed an even tenor. After 21 daily doses of penicillin, the only neurological abnormality was bilateral papilledema. Dexamethasone was gradually withdrawn during this period. A CT scan at the end of treatment period showed a small area of enhancement in the anterior corpus callosum; however, the frontotemporal lesion and the cerebral edema had resolved (Fig. 2 left). Lumbar cerebrospinal fluid (CSF) contained 4 lymphocytes/cu mm and a protein level of 350 mg/liter without excessive globulin; testing for TPHA was positive. The original skin biopsy was reviewed by Professor R. O. Weller, who reported extensive vascular cuffing by lymphocytes and plasma cells around superficial and deep blood vessels in the dermis, with some endothelial cell proliferation and a small epithelioid granuloma. The appearance was characteristic of the papular lesion of secondary syphilis.

Follow-Up Examination. A CT scan repeated 6 weeks after the end of treatment showed increased enhancement in the corpus callosum but without mass effect. Eight months later, this abnormality had resolved (Fig. 2 center and right). The patient remains well, apart from a rather uninhibited affect, as illustrated by her parting comment to the house staff in October, 1979: "You lovely boys must come to ... and visit me." The patient had been a widow for 8 years, her husband having died of cirrhosis, and she denied sexual contact since his death.

Discussion

The VDRL titer of 1:128 and transient eruption disappearing without trace are suggestive of secondary cutaneous syphilis. This possibility is supported by the histological findings, although the profound itching and absence of constitutional symptoms are more usual in tertiary syphilis. The positive TPHA test of the CSF indicates that the patient had neurosyphilis, as false-positive reactions to the specific antibody tests applied to CSF are rare. Neurosyphilis is classified as asymptomatic, meningovascular, or parenchymatous; the division between the last two categories is not absolute. Meningeal involvement may occur within 2 years of the primary infection, occasionally coinciding with the secondary rash, and taking the form of acute syphilitic meningitis. In the present case, the absence of fever, meningism, and cranial nerve palsies makes an acute meningitis unlikely. Cerebral gummas are very rare and present as cerebral tumors. Multiple pea-sized meningeal gummata are rarer still; they are surrounded by extensive cerebral edema and ischemic softening consequent upon meningeal arteritis and phlebitis. It is proposed that the patient described here had this latter form of meningovascular syphilis, which responded to penicillin, in contrast with classical cerebral gummata, which do not resolve with penicillin and require excision.

There are few descriptions of the radiological appearances of cerebral gummata. Cerebral angiography shows an avascular mass with narrowing or dilatation of local arteries; occasionally, a hypervascular mass is demonstrated. On CT scanning, an en-
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hancing mass without edema is seen. The CT scan in
the present case showed small enhancing foci with
extensive white-matter edema, and this appearance
complements the pathological description of small
meningeal gummata. Fortuitous circumstances by
way of multiple lesions coupled with a misleading
history of lymphoma delayed surgical intervention.
Routine serological tests proved of value in reaching
the correct diagnosis and appropriate treatment.

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