Neurocysticercosis

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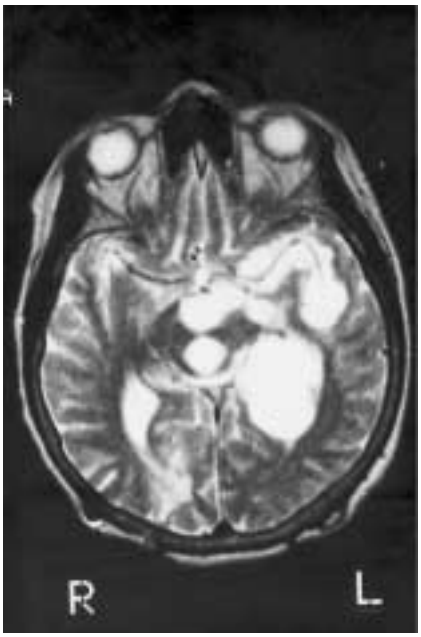
Neurocysticercosis is caused by the larval form of the pork tapeworm, *Taenia solium*. Most cases of neurocysticercosis are due to cysts within the cerebral parenchyma and generally carry a good prognosis. Extraparenchymal neurocysticercosis results from cysts within the cerebrospinal fluid. In contrast to parenchymal disease, its neurologic outcome is poor and it can cause death [1,2]. Chronic inflammation in the subarachnoid space caused by cysticercal cysts mimics chronic meningitis from other etiologies and may lead to erroneous diagnosis and delayed treatment [3]. The specific diagnosis of neurocysticercosis may prove difficult, as history, examination, and laboratory studies may be non-specific.

We present a case of extraparenchymal neurocysticercosis and review the literature regarding diagnosis and management. Based on a survey of the literature, we believe that this is the first case reported in Israel.

Patient Description

A 49 year old woman presented with an 18 month history of headaches, nausea, vomiting, and progressive cognitive decline. She was born in Russia and immigrated to Israel 5 years previously. Her past medical history included pulmonary tuberculosis that was treated twice (the treatment protocols and duration are not known). In the previous year, she was hospitalized repeatedly in several hospitals and underwent thorough examinations. Computed tomography and magnetic resonance imaging scans showed obstructive hydrocephalus and multicystic lesions in the basal cisterns. Cerebrospinal fluid examination showed hypoglycorrachia (32–37 mg/dl), elevated protein (96–125 mg/dl) and lymphocytic pleocytosis (42–67 cells/L). Because of the combination of known past pulmonary tuberculosis and evidence of chronic meningitis, she was diagnosed as suffering from tuberculous meningitis, and empiric anti-tuberculous treatment with isoniazid, rifampicin and pyrazinamid was initiated. She also underwent ventriculo-peritoneal shunt insertion to alleviate the hydrocephalus. Several shunt revisions were carried out due to continued neurologic deterioration, but they were unhelpful. She was admitted to our hospital because of continued cognitive decline. Her neurologic examination revealed lethargy, bilateral optic atrophy, and ataxic gait. Cognitive evaluation showed an apathetic and bradiphrenic patient, with markedly reduced functions such as attention, concentration, abstraction, short and long-term memory, left-right discrimination, anomia, acalculia, and alexia. Mini-mental status examination score was 11/30. Blood tests were normal. The MRI scan was remarkably abnormal and showed many multicystic lesions located in the interpeduncular cistern, the choroidal and transverse fissures, and the left sylvian fissure [Figure]. The lesions had increased in size as compared to those visualized in the previous scans. Studies of the lumbar CSF showed high protein levels (244–404 mg/dl), low glucose levels (5–9 mg/dl) and lymphocytic and eosinophilic pleocytosis (50–163 cells/mm^3). CSF obtained by a ventricular tap was normal. Direct smears, cultures and serologic tests were negative for a variety of bacteria, viruses and fungi. Mycobacteria were not revealed in repeated CSF samples, which were stained with Ziehl-Nielsen and cultured. The Gene-probe Amplified Mycobacteria Tuberculosis Direct test was also performed. The patient became severely lethargic de-



MRI of the brain, axial section, T2-weighted image. Polycyclic, multicystic lesions in the basal cisterns and the left sylvian fissure.

CSF = cerebrospinal fluid

spite continued anti-tuberculous treatment and third ventriculostomy due to obstructive hydrocephalus.

A careful history revealed possible exposure to *Taenia solium*. The diagnosis of neurocysticercosis was then suspected. She underwent left temporoparietal craniectomy and a biopsy specimen was taken from one of the cystic lesions. The histologic examination showed a cysticercal cyst. The patient was treated with praziquantel and steroids, which led to an early improvement followed by stabilization of her condition (6 months of follow-up). Improvement was also noted in CT imaging.

Comments

We describe a case of extraparenchymal neurocysticercosis that presented with headaches, nausea, vomiting, and progressive cognitive decline. This disease is endemic in many areas worldwide, but has never been reported in Israel.

Four types of neurocysticercosis are known [1]. The most frequent clinical form is parenchymal cysticercosis, which carries a good prognosis. Parenchymal cysts are usually found in the cerebral cortex, including the cortical-subcortical junction. Meningeal cysts are frequent in the meninges overlying the base of the brain. Ventricular cysts are usually located in the fourth ventricle and can cause hydrocephalus. Spinal cord cysts also exist but are rare. The extraparenchymal form, as in our patient, carries a much poorer prognosis and may even be fatal [2]. As occurred in our patient, extraparenchymal cysticercosis is frequently complicated by hydrocephalus, increased intracranial pressure, optic atrophy and arachnoiditis [2]. Entrapment of the circle of Willis, which may cause infarctions, was also reported. Fever, meningeal irritation and cranial neuropathies are rare [3]. Impairment of intellectual function may be significant [3]. Reported symptoms include apathy, amnesia, emotional lability, and hallucinations [3]. Our patient also presented with marked intellectual impairment; she became apathetic, bradiphrenic, and had severe memory and attention deficits.

The diagnosis of neurocysticercosis was deferred in this patient because of a tentative diagnosis of tuberculous meningitis. The clinical history (including previous history of tuberculosis), neurologic examination and laboratory tests (particularly the CSF findings) could be compatible with tuberculous meningitis [4]. MRI findings of CNS tuberculosis include intraparenchymal tuberculomas, thickening and enhancement of meninges especially in basilar regions, exudates in the basal cisterns and in the sylvian fissure, gyral enhancement, and infarcts (due to blood vessel entrapment) [4]. Hydrocephalus is a known complication of CNS tuberculosis [4]. While the MRI findings in this patient could be related to CNS tuberculosis, the multicystic lesions were more compatible with extraparenchymal neurocysticercosis. The diagnosis of neurocysticercosis was also suspected due to continued clinical deterioration and CSF abnormalities despite prolonged anti-tuberculous treatment, and lack of detection of *Mycobacterium tuberculosis* in repeated CSF samples. CSF eosinophilia, as found in our patient, is characteristic of parasitic diseases (including cysticercosis), fungal infections, neoplastic diseases, sarcoidosis, and hypereosinophilic syndromes. In this patient, CSF eosinophilia was suggestive of neurocysticercosis [3]. Differences between ventricular and lumbar CSF, as seen in our case, were also described in other patients with chronic arachnoiditis caused by cysticercosis [5]. Exposure history is essential in evaluating the potential for neurocysticercosis, as shown in this case. The diagnosis was finally achieved by brain biopsy.

The anti-helminthics praziquantel and albendazole are accepted as effective treatment in patients with neurocysticercosis [2,3]. Addition of corticosteroids may be required to prevent initial exacerbation of neurologic symptoms [3]. Because most studies of medical therapy focus on parenchymal neurocysticercosis, knowledge about the therapy for extraparenchymal disease is still lacking. Patients with arachnoiditis may not respond well to anti-parasitic therapy [2,3]. Surgical intervention and multiple courses of anti-parasitic drugs are usually necessary. Many of the deaths due to subarachnoid neurocysticercosis were associated with hydrocephalus [2,3]. Thus, diversion procedures, as were done in this case, are critical. Craniotomy and excision of subarachnoid cysts have also been advocated, but given the high morbidity and mortality, medical treatment is recommended before considering surgical therapy [2].

This case illustrates the need for increased awareness to neurocysticercosis in Israel, as emigration from or travel to endemic areas can result in exposure to *Taenia solium*. The specific diagnosis of neurocysticercosis may be difficult, and in some cases brain biopsy may be necessary to establish the diagnosis of this potentially treatable neurologic disease.

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