Intracranial sparganosis: an uncommon infection

Case report

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The first case of intracranial sparganosis to be reported from the United States is presented. The patient, a 27-year-old woman, complained of focal seizures involving the right lower extremity. A left parietal parasagittal craniotomy was performed, and a granuloma containing a sparganum was excised from the parietal lobe. The clinical and pathological features of sparganosis are reviewed. Only five cases of intracranial sparganosis have previously been described.

KEY WORDS • sparganum • Spirometra • parasite • intracranial granuloma

SPARGANOSIS is an infection caused by a sparganum, the migrating plerocercoid larva of a tapeworm of the genus Spirometra. Human sparganosis has been reported worldwide, but is most common in China, Japan, and Southeast Asia. Approximately 65 cases have been described in the United States, especially in the southeastern states. The majority of infections involve the subcutaneous tissues. Rarely, however, the organism may invade the central nervous system. This report concerns the first case of intracranial sparganosis to be reported from the United States.

Case Report

This 27-year-old woman, who worked as a health educator, complained of seizures. Her present illness dated from February 23, 1982, when she became aware of an abnormal pulsating sensation in her right lower extremity, and some associated difficulty in the use of the limb. The acute episode lasted approximately 4 minutes, with delayed clearing of abnormal sensations and "weakness" in about 15 minutes. A second similar but more intense episode occurred 4 days later, accompanied by a grand mal seizure and followed by amnesia and a sore tongue. In the following week the patient had two additional brief episodes of focal impairment of extremity sensation. The only significant feature in an otherwise unremarkable history was a complaint of headaches for several years. Her father has Parkinson’s disease.

She was born in Greece and, until she was 9 years old, lived there in a rural setting in the presence of common farm animals (sheep, pigs, dogs, and cats). She subsequently lived in Los Angeles, but each summer following her emigration to the United States returned to Greece for prolonged visits, again in the rural setting. Her only medication was phenobarbital for seizure control. She was unaware of any contact with tuberculosis.

Examination. A general physical examination revealed no abnormalities. She was a bright intelligent woman with normal mentation. Cranial examination and cranial nerve survey were unremarkable. There was no speech disorder. Motor development, tone, power, coordination, gait, and stance were normal. Sensory examination was unrevealing. Abdominal reflexes were absent, and either a neutral or equivocal plantar response was regularly elicited on the right side.

High sections of the computerized tomography (CT) scan with a double dose of contrast material revealed an enhancing nodule in the upper left parasagittal parietal region (Fig. 1 left). Examination of the lumbar cerebrospinal fluid disclosed no cells and a protein concentration of 24 mg%. Cerebral angiography was not abnormal, but a technetium isotope scan demonstrated uptake in the region of the parietal nodule.
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**Fig. 1.** Computerized tomography scans with contrast enhancement. Left: The scan in March, 1982, shows an enhancing lesion in the left parasagittal parietal cortex. Right: The scan in November, 1982, reveals enlargement of the lesion with a surrounding low-density region suggestive of cyst formation.

Routine hematological and chemical investigations were normal. The patient's purified protein derivative was strongly positive and the possibility of tuberculosis was entertained. A chest roentgenogram and excretory urogram revealed no lesions. A second CT scan 6 weeks following the first study was unchanged.

**First Operation.** Isoniazid, rifampin, and phenobarbital were given and a left parietal parasagittal craniotomy was performed. Firm, tough tissue was localized with the aid of ultrasonography, and biopsies of the parietal lobe were obtained. The pathological examination revealed chronic inflammation with eosinophils. A parasitic process was suggested, but the precise etiology could not be established. Viral, fungal, bacterial, and mycobacterial cultures from the biopsy specimens were all negative. A transient eosinophilia to 13% was noted postoperatively. Serological investigations for *Toxoplasma*, cytomegalovirus, *Echinococcus*, and *Cysticercus* were all negative.

Postoperatively, there was no change in the patient's neurological status. Follow-up CT scans in May and July, 1982, demonstrated the lesion to be slightly smaller. She was maintained on rifampin, isoniazid, and phenobarbital. Two months postoperatively, she suffered a series of focal seizures, each associated with a sense of numbness in the right lower extremity. She also described recurrent episodes of transient disorientation or "depersonalization" in which she believed she had an altered relationship to space. A transient splinter hemorrhage was observed in the right fundus in September, 1982.

By the 3rd week of October, 1982, a CT scan demonstrated that the lesion in the high left parietal region had increased in size. There was an interictal background sense of numbness in the right lower extremity, although no functional deficit was observed. In mid-November, 1982, the patient experienced a series of sensory seizures which, for the first time, were associated with weakness in the extremity. She was hospitalized and a CT scan on November 18 revealed that the parietal lesion was even larger than in October (Fig. 1 right). A low attenuation area was suggestive of cyst formation.

**Second Operation.** On November 22, 1982, the left parasagittal parietal region was reexplored. At a depth of approximately 1 mm, a glistening cyst inside the parietal lobe was emptied of approximately 10 ml of non-clotting yellow fluid. In the wall of the cyst, and abutting the falk and the longitudinal sinus, a solid nodular tumor was defined. It was not attached to the dura mater, but was embedded in the brain parenchyma and surrounded by a thin shroud of attenuated brain tissue. Several vascular channels were associated with it. The lesion was totally delivered with ease and removed. The patient has remained well in the 15 months following the second operative procedure and has no neurological deficits.

**Pathological Examination.** The tissue from the first craniotomy consisted of six small biopsy fragments obtained from the parietal lobe cortex. All of these revealed low-grade reactive gliosis and a moderate perivascular infiltrate of chronic reactive cells consisting of lymphocytes, plasma cells, and eosinophils. No organisms, viral inclusions, or neoplasia were noted.

The mass obtained from the second craniotomy was an irregularly shaped yellow-and-tan nodule measuring 1.5 × 1.5 × 1.0 cm. The tissue was firm and sharply delineated from a thin rim of soft hyperemic cerebral cortex (Fig. 2). Gram and acid-fast stains revealed no
organisms. Because of the nature of the cellular infiltrate, step sections were carried out and a single, small but solid, folded worm was microscopically identified (Fig. 3). Underlying a regular integument within the organism were columnar ("tegumental") cells, bundles of longitudinal smooth-muscle fibers, and longitudinally arranged excretory ducts in a predominantly fluid-filled parenchyma. A few calcareous corpuscles were noted, but a scolex was not present. The parasite was surrounded by an infiltrate of polymorphic neutrophils, with a concentric collection of plasma cells, lymphocytes, histiocytes, and eosinophils in a dense connective tissue matrix. No epithelioid macrophages or giant cells were encountered. The surrounding margin of brain was edematous, congested, and gliotic, and housed numerous chronic inflammatory cells.

Discussion

Sparganosis was first described in China by Manson in 1882. Since then, details of over 300 cases have been published. In 1908, Stiles reported the first case in man (Sparganum proliferum) in the United States. Subsequently, approximately 65 cases have been described in this country, the majority occurring in the southeastern states. Only one case has previously been identified in the state of California; this was found in a Filipino who presumably acquired the disease before immigrating.

Two distinct forms of sparganosis exist. The classical type, representing the vast majority of cases, is nonproliferative. The larvae are capable of slow growth in man, but cannot reproduce. This entity is caused by the larvae of tapeworms of the genus Spirometra, which are closely related to the fish tapeworm Diphyllobothrium latum. In contrast, Sparganum proliferum is a rare disease in which the larva grows by branching and budding, and is capable of forming new organisms. Although this parasite has the histological components of a typical sparganum, it is irregular and asymmetrical. An adult stage of this parasite is unknown. Approximately eight cases of infection with Sparganum proliferum have been recognized, the majority occurring in Japan. This aberrant larva usually causes death by massive proliferation throughout virtually all host tissues, and may involve the brain.

The life cycle of Spirometra has been well documented. Definitive hosts are domestic and wild cats and dogs. As with most tapeworms, eggs are passed in the feces from the intestinal parasite. These eggs...
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<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors, Year</th>
<th>Sex, Age (yrs)</th>
<th>Location</th>
<th>Signs &amp; Symptoms</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Takeuchi, 1918*</td>
<td>M, 33</td>
<td>intracerebral (rt frontal)</td>
<td>seizures, lt hemiparesis, headache</td>
<td>death due to cerebral infarction; autopsy</td>
</tr>
<tr>
<td>2</td>
<td>Kuroiwa, 1951</td>
<td>M, 45</td>
<td>intracerebral (rt temporal)</td>
<td>fever, depressed level of consciousness, headache, diplopia, poor vision, weakness, verticerebral hemiparesis, vomiting, rt hemiparesis</td>
<td>death due to subarachnoid hemorrhage; autopsy</td>
</tr>
<tr>
<td>3</td>
<td>Pradatsundarasar, et al., 1971</td>
<td>F, 46</td>
<td>subarachnoid (basal cisterns)</td>
<td>seizures</td>
<td>death due to subarachnoid hemorrhage; autopsy</td>
</tr>
<tr>
<td>4</td>
<td>Monolo, et al., 1976</td>
<td>M, 28</td>
<td>subarachnoid (lt frontal)</td>
<td>seizures, surgery, well postop</td>
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<tr>
<td>5</td>
<td>Mineura &amp; Mori, 1980</td>
<td>F, 33</td>
<td>subdural (rt frontal)</td>
<td>surgeries, well postop</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Anders, et al., 1984</td>
<td>F, 27</td>
<td>intracerebral (lt parietal parasagittal)</td>
<td>surgeries, well postop</td>
<td></td>
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</tbody>
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* Cited by Kuroiwa.18

**TABLE 1**

Summary of six cases of intracranial sparganosis

Man is an accidental intermediate host, but can acquire the infection in three ways: 1) by drinking water contaminated with infected copepods (apparently the most common route of infection in the United States5-20); 2) by ingestion of raw or inadequately cooked meat infected with the sparganum; and 3) by the application of the flesh of an infected intermediate host as a poultice to an open wound into which the sparganum can then enter. This latter method has been implicated as causing orbital sparganosis in Korea. Although the larvae penetrate the human intestine and migrate to various locations, they cannot develop into adult worms. The most likely source of infection in the present patient was by drinking contaminated well water while visiting her parents' farm in Greece.

The diagnosis of sparganosis requires gross or microscopic morphological confirmation.25,26 Lesions are usually well circumscribed, small, firm, fibrous masses that are gray, brown, or yellow in color. Foci of hemorrhage, necrosis, and cyst formation are often present, and occasionally the sparganum can be grossly identified. Spargana are thin white solid ribbon-like worms which may measure variably from several millimeters to over a meter in length. Width is variable, but is usually only a few millimeters. The anterior end is invaginated and slightly bulbous. The body is ridged at fairly regular intervals. Microscopically, the larva is surrounded by a dense inflammatory response containing lymphocytes, plasma cells, histiocytes, neutrophils, and occasionally foreign-body giant cells. Eosinophils are present in variable numbers and Charcot-Leyden crystals may be seen. The sparganum's body wall consists of a ridged regular integument, 5 to 15 μ thick. Important characteristics that distinguish the spargana from the Cysticercus, Echinococcus, and Coenurus larvae are the presence of a solid noncavitated body, lack of bladder walls, and absence of a hooked scolex.

The majority of patients with sparganosis are adults. Patients develop slowly growing, tender, sometimes migratory subcutaneous nodules which may persist for several weeks to years.20,27 Fever, chills, erythema, edema, and eosinophilia may be present. The eosinophilia may be especially apparent when the worm is migrating through tissue.2 Symptoms are predominantly related to those caused by a space-occupying lesion. Although the vast majority of infections occur subcutaneously, the parasite has also been described in the eye,13 lung, epididymis, urethra, and vertebral canal.11,21 Intracranial involvement is very rare. Only five cases have been previously described10,15,16,22 (Table 1). The patients have ranged in age from 27 to 46 years, with equal numbers of men and women reported. The most frequent early symptoms have been headache and seizures, with or without focal neurological signs. The course may be quite acute, mimicking other causes of encephalitis (Case 2) or subarachnoid hemorrhage (Case 3). More commonly, however, patients present with an illness lasting from months to years. In all probability, in intracranial cases the parasite may be anywhere, but the majority have been intracerebral, subarachnoid, or subdural in location.

Three cases have been treated surgically with good results (Table 1). As there is no effective medical therapy and since the illness may terminate fatally, appropriate surgery is indicated.

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References


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