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The Brief Case: Central Nervous System Sparganosis in a 53-Year-Old Thai Man

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CASE

A 53-year-old Thai man with a history of pemphigus vulgaris on chronic prednisolone (30 mg/day) presented to a hospital in Thailand with a 5-month history of lower back pain. He had initially been treated with tramadol, amitriptyline, and gabapentin without relief. Two months prior to presentation, he had developed weakness of the right leg, and he presented when weakness in his right foot made it difficult for him to keep his sandal on. He denied numbness, paresthesia, urinary retention, or bowel incontinence. On physical exam, he appeared well, with a temperature of 36.6°C, a blood pressure of 141/72 mm Hg, and a pulse of 74 beats per minute. Neurologic exam showed a knee flexion score of 4/5 and a foot dorsiflexion score of 3/5 on the right and a knee flexion score of 4/5 and foot dorsiflexion on the left. Sensation was intact bilaterally. Patellar reflexes were 1+ bilaterally, with downward plantar reflexes. Initial laboratory investigation showed a white blood cell count of 11,000 cells/mm^3 (normal, 4,000 to 11,000 cells/mm^3), with 81.1% neutrophils and 0.3% eosinophils (absolute eosinophil count, 33 cells/mm^3). Three serial stool specimens sent for microscopic ova and parasite identification were negative.

Gadolinium-enhanced magnetic resonance imaging (MRI) of the lumbosacral spine was performed (Fig. 1A), demonstrating arachnoiditis with a nonenhancing, loculated cystic lesion attached to the left aspect of the cauda equina. Based on this appearance, a parasitic infection was suspected, and neurosurgical consultation was requested. The patient was taken to the operating room for removal of the structure. The cystic lesion was identified in the intradural space, and within was found a macroscopic white helminth (Fig. 1B). The exact length of the specimen could not be determined due to fragmentation during extraction but was greater than 3 cm. Gross pathology showed a helminth with pseudosegmentation evidenced by various circumferences, while microscopic specimens demonstrated a tegumental brush border, calcareous bodies, and a lack of organoid structures (Fig. 1C). With the combination of clinical presentation and pathological findings, a diagnosis of sparganosis was made. Subsequent MRI of the brain also showed evidence of cerebral and cerebellar involvement, with white matter enhancement and serpiginous tunneling (Fig. 1D). On further history, the patient acknowledged that he frequently consumed both raw frog and raw snake meat. He again denied any other neurologic symptoms apart from those mentioned previously, and there was no evidence of cerebellar or cerebral dysfunction on exam.

DISCUSSION

Sparganosis is a zoonotic infection caused by cestodes of the genera *Spirometra* and *Sparganum*, members of the Diphyllobothriidae family (1; DPDx, sparganosis [Centers

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For answers to the self-assessment questions and take-home points, see page 658 in this issue (https://doi.org/10.1128/JCM.01337-16).

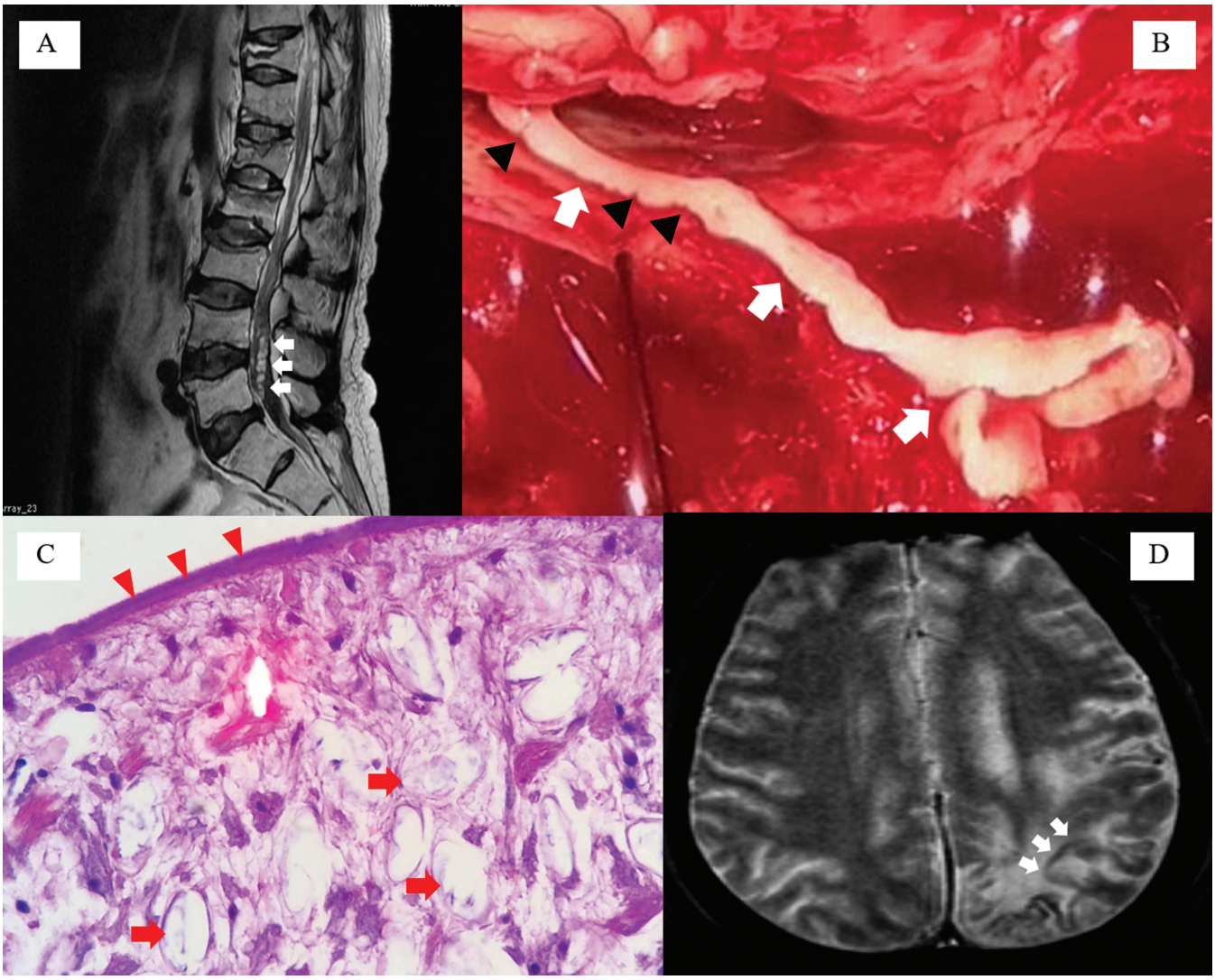

FIG 1 (A) Magnetic resonance imaging of the lumbosacral spine demonstrates a loculated, cystic lesion (arrows) attached to the left cauda equina. (B) An intraoperative photograph shows a white, elongated plerocercoid (white arrows) being removed from the cyst. Note the variation in circumferences, indicating the presence of pseudosegmentation (black arrowheads). (C) Tissue sections of the plerocercoid demonstrate faintly purple-staining fragments of calcareous bodies (arrows), a lack of organoid structures, and the tegumental fine brush border (arrowheads) characteristic of cestodes (hematoxylin and eosin stain; ×400 magnification). (D) T2-weighted gradient echo sequence magnetic resonance imaging of the brain shows white matter edema of the left temporoparietal and occipital regions with tunneling (arrows) indicative of parasitic migration through the tissue.

for Disease Control and Prevention, http://www.cdc.gov/dpdx/sparganosis/]). Cases have occurred worldwide, though the majority of reports come from East and Southeast Asia, where most cases are caused by *Spirometra erinaceieuropaei*. *Sparganum proliferum* is the cause of the rarer proliferative form of the disease. Adult worms reside in the intestines of multiple mammalian species, including dogs and cats. Unembryonated eggs are shed in the feces of infected animals and have an elliptical shape measuring approximately 65 by 35 μm. The eggs hatch in water, releasing coracidia that are subsequently ingested by copepods, a subclass of crustaceans that are the first intermediate hosts. There, the parasites develop into procercoid larvae. When the copepod is eaten by a second intermediate host (often fish, reptiles, or amphibians), the procercoid larvae are released and develop into plerocercoid larvae (termed sparagana). Humans act as accidental hosts and are infected by the procercoid or plerocercoid larval forms, which cause illness by migrating into tissues. Humans are almost always intermediate hosts, only rarely developing intraintestinal adult worms (1, 2). Transmission is most commonly from ingestion of raw or undercooked frog or snake

meat or contaminated water, and human-to-human transmission is not seen. Historically, infection has also been seen from the use of traditional poultices involving raw meat (especially raw frog) applied to inflamed eyes, skin, and teeth (2, 3). Transmission via this route has decreased substantially over the last decade, however, likely due to a decline in these practices.

The clinical presentations of sparganosis are widely varied and dependent upon the location of infection. Larvae have been identified throughout the body, including in the brain, spine, eyes, skin, lungs, abdominal viscera, and genitourinary tract, and can live in humans for up to 20 years (2). Central nervous system (CNS) infection can present with headache, seizure, confusion, weakness, and/or paresthesia, depending on the location of the larvae and migratory path (4). Cerebral hemorrhage has also been described (5). Spinal disease symptoms can include back pain and weakness, as in our patient, and occasionally in urinary retention or bowel incontinence, depending on the size and location of the lesion (6).

Computed tomography (CT) and MRI imaging can assist in diagnosis of sparganosis. In brain MRI, white matter enhancement and serpiginous tunneling are characteristic, and sequential imaging may demonstrate migratory lesions (1). In spinal disease, nodular mass lesions with minimal enhancement are seen upon MRI (6). Based on the history, imaging, and local epidemiology, our differential for this case included neurocysticercosis, echinococcosis, and gnathostomiasis. Neurocysticercosis often presents with multiple cystic lesions, though single lesions can be seen. Spinal echinococcosis is more likely to involve the vertebral body, while gnathostomiasis lesions are less well circumscribed. Other infectious mass lesions of the spine, such as tuberculosis and bacterial abscess, more commonly show enhancement of the lesion after the administration of contrast, which was not observed in our patient.

Diagnosis is most often made by identification of the sparganum after extraction or upon tissue pathology. Gross pathology will demonstrate a white, pseudosegmented, ribbon-like helminth resembling an adult cestode, approximately 1 to 2 mm wide and varying from 3 to 30 cm long. The length and pseudosegmentation help distinguish *Spirometra* from other cestodes. Microscopy of the organism shows a tegumental brush border for nutrient absorption, calcareous bodies, and a lack of a gastrointestinal tract. These features are common to all cestodes and are diagnostically useful in differentiating them from other helminths. Pathology generally allows only a genus-level identification of *Spirometra*; species-level identification is not routinely available but can be achieved by PCR, which targets primarily ribosomal gene sequences (1). Enzyme-linked immunosorbent assays (ELISAs) are also available, though cross-reactivity with clonorchiasis, cysticercosis, paragonimiasis, and other helminth antigens can occur depending upon the assay used (1). Both PCR and ELISAs for clinical purposes are unavailable outside Asia. Imaging can aid in the diagnosis of CNS sparganosis, which may demonstrate cystic lesions or tunneling from migration of the larvae, along with associated edema (4).

The majority of cases are treated with surgical excision of the larvae. The entire organism must be removed, as retained scolex may lead to recurrence of disease (1). For cases where excision is not possible, some have been treated with localized injection of 40% ethanol procaine or antichymotrypsin. High-dose praziquantel and mebendazole have been used in certain cases with limited success. Albendazole has also been studied in the mouse model but does not appear to be effective.

After surgical removal of the parasite, our patient was treated with a 2-day course of high-dose praziquantel. At the 12-month follow-up, he had regained complete neurologic function, with only mild residual back pain. Though uncommon, sparganosis should be considered in the differential for patients traveling from Southeast Asia who present with cystic lesions of the CNS and a history of exposure to exotic animals. A thorough assessment of food and water exposures, particularly frog and snake meat, can assist in diagnosis.

SELF-ASSESSMENT QUESTIONS

1. Humans usually acquire sparganosis from ingestion of or exposure to raw or undercooked meat from which animals?
    A. Pigs and cows
    B. Dogs and cats
    C. Frogs and snakes
    D. Sheep and goats
2. Which life cycle stage of the *Spirometra* spp. is infectious to humans?
    A. Unembryonated eggs
    B. Procercoid larvae
    C. Embryonated eggs
    D. Coracidia
3. How is species-level identification of *Spirometra* spp. achieved?
    A. Examination of gross and microscopic pathology
    B. Serologic assays
    C. Magnetic resonance imaging
    D. PCR

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