84

A 64-Year-Old Japanese Man With Generalized Tonic-Clonic Seizures

MASAHIDE YOSHIKAWA, FUMIHIKO NISHIMURA, YUKITERU OUJI AND HIROYUKI NAKASE

Clinical Presentation

History

A 64-year-old Japanese man is admitted to a local hospital in Japan because of generalized tonic-clonic seizures followed by weakness of the right lower extremity.

He has a medical history of hypertension and right cerebellar infarction; however, recent simple partial seizures were well controlled by administration of an antiepileptic agent.

Clinical Findings

On admission, the patient is afebrile, GCS is 15/15 and the neurological examination does not reveal any abnormality.

Laboratory Results

Laboratory results show no abnormalities, including no eosinophilia or leukocytosis.

Imaging

Head CT reveals a small high-density nodule with an enhanced perifocal low-density area in the left occipital lobe.

MRI shows a ring-enhancing, tunnel-shaped lesion in the left occipital lobe (Fig. 84.1).

Angiography findings are normal, except for right vertebral artery occlusion, reflecting the history of right cerebellar infarction.

Further Investigations

Open surgery with craniotomy targeting the lesion is performed. With manipulation of the aspirator and forceps, a white tape-like body at the centre of the targeted lesion is exposed, and complete removal of a live worm-like structure is carefully performed (Fig. 84.2A-C). A presumed diagnosis of sparganosis is made.

Questions

1. What is the cause of sparganosis?
2. What is the treatment of choice for cerebral sparganosis?

Discussion

A 64-year old Japanese man presents with a generalized tonic-clonic seizure. Imaging reveals an elongated structure in the right occipital lobe. During neurosurgery, a live worm-like structure is removed from the brain, identified as a sparganum.

Answer to Question 1

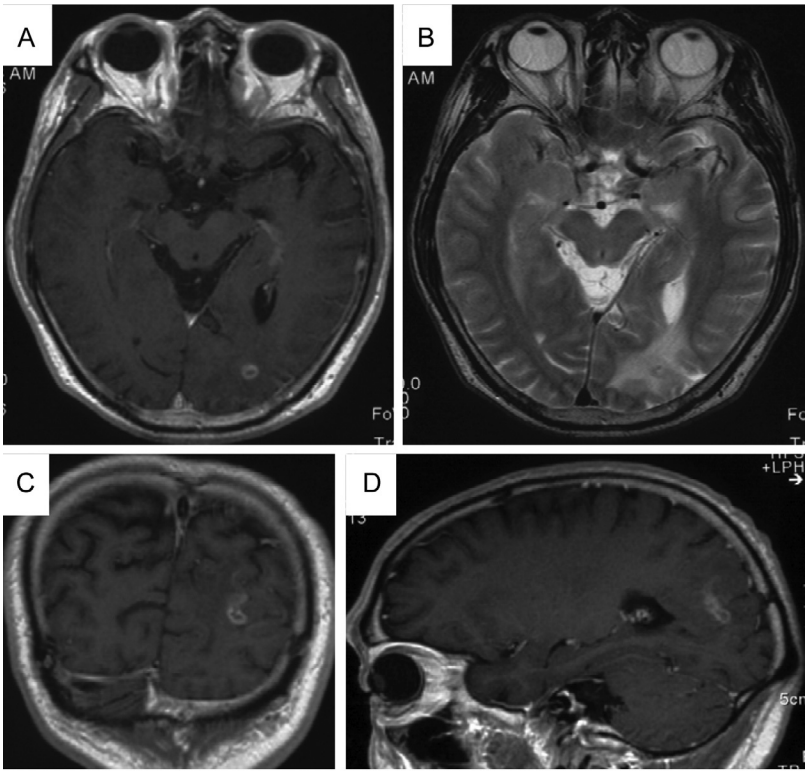
What is the Cause of Sparganosis?

Spargana are the larvae of zoonotic tapeworms of the genus *Spirometra.* Adult worms inhabit the intestines of cats and dogs, and produce eggs that are discharged in faeces. In fresh water, coracidia hatch from the eggs to be ingested by copepods and develop into first stage (procercoid) larvae.

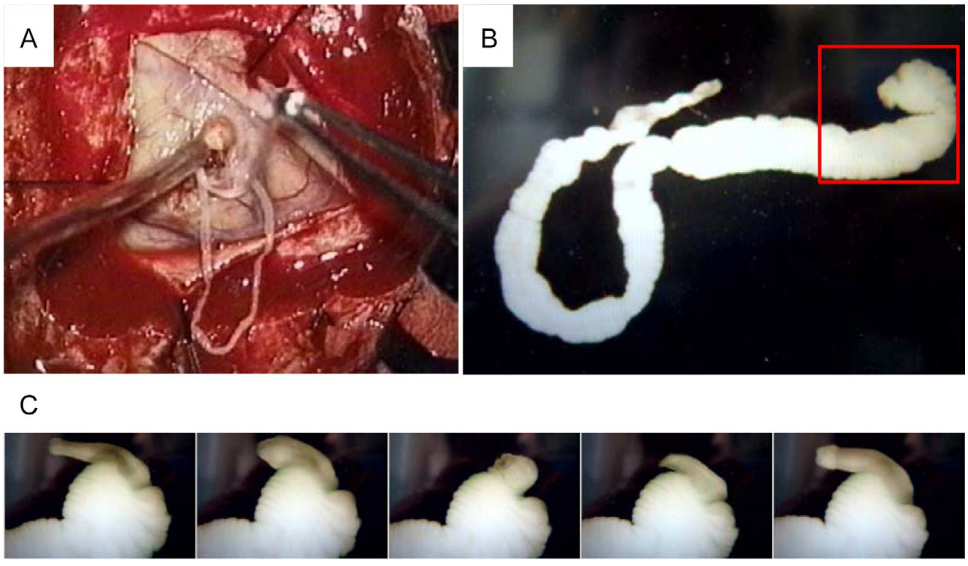
When copepods are ingested by the second intermediate host (e.g. frogs, fish, snakes, birds), the procercoid larvae mature into plerocercoid sparganum larvae and migrate to the muscle and subcutaneous tissue. The cycle is completed when a carnivorous mammal eats the second intermediate host.

Humans can become infected by ingesting the first or second intermediate host, i.e. by drinking water containing infected copepods, eating undercooked meat from infected snakes, frogs, or birds, or by the use of poultices produced from infested frog or snake flesh or skin on open wounds or eyes.

In the human body, the sparganum commonly migrates into subcutaneous tissue or muscle, and sometimes into the eye, the spinal cord or brain, the latter resulting in cerebral sparganosis. The infection route in the present case was probably through consumption of raw chicken meat, which the patient ate approximately once a month before the generalized seizure.



• **Fig. 84.1** MRI A. T1-weighted MRI showing a ring-shaped nodule with gadolinium enhancement in the left occipital lobe. B. T2-weighted image showing an area of hyperintensity associated with an adjacent ventricular dilation. C, D. Post-contrast coronal and sagittal T1-weighted images demonstrating a tunnel-shaped enhancement, so-called tunnel sign.



• **Fig. 84.2** Images of the Sparganum A. Intraoperative photograph showing a live sparganum during removal from the cortex. B. Whole body of extracted sparganum with anterior end enclosed by red square. The sparganum was wrinkled and whitish, and measured 1 to 3 mm in width and up to 12.5 cm in length at maximum extension. C. Five successive images from video recording. The anterior end showed active repeated movements of extension and constriction.

Cerebral sparganosis is a rare disease, but should be considered in patients from endemic areas presenting with symptoms suggestive of a space-occupying lesion. Cases have been mainly reported from Asian countries, especially China, South Korea, Thailand and Japan, and the most commonly observed clinical manifestation is epileptic seizures. Diagnosis is based upon dependable history taking and serological investigations. Neuroimaging, especially MRI, is helpful

for the diagnosis of cerebral sparganosis. The most characteristic finding is the "tunnel sign" in post-contrast MRI, which appears as hypointensity in T1-weighted and hyperintensity in T2-weighted images.

Answer to Question 2

What is the Treatment of Choice for Cerebral Sparganosis?

The treatment of choice is the removal of the sparganum from the brain. However, in a small-sample non-randomized retrospective study, high-dose praziquantel had an efficacy similar to that of surgical removal with respect to the primary outcome (complete disappearance of active lesions in cerebral MRI findings).

The Case Continued…

After surgical removal of the worm, the patient was well without seizures at the most recent examination. Enzyme-linked immunosorbent assay (ELISA) results of serum and cerebrospinal fluid revealed strong positivity for Spirometra mansoni.

SUMMARY BOX

Cerebral Sparganosis

Cerebral sparganosis is the infestation of the human brain by larvae of a zoonotic cestode of carnivorous mammals. It is a food-borne zoonosis acquired by drinking untreated freshwater or eating undercooked meat of snakes and frogs or by application of infested flesh on open wounds or the eyes during medical procedures. The incubation period is 6 to 11 days but may be longer. The larvae can survive in the human host for up to 20 years.

Most cases occur in Asia, but sporadic cases have been reported from many other parts of the world.

Cerebral infestation may manifest like any space-occupying lesion with headache, confusion, seizures, paraesthesias or palsies. Spinal sparganosis may lead to paraplegia, as well as urinary and bowel incontinence.

Diagnosis of cerebral sparganosis is made on the basis of a combination of clinical history, laboratory tests (eosinophilia, positive serology), CT and MRI, and histopathology findings including the identification of the larva itself, if available. Recent advances in imaging technology, especially MRI, are key for diagnosis.

Differential diagnoses include brain tumours or metastases, neurocysticercosis, cerebral schistosomiasis and paragonimiasis.

An HIV test should be done in any person with unclear CNS lesions. In immunocompromised patients, cerebral toxoplasmosis has to be considered, as well as cerebral cryptococcomas and tuberculomas.

Further Reading

1. Baily G, Garcia HH. Other cestode infections: intestinal cestodes, cysticercosis, other larval cestode infections. In: Farrar J, editor. Manson's Tropical Diseases. 23rd ed. London: Elsevier; 2013 [chapter 57].
2. Liu Q, Li MW, Wang ZD, et al. Human sparganosis, a neglected food borne zoonosis. Lancet Infect Dis 2015;15(10):1226–35.
3. Song T, Wang WS, Zhou BR, et al. CT and MR characteristics of cerebral sparganosis. Am J Neuroradiol 2007;28(9):1700–5.
4. Li YX, Ramsahye H, Yin B, et al. Migration: A Notable Feature of Cerebral Sparganosis on Follow-Up MR Imaging. Am J Neuroradiol 2013;34(2):327–33.
5. Hong D, Xie H, Wan H, et al. Efficacy comparison between long-term high-dose praziquantel and surgical therapy for cerebral sparganosis: a multicenter retrospective cohort study. PLoS Negl Trop Dis 2018;12(10):e0006918.