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Case Report

Cerebral Sparganosis

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Abstract

A 47-year-old woman presented with severe headache associated with nausea for 2 weeks. There was no fever, neck pain, dizziness, vertigo, unilateral limb weakness, blurred vision or trauma history. She had no history of systemic disease or medical problems. Brain computed tomography showed left side diffuse brain edema. Brain contrast magnetic resonance imaging revealed a mixed intensity mass lesion with heterogeneous abnormal enhancement in the left parietal lobe. The differential diagnosis included neoplasm and a lesion mimicking a neoplasm. An operative biopsy showed a grossly yellow, 3 cm, left occipital tumor. Pathological findings showed granulomatous inflammation in the brain tissue and a dead parasite body was noted. Calcareous bodies were evident inside the parasite. Cerebral sparganosis, which is an infection from *Spirometra mansoni*, was diagnosed. Two weeks later, she was discharged and had recovered completely. She may have been infected by eating raw meat and drinking unboiled water. Cerebral sparganosis is extremely rare and should be considered in the differential diagnosis of metastatic brain tumor, especially in endemic areas. (*Tzu Chi Med J* 2010;22(2):115–118)

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1. Introduction

Acute headache is common in the emergency department. Most cases are induced by virus or migraine, and require only medical therapy. Some are emergency cases, such as aneurysm rupture, intracranial hemorrhage or tumor with increased intracranial pressure. But sometimes, rare cases present in the ER. We describe a patient who visited our ER with the chief complaint of headache with no neurologic deficit. She had a rare case of cerebral sparganosis. Most ER physicians are not familiar with this disease. ER physicians need to be alert to diagnose emergency cases with headache. Many people in eastern Taiwan drink unboiled water and eat raw meat. When encountering a case of brain tumor, parasite infection should be included in the differential diagnosis in endemic areas.

2. Case report

A 47-year-old woman presented in the ER with severe headache of 2 weeks' duration, associated with nausea. She denied fever, neck pain, dizziness, vertigo, unilateral limb weakness, blurred vision and trauma history. She had no history of systemic disease or

medical problems and had not had this type of headache previously. She denied drinking alcohol or smoking. She was an indigene, and lived in Hualien county. Medical management did not relieve the severe headache and her systolic blood pressure was elevated to 152 mmHg. Unenhanced brain computed tomography (CT) revealed a large area of cerebral edema in the left temporal-parietal lobes with mass effect (Fig. 1A). Under the impression of increased intracranial pressure and brain tumor, a neurologic surgeon was consulted. Brain magnetic resonance imaging (MRI) with contrast showed a 3.6×3.5×3.4 cm mixed intensity mass lesion occupying the left occipital lobe (Figs. 1B & 1D). After contrast injection, there was heterogeneous abnormal enhancement (Fig. 1C). The differential diagnosis included neoplasm, abscess, or a lesion mimicking a neoplasm. Although a metastatic tumor was suspected, a primary tumor could not be found. A biopsy done during surgical intervention revealed a 3-cm left occipital tumor that was rubber hard and grossly yellow. Pathological findings showed granulomatous inflammation in the brain tissue with a dead parasite body. Calcareous bodies were evident inside the parasite (Fig. 2). Sparganosis was considered. Two weeks later, she was discharged and she had recovered completely.

3. Discussion

Spirometra mansoni is a cestode (also known as a tapeworm). It has two intermediate hosts. The eggs of *S. mansoni* hatch in water and become coracidium (ciliated larvae), which are consumed by *Cyclops* (first intermediate host). Coracidium develop into procercoids in *Cyclops*. When a fish, snake, frog or amphibian (second intermediate host) drinks water containing *Cyclops*, the procercoids penetrate the intestine and migrate to various organs to become a plerocercoid

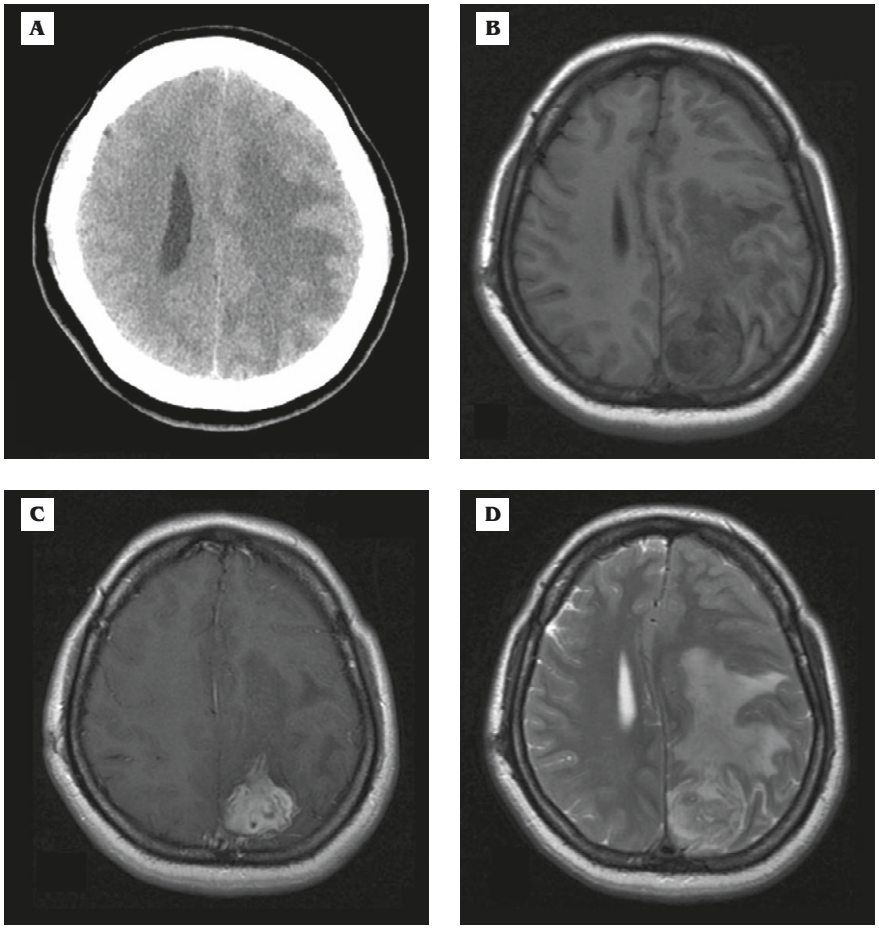

Fig. 1 — Brain imaging of a 47-year-old woman with a history of headache presenting in the ER. (A) An extensive area of white matter hypoattenuation is seen on unenhanced computed tomography with mass effect. (B) A mixed intensity lesion and hypointensity in the white matter are seen on T1-weighted fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging (MRI) and mass effect is noted. (C) T1-weighted FLAIR postcontrast MRI shows a heterogeneous enhanced mass in the left parietal lobe. (D) T2-weighted MRI of the same section as (B) shows a hyperintense area in the left parietal lobe with a heterogeneous mass near the occipital cortex.

called a sparganum. The human body is an accidental intermediate host [1–3]. People get infected by drinking unboiled water containing *Cyclops* or by ingesting inadequately cooked meat, or by applying the raw flesh of the infected host as a poultice to an open wound. Our patient lived in the countryside in eastern Taiwan and may have been infected by drinking unboiled water or eating uncooked meat.

Sparganosis most frequently involves the subcutaneous tissue and muscle. It is most frequently observed in China, Korea, Japan, and Southeast Asia [2]. Infections with central nervous system involvement are extremely rare [2]. The disease most frequently involves the cerebral hemisphere, especially the frontoparietal lobes. In some cases of cerebral sparganosis, a live worm was removed from the cerebrum [4–7]. Contralateral migration of cerebral sparganosis through the splenium has been observed [8].

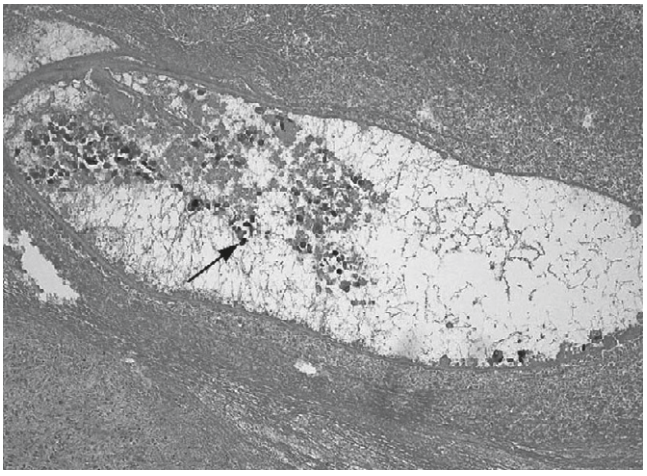

Fig. 2 — Granulomatous inflammation in the brain tissue with a dead parasite body noted (hematoxylin & eosin, 200×). Calcareous bodies (arrow) are evident inside the parasite.

Six patients with this diagnosis have been reported in Taiwan. The details are summarized in Table 1 [9–13]. There were three men and three women ranging in age from 19 to 74 years. Presenting symptoms included seizures (n=4), limb weakness or clumsiness (n=5), headache (n=1), and sudden loss of consciousness (n=1). The duration of symptoms before diagnosis ranged from 1 month to 12 years. All patients underwent surgery and four patients received stereotactic techniques with good outcomes. Only one of the six patients had an ELISA study of the cerebrospinal fluid for sparganum, which was performed in Korea.

Our patient suffered from a severe headache for about 2 weeks without obvious neurologic signs and accompanying symptoms when she presented in the ER. The symptoms and signs depend on the location. It may start with a severe headache, as in our case, mimicking encephalitis or subarachnoid hemorrhage. Most patients have longer durations of illness than our patient, with some cases lasting for months to several years. A long history of seizures is the most common clinical manifestation [1,2,14]. Because of the prolonged course of headache and poor response to analgesics, imaging studies were done for our patient. According to the literature, the main CT findings consist of white matter hypodensity with adjacent ventricular dilatation, an irregular or nodular enhancing lesion and small punctuate calcifications [8]. The most prominent MRI findings are widespread white matter degeneration and cortical atrophy with consequent ipsilateral ventricular enlargement and a decreased volume of the ipsilateral crus cerebri [2]. The most characteristic finding is a tunnel sign on postcontrast MRI. The most common finding is bead-shaped enhancement [15]. The MRI findings in our patient were similar to reported characteristics. However, ipsilateral ventricular compression and mass effect with a midline shift to the right was found in our case. Why was there a mass effect instead of white matter atrophy with

Table 1 — Summary of clinical characteristics of 6 patients of cerebral sparganosis reported from Taiwan

Ref.	Age (yr)/sex	Symptom	Duration of symptoms	History of ingestion of infected food	ELISA		Treatment
					Serum	CSF	
9	52/F	Seizure, leg clumsiness	3 mo	Underground water	NP	NP	Removal of live worm*
9	49/F	Headache, right upper limb weakness	3 mo	Unboiled water	NP	NP	Removal of live worm*
10	67/F	Intermittent right limb weakness, sudden onset of change in consciousness	2 mo	Unknown	NP	NP	Removal of degenerated worm
11	74/M	Seizure, right side weakness	1 mo	Drinking inadequately treated water	NP	NP	Removal of degenerated worm*
12	19/M	Seizure	12 yr	Unknown	NP	†‡	Removal of live worm
13	64/M	Seizure, left leg weakness	4 mo	Underground water	NP	NP	Removal of degenerated worm*

*Removal by stereotactic technique; †ELISA test of CSF performed by Yonsei University College of Medicine, Korea; ‡seropositivity for *Spirometra mansoni*. ELISA=enzyme-linked immunosorbent assay; CSF=cerebral spinal fluid; NP=not performed.

adjacent ventricular dilatation? Almost all previously documented patients were ill for a longer period of time than was our patient. The mass effect in our patient was probably related to acute inflammation of the brain. Serial images are needed to observe if the mass effect resolves with time to be replaced by white matter atrophy.

A preoperative diagnosis is difficult to make because the disease is very rare and there tends to be no specific clue in the hematologic and biochemical laboratory data. Also, doctors are not familiar with the CT and MRI findings of cerebral sparganosis. ELISA (serum and cerebral spinal fluid) is very sensitive and essential for diagnosis [2,4].

In the histological investigation of cerebral sparganosis, it was noted that there were variable degrees of mummification of worms found in the center of granulomatous lesions, reflecting differences in the time that had elapsed between ingestion and removal of the worm from the brain [2]. The degenerated worm is reported to contain multiple characteristic calcareous bodies (calcospherules), as in our patient.

Good long-term outcomes were only achieved in patients who had undergone complete removal of the granuloma including the live or degenerated worm. Early detection and complete surgical removal of the sparganum, together with the surrounding granuloma, is the treatment of choice [2]. In one case report of a patient with a long history of neurologic deficit, the removal of the granuloma containing the degenerated worm was not expected to improve the symptoms. However, the patient had been suffering from motor aphasia, right hemiparesis and epilepsy for 10 years; therefore, he strongly requested a biopsy to obtain a diagnosis. After removal of the small ring-enhanced mass, the patient's intractable seizures became controllable [1].

Cerebral sparganosis is extremely rare but should be considered in the differential diagnosis of metastatic brain tumor. Chronic headache with seizures for months to years is the classic chief complaint. Imaging studies and ELISA of the serum and cerebral spinal fluid can help in diagnosis. Humans can be infected by drinking unboiled water or ingesting inadequately cooked meat containing *Cyclops*, or by applying the raw flesh of an infected frog to an open wound. In Hualien, which is in eastern Taiwan, many people live in rural areas and on the mountains, and are used to drinking unboiled water and eating raw frog and boar meat. Emergency physicians in this city must consider these local customs and their relationship with parasite infection.

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