Cerebral Paragonimiasis
—Report of Five Cases—

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Abstract

Five cases of cerebral paragonimiasis presenting with hemianopsia, convulsion, and gait disturbance are discussed. The cases were all in the chronic stage. The intradermal paragonimiasis reaction, complement fixation, and Ouchterlony tests were not useful for diagnosis. Computed tomography demonstrated calcifications in all cases in sites consistent with the foci of symptoms. Surgical treatment in two cases failed to improve symptoms.

Key words: intracranial paragonimiasis, Paragonimus pulmonalis, computed tomography, surgical indication, Ouchterlony test

Introduction

Cerebral paragonimiasis is the parasitic infectious disease most frequently encountered by neurosurgeons. However, recent developments in epidemiology and antiparasitic agents have considerably reduced the incidence of this and other parasitic infectious diseases. Nevertheless, even today cerebral paragonimiasis occasionally occurs and it remains an important differential diagnosis of intracranial diseases with calcification.

Here, we report five cases of cerebral paragonimiasis which occurred during the past 5 years and discuss the method of diagnosis and the indications for surgical treatment.

Case Reports

Case 1: This 48-year-old female had eaten freshwater crabs during childhood. At about 6 years old, she had two episodes of generalized convulsive seizures. An episode of transient left hemiplegia occurred at about 12 years old. About 2 years ago, she noticed a defect in her visual field. An ophthalmologist found left homonymous hemianopsia.

No other neurological abnormalities were found on admission to our hospital. Plain skull x-rays showed an egg shell-like calcification in the right temporal region (Fig. 1A). A computed tomographic (CT) scan revealed multiple, oval-shaped high-density areas surrounded by low-density areas from the right temporal to parietal lobes (Fig. 2A). A plain chest x-ray showed calcification in the pulmonary field. An intradermal test for paragonimiasis was questionably positive, while both the complement fixation and Ouchterlony tests were negative.

The calcified mass was removed through a right temporal craniotomy. Dead Paragonimus eggs were found in the resected cyst (Fig. 3). The mean size of the eggs was 49.7 × 83.8 μm (n = 15). Some egg shells thickened at the operculum and approximately half had the maximum width on the operculum side. The low-density area on the CT scan surrounding the cyst was fibrous gliosis.

Postoperatively, the left homonymous hemianopsia and the neurological symptoms remained unchanged.

Case 2: This 36-year-old male had eaten freshwater crabs during childhood and sometimes coughed up bloody sputum. He also repeatedly experienced convulsive seizures. After childhood, no symptoms manifested until he had an attack of generalized convulsive seizures with loss of consciousness. He was admitted to our hospital.
There were no abnormal neurological findings except for a slight mental deterioration. Plain skull x-rays revealed faint, poorly defined calcifications scattered over the right frontal area (Fig. 1B). A CT scan showed oval-shaped high-density masses surrounded by low-density areas in the right frontal lobe (Fig. 2B). Chest x-rays found no abnormalities. The intradermal test for paragonimiasis, complement fixation test, and Ouchterlony test were all negative.

The calcified masses were removed through a right frontal craniotomy. Dead Paragonimus eggs were detected in the resected cysts (Fig. 4). The mean size of the eggs was $52.0 \times 74.8 \mu m$ ($n = 25$). Eggs were also detected in the tissue preparations. In nine of 16 eggs which had thickness, the egg shell was thickened opposite the operculum, and the mean size of 16 eggs was $48.5 \times 79.4 \mu m$. In 18 of 25, the width was maximum in the middle, and five of 25 at the operculum.

Anticonvulsant agents were administered postoperatively. No recurrence of convulsive seizures occurred.

**Case 3:** This 48-year-old male had episodes of generalized convulsive seizures during the lower grades in elementary school and coughed bloody sputum at 12 years old. Paragonimiasis was diagnosed at a hospital and worm cysts resected. Recently, disturbance of memory and gait manifested and he visited another hospital. A CT scan demonstrated enlargement of cerebral ventricles. There was no history of consumption of freshwater crabs.

On presentation at our hospital, he was predemential (17.5 points on the Hasegawa dementia scale) and demonstrated memory disturbance. Right homonymous hemianopsia was also observed. He walked with a broad gait (brachybasia). Plain skull x-rays showed calcification in the left occipital region (Fig. 1C). A CT scan revealed marked enlargement of the lateral and third ventricles and a faint low-density area around the ventricles. A high-density area composed of small, multiple oval-shaped masses was present in the left occipital lobe (Fig. 2C). The intradermal paragonimiasis test was questionably positive, while the complement fixation test, Ouchterlony test, and enzyme-linked immunosorbent assay were negative.

A ventriculoperitoneal shunt was performed under a diagnosis of hydrocephalus. Postoperatively, the gait disturbance improved and the memory disturbance gradually improved. However, the homony-
mous hemianopsia did not change.

**Case 4:** This 73-year-old female had eaten freshwater crabs at around 25 years old, but no abnormalities in the nervous and respiratory systems had occurred. At around 70 years old, diplopia developed. Recently, paralysis of the right facial nerve appeared.

Neurological examination detected diplopia, paralysis of the right facial nerve, and hypacusis of the left ear. Plain skull x-rays found faint calcification in the left temporal region (Fig. 1D). A CT scan revealed multiple, oval-shaped high-density masses at the left Sylvian fissure (Fig. 2D). Plain chest x-rays found no abnormality. The intradermal paragonimiasis test was questionably positive, while the complement fixation and Ouchterlony tests were negative.

No treatment was given but her course was carefully followed. After 3 months, the paralysis of the right facial nerve improved, but the diplopia and hypacusis of the left ear did not change.

**Case 5:** This 75-year-old female had eaten freshwater crabs during childhood. Left hemiplegia appeared at around 5 years old, but improved fairly well by elementary school. Generalized convulsive seizures with loss of consciousness occurred frequently at between 18 and 40 years old. She received anticonvulsant drugs. She presented at our hospital with disturbance of gait.

Neurological examination found slight left hemiplegia and cerebellar symptoms. Plain skull x-rays found irregular-shaped, poorly defined calcifications in the vermis and right hemisphere of the cerebellum (Fig. 1E). A CT scan revealed multiple, small, oval-shaped high-density masses surrounded by low-density areas in the same locations (Fig. 2E). Plain chest x-rays showed calcification in the liver. The intradermal paragonimiasis test was positive, but the complement fixation and Ouchterlony tests were negative.

No treatment was initiated and the course of the disease was followed.

The clinical features of these five cases are summarized in Table 1.

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**Fig. 2 CT scans.** A: Case 1, B: Case 2, C: Case 3, D: Case 4, E: Case 5.
Fig. 3 Case 1. upper: Macroscopic appearance of excised tumor tissue. lower: Microscopic appearance of *Paragonimus pulmonalis* egg. ×400.

Fig. 4 Case 2. upper: Macroscopic appearance of excised tumor tissue. lower: Microscopic appearance of *Paragonimus pulmonalis* egg. ×400.

Table 1 Summary of five cases with intracranial paragonimiasis

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Neurological symptoms</th>
<th>Age at onset (yrs)</th>
<th>Location of calcification</th>
<th>History*</th>
<th>ST</th>
<th>CFT</th>
<th>OT</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>48</td>
<td>F</td>
<td>Lt hemianopsia</td>
<td>6</td>
<td>rt temporal to parietal</td>
<td>+</td>
<td>±</td>
<td>−</td>
<td>−</td>
<td>removal of granuloma</td>
</tr>
<tr>
<td>2</td>
<td>36</td>
<td>M</td>
<td>episode of convulsion</td>
<td>6</td>
<td>rt frontal</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>removal of granuloma</td>
</tr>
<tr>
<td>3</td>
<td>48</td>
<td>M</td>
<td>memory disturbance, gait disturbance</td>
<td>7</td>
<td>lt occipital</td>
<td>−</td>
<td>±</td>
<td>−</td>
<td>−</td>
<td>VP shunt</td>
</tr>
<tr>
<td>4</td>
<td>73</td>
<td>F</td>
<td>diplopia</td>
<td>?</td>
<td>lt Sylvian fissure cerebellum</td>
<td>+</td>
<td>±</td>
<td>−</td>
<td>−</td>
<td>no</td>
</tr>
<tr>
<td>5</td>
<td>75</td>
<td>F</td>
<td>gait disturbance</td>
<td>5</td>
<td></td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>no</td>
</tr>
</tbody>
</table>


Discussion

Paragonimiasis is a parasitic disease distributed worldwide. In Japan, infection by *Distoma westermani* and *Paragonimus Miyazaki* are well known. Human infection occurs by intake of raw freshwater crabs, the second intermediate host, through contact with utensils used to cook the crabs,
or by ingesting metacercaria while playing with the crabs. Four of five our cases had a history of intake of crabs, of which two had consumed both Eriocheir japonicus and Geothelphusa dehaani.

Although the original infection site of Paragonimus is the lung, heterotopic parasitism is not uncommon. The most difficult problem of heterotopic paragonimiasis is erratic parasitism in the brain, which accounts for 30–60% of cases. The frequency of published articles dealing with paragonimiasis has declined considerably since the 1970s with the marked decrease in paragonimiasis, and cases of cerebral paragonimiasis are now rare. None of our cases represent fresh infection, but have a history of more than 30 years since initial infection.

Miyazaki named the large type of Distoma westermani Paragonimus pulmonalis and the small type which gamogenizes Paragonimus westermani. The intermediary host of the former is Eriocheir japonicus, and that of the latter is mainly Geothelphusa dehaani. It was therefore interesting to identify the species of Distoma westermani causing the cerebral paragonimiasis. The size of worm eggs in the tissue does not represent the true size. However, examining the worm eggs detected showed that the eggs in Case 1 were larger than those of Paragonimus westermani and the eggs in Case 2 were also a little larger, and more like the eggs of Paragonimus pulmonalis. Furthermore, in some eggs the width was maximum at the operculum and the egg shell thickened opposite the operculum. Therefore, the assumption that these two cases were caused by Paragonimus pulmonalis is reasonable.

Symptoms of acute paragonimiasis include fever, headache, nausea, vomiting, hemiplegia, and convulsive seizures, etc. These are thought to result from the acute brain inflammation caused by erratic parasitism. Chronic symptoms include convulsive seizures, particularly the Jackson type, disturbance of vision, visual field, and movement, and perception. Most symptoms result from the space-occupying lesions due to paragonimiasis. In four of our cases, acute symptoms manifested during infancy or childhood suggesting that infection occurred at this period. In the other one case, freshwater crabs was eaten in later years and the acute symptoms were not clearly demonstrated. In four cases, there was a fairly long period without symptoms of nervous system involvement after subsidence of the invasion or acute symptoms. Then the chronic stage with characteristic symptoms as chief complaints began. The chronic symptoms corresponded roughly to the calcification nidus at the site of paragonimiasis.

Paragonimiasis is diagnosed by the historical, neurological, radiological, and serum immunological examinations. Plain skull x-rays demonstrate no abnormalities during the acute stage, but calcification is detected during the chronic stage. The final calcification due to cerebral paragonimiasis is into four types. Round or oval-shaped masses of cystic calcification are characteristic of cerebral paragonimiasis. About 20% of calcifications due to cerebral paragonimiasis are of this type and are decisive evidence for diagnosis. However, CT is now the most useful radiological modality for the cerebral disorders. Gondo et al. reported that multilocular, ring-like enhancement with surrounding low-density areas are characteristic of the acute stage of cerebral paragonimiasis. Udaka et al. reported enlarged brain sulci and cerebral ventricles, multiple, mainly oval-shaped of various sizes, and extensive low-density areas around and connected to the calcification as characteristic of the chronic stage. All our chronic cases demonstrated the multiple, round or oval-shaped calcifications. The low-density areas surrounding the calcifications were not extensive and some cases did not demonstrate enlarged cerebral ventricles and brain sulci. Multiple, round or oval-shaped calcifications do not occur in other diseases associated with calcification on CT scans and, therefore, are characteristic of cerebral paragonimiasis.

Serum immunological examinations include the intradermal paragonimiasis reaction, complement fixation reaction, and Ouchterlony method. The intradermal paragonimiasis method is easy to perform and highly specific and is the most useful test for differential diagnosis. The reaction becomes positive about 4 months after infection and remains positive long after complete cure. However, if over 10–20 years have passed since death of the worm, the reaction will be negative. Case 2 demonstrated a negative intradermal test and dead eggs were obtained from the calcified lesions. The positive complement fixation reaction becomes negative soon after the death of the worm and, therefore, is useful for judging the efficacy of the treatment. The Ouchterlony or immunoelectrophoretic method is more specific than the intradermal paragonimiasis reaction and is useful long after the death of the worm. Our cases had intervals of over 30 years since infection. The intradermal reaction was positive in one, questionably positive in three, and negative in one, and no positive reaction to the complement fixation or Ouchterlony test was observed. Therefore, CT findings are most useful for diagnosis in cases with long intervals after infection.

Indications for surgical treatment include a history

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of less than 2 years after onset, symptoms of progressive elevation of intracranial pressure, difficult differential diagnosis from brain tumor, focal lesions of the epilepsy or convulsive seizures, etc. Chronic neurological symptoms have not been reported to improve postoperatively, although anticonvulsant dosage may be reduced, continued administration is necessary. None of our surgical cases showed improved neurological symptoms after treatment.

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References


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