

Original Article

Cerebral paragonimiasis with subcutaneous paragonimiasis on the face: a case of ectopic paragonimiasis in a child

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Abstract: *Objectives:* Although the primary focus of *Paragonimus* infection is the lung, erratic migration of the larvae might cause ectopic paragonimiasis. The most frequent locations of ectopic paragonimiasis are the brain, spinal cord, eyes, genital organs, abdominal cavity, and subcutaneous tissues. Collecting a detailed travel and dietary history is essential for the diagnosis and treatment of ectopic paragonimiasis, but physicians often ignore this because ectopic paragonimiasis is rare and can be missed easily. In this article, we report a complicated ectopic paragonimiasis case of a 12-year-old boy who presented with cerebral *Paragonimus westermani* (*P. westermani*) and subcutaneous paragonimiasis on his face. *Case presentation:* A 12-year-old Chinese boy with fever and severe headache for 3 months and facial swelling for 15 days was admitted to the emergency room in May 2010. He had a history of annual exposure to grilled freshwater crabs for 8 years. Serum antibody test for paragonimiasis was positive. Fecal examinations for *P. westermani* eggs was negative. Chest x-ray and computed tomography (CT) showed cavity-like lesions in the right upper lung. Brain magnetic resonance imaging (MRI) showed multiple signal abnormalities in the corpus callosum, right temporal lobe, and frontal lobes. Biopsy of subcutaneous nodule in the facial skin lesion revealed marked eosinophil infiltration. Based on the history, clinical symptoms, and investigations after admission, treatment with praziquantel and prednisone was initiated. The patient responded well to the treatment. *Conclusions:* This patient was given a diagnosis of ectopic paragonimiasis presenting with cerebral paragonimiasis and subcutaneous paragonimiasis on the face. To our knowledge, this is the first report of ectopic paragonimiasis involving both brain and facial skin lesions. The patient responded well to praziquantel and prednisone therapy.

Keywords: Ectopic paragonimiasis, *paragonimus westermani*, cerebral paragonimiasis, skin lesion, MRI, praziquantel

Introduction

Paragonimiasis occurs after ingestion of Crustacea carrying *P. westermani* or other species. It is endemic in South Asia and the Far East and remains highly prevalent in Japan, Korea, China, and Taiwan [1, 2]. An infection with paragonimiasis is caused by eating raw or insufficiently cooked freshwater crabs, shrimp, or crayfish infected with *metacercariae*. The *metacercaria* excysts in the host's small intestine and penetrates into the abdominal cavity and then migrates through the diaphragm into the lungs [3]. The main habitat of *Paragonimus* is within the lung. Although the primary focus of *Paragonimus* infection is the

lung, erratic migration of the larvae might cause ectopic paragonimiasis. The most frequent locations of ectopic paragonimiasis are the brain, spinal cord, eyes, genital organs, abdominal cavity, and subcutaneous tissues [4]. Among all these ectopic sites, cerebral paragonimiasis has been described as the most serious and sometimes fatal form of extrapulmonary paragonimiasis. In the cases of cerebral paragonimiasis, *P. westermani* enters the cranial cavity through the jugular foramen or carotid canal and usually invades temporal and occipital lobes of the brain [3].

Compared to cerebral paragonimiasis, cutaneous infestation is a rather rare form of ectopic

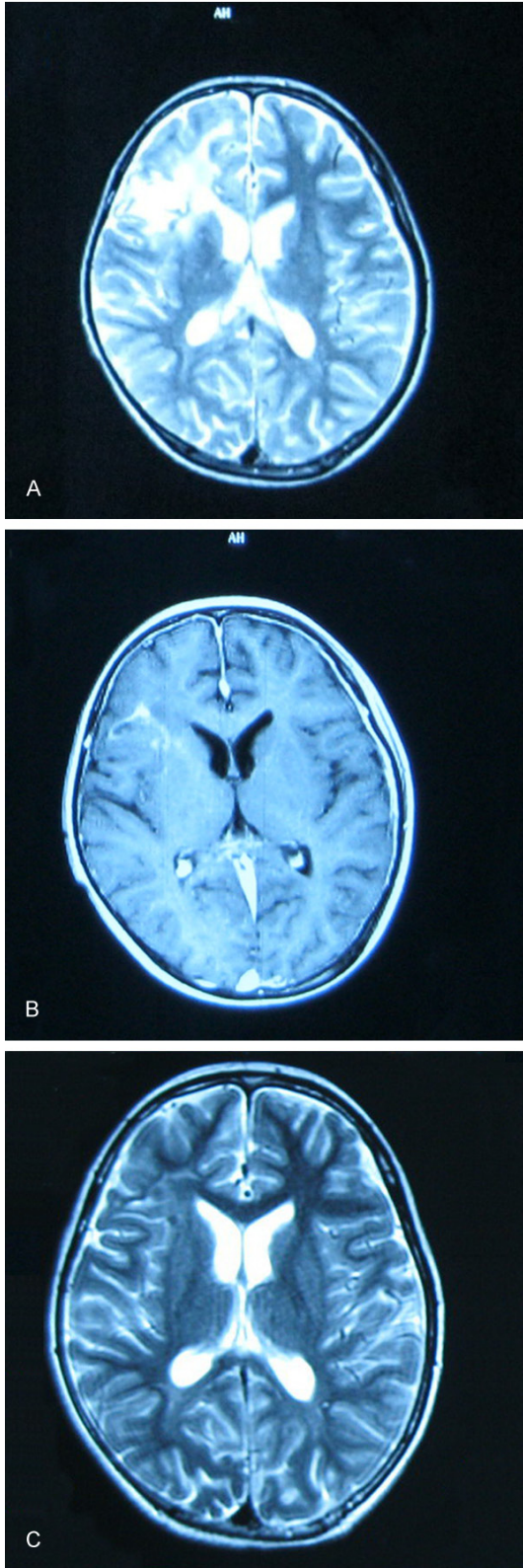


Figure 1. Brain magnetic resonance images. A. T2 weighted axial image (T2WI) showed multiple signal abnormalities (long T2) in corpus callosum, right temporal lobe, and frontal lobes. B. T1 weighted ax-

ial image (T1WI) showed marked radial contrast enhancement around brain lesions. C. After 6 months, the second brain MRI showed significant improvement on intracranial lesions.

paragonimiasis [5, 6]. Cutaneous paragonimiasis usually appears on the skin of the abdomen, groin, or upper chest wall. Other locations of infection are exceptional, like the fingers or face [7]. Here, we report a case of a 12-year-old Chinese boy who was diagnosed with cerebral *P. westermani* and subcutaneous paragonimiasis on his face. This is the first report on ectopic *P. westermani* involving both the brain and face in children.

Case presentation

A 12-year-old Chinese boy was admitted to the hospital in May 2010 due to ongoing fever, headache, and left facial swelling. The intermittent fever and severe headache started 3 months earlier, and then the boy developed facial swelling 15 days prior to admission. His highest body temperature during the course was 39.3°C, but there was no particular fever pattern. There was also no correlation between fever and paroxysmal headache. He had no nausea, vomiting, or lethargy. There was no seizure, giddiness, speech difficulty, hemiplegia, hemianopia, vision change, or other neurological concerns. No respiratory symptoms presented. His skin lesion was first noticed 15 days before admission, and he presented with facial swelling from the left eye to left cheek. Besides the swelling, the left side of the face showed no color change. The boy is a local resident from a mountainous area in southern China and had a history of eating grilled freshwater crabs every autumn over the past 8 years.

His first CBC showed an absolute RBC count of $4.19 \times 10^{12}/L$, absolute WBC count of $5.0 \times 10^9/L$ (neutrophil 17.7%, lymphocyte 45.4%, monocyte 3.4%, eosinophil 31.8%, and basophil 1.7%), Hb level of 110 g/L, and platelet count of $238 \times 10^9/L$. This result revealed that the patient had eosinophilia with an absolute eosinophil count of $1.6 \times 10^9/L$, which accounted for 31.8% of total white blood cells. Serum immunoglobulin levels were unremarkable. Erythrocyte sedimentation rate level was slightly elevated (24 mm/L). Testing on cerebrospinal

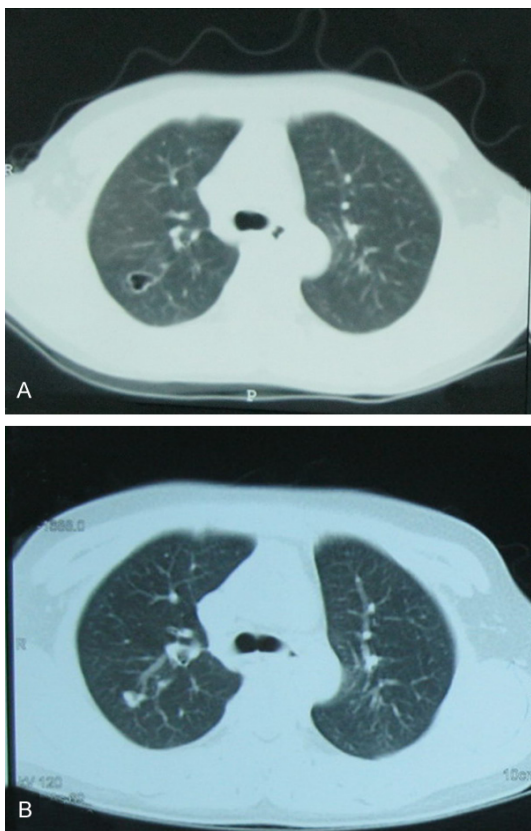


Figure 2. Chest CT images. A. Chest CT showed cavity-like lesions in the right upper lung. B. After 1 month, the second chest CT showed cavity-like lesions in the right upper lung have been completely absorbed.

fluid (CSF) showed normal intracranial pressure and absolute WBC count of $4 \times 10^6/L$ (mononuclear leukocyte accounted for 80%). All the cultures were negative. Tuberculin skin test was negative. Chest x-rays showed patching shadow with obscure boundary in the second intercostal space of the right upper lobe. Chest CT showed cavity-like lesions in the right upper lung (**Figure 2A**). Brain MRI revealed multiple signal abnormalities (long T1 and long T2) in the corpus callosum, right temporal lobe, and frontal lobes. Radial contrast enhancement around these lesions was also seen on MRI scan (**Figure 1A, 1B**).

Because of the patient's dietary history, clinical symptoms, and initial laboratory and radiologic investigations, few specific tests for paragonimiasis were applied. Microscopic examination of a respiratory specimen (single sputum specimen test) and a stool specimen (stool examination for parasite eggs, protozoan cysts, and trophozoites) were all negative. Se-

rum enzyme-linked immunosorbent assay (ELISA) antibody test for paragonimiasis was positive. The biopsy of a subcutaneous nodule in the facial skin lesion revealed eosinophilic granuloma with marked eosinophil infiltration (**Figure 3**).

Based on all of these findings, a diagnosis of cerebral paragonimiasis with subcutaneous paragonimiasis on the face was reached. An effective antihelminthic therapy against paragonimiasis was initiated. The patient underwent three consecutive courses of deworming treatment in a month. For each course, he received praziquantel at 35 mg/kg and prednisone at 1.5 mg/kg daily for 4 consecutive days. The patient's symptoms, including fever and headache, went away once the first therapy course was completed. His facial swelling significantly improved. One month after initiation of treatment, another chest CT was done to follow up on the lesions located in the right upper lung. The f/u CT showed the lesions were completely absorbed (**Figure 2B**). The left side of his face returned to normal after 3 months. Half a year after discharge, a follow-up brain MRI showed significant improvement on the multiple intracranial lesions (**Figure 1C**).

Discussion

Paragonimiasis is a parasitic disease that strikes carnivores, causing a subacute to chronic inflammatory process of the lung. Humans become infested by eating raw, pickled, or half-boiled freshwater crab or crayfish, carrying encysted *metacercariae* of *P. westermani*. The larvae could penetrate intestinal walls first and then migrate to the lungs. Aberrant migration of larvae can cause ectopic paragonimiasis. The extrapulmonary forms were found in about 2% of all cases [9]. The brain, unfortunately, is the primary site of ectopic paragonimiasis. The larvae might traverse the mediastinum and neck and migrate to the brain through one of the basal foramina [8].

No good epidemiological study has focused on paragonimiasis in China; therefore, we do not know this disease's exact incidence. However, we do know the endemic areas of paragonimiasis were sporadically spread over the villages close to small streams in southwestern and eastern China. In this case, the patient was 12 years old, and he and his male friends

Ectopic paragonimiasis in a child

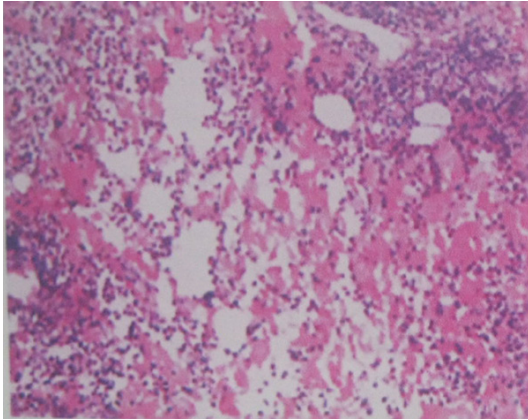


Figure 3. The biopsy of subcutaneous nodule in the facial skin lesion revealed eosinophilic granuloma with marked eosinophil infiltration. (HE staining, $\times 200$).

had eaten grilled freshwater crabs caught in the stream near the village every autumn since he was 4 years old. This may be the reason why cerebral paragonimiasis is predominantly affecting males more than females among children.

Although pulmonary paragonimiasis is often a benign illness, cerebral paragonimiasis is a serious and sometimes fatal disease. In the early acute stage, the predominant symptoms usually resemble meningoencephalitis [4]. The boy from our case had fever and headache for 3 months, and he received treatment for suspected meningitis in a local hospital until he developed facial swelling. Shin Joong Oh reviewed 62 cases of cerebral paragonimiasis [10], and he found that seizures, headache, visual disturbance, and motor and sensory disturbance were the five most common symptoms. He also noticed that mental deterioration, hemiplegia, unilateral hemihypaesthesia, homonymous hemianopsia, and optic atrophy were the five major signs for cerebral paragonimiasis. In our case, the patient's general condition was relatively good. He had no seizures, visual impairment, speech difficulty, hemiplegia, hemianopia, or other disturbance, although he had multiple intracranial lesions according to the brain MRI scan.

Extrapulmonary paragonimiasis can be divided into cerebral, abdominal, subcutaneous, and miscellaneous forms. The brain is the most common extrapulmonary site of infection; subcutaneous paragonimiasis is very rare, and it is commonly identifiable by the appearance

of subcutaneous nodules. Several cases of cutaneous paragonimiasis have been reported [6, 11]. Some case reports found that cutaneous symptoms often occurred before organ involvement. A few other reports showed organ involvement that was followed by cutaneous paragonimiasis or simultaneously detection of both processes. In our case, the facial swelling was followed by neurological symptoms. Hypereosinophilia was then detected upon admission. To our knowledge, this is the first case of subcutaneous paragonimiasis on the face with cerebral paragonimiasis.

To confirm the diagnosis of cerebral paragonimiasis, imaging examinations (e.g., CT MRI), laboratory tests (ELISA and eosinophil counting), and parasitology tests are needed³. We can diagnose paragonimiasis if eggs can be detected in feces, body fluid, or tissue. However, this is not easy for extrapulmonary paragonimiasis. When cerebral paragonimiasis is suspected, CSF tests are highly recommended, as well as brain CT or MRI. Conglomerates of multiple ring-like shadows or enhancements in one hemisphere on contrast MRI, which can also be called grape cluster or soap bubble, are important for diagnosis of chronic cerebral paragonimiasis [3, 12].

In our case, MRI revealed multiple conglomerated long T1 and long T2 signals in the corpus callosum, right temporal lobe, and frontal lobes. Marked radial contrast enhancement around these lesions was also seen on MRI scan. However, these findings were not typical MRI changes for cerebral paragonimiasis, which is probably because the disease stage was still acute instead of chronic. After 6 months, the second brain MRI showed significant improvement on the intracranial lesions.

Eosinophilia suggests the diagnosis of parasitic diseases; it is an important indicator in the suspicion of parasite infection at the early stages. In Robertson's study, peripheral eosinophilia was present among 25% of all patients [13]. In this case, the patient's eosinophil numbers accounted for 31.8% of all WBC on admission. After the first course of therapy, the eosinophil count was normalized. Not every case of paragonimiasis presents with eosinophilia. Eosinophilia is always resolved in the chronic stage [14]. Serum ELISA has been widely applied to diagnose paragonimiasis. This test has very good sensitivity and specificity

in terms of the diagnosis for pulmonary and extrapulmonary paragonimiasis [2]. In the present case, a confirmed diagnosis was made by a positive ELISA result for *P. westermani*.

Praziquantel is known to be effective for treating paragonimiasis. It has a more than 95% efficiency and is definitely effective in the early stage of active cerebral paragonimiasis. In our case, three courses of high-dose praziquantel were applied. The patient received praziquantel at 35 mg/kg per day for 4 days for each course. No adverse effects were noticed during the treatment. In addition, corticosteroids were added to the treatment to reduce inflammatory changes. Prednisone 1.5 mg/kg per dose was applied with praziquantel. The patient responded very well to treatment.

Conclusions

Based on the dietary history, clinical findings, radiologic results, histopathologic analysis, and laboratory testing, this case was diagnosed as cerebral *P. westermani* with subcutaneous paragonimiasis on the face. This is the first case report of cerebral *P. westermani* involving facial skin lesion in children. The patient responded well to praziquantel and prednisone therapy.

Acknowledgements

Written informed consent was obtained from the patient's legal guardian(s) for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Disclosure of conflict of interest

None.

Abbreviations

MRI, Magnetic resonance imaging; CT, Computed tomography; HE, Hematoxylin and eosin; RBC, Red blood cell; WBC, White blood cell; Hb, Hemoglobin; ESR, Erythrocyte sedimentation rate; CSF, Cerebrospinal fluid; ELISA, Enzyme-linked immunosorbent assay.

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