□ Case Report □

Two Cases of Paragonimiasis in Young Siblings Presenting with Pleural Effusion and Subcutaneous Nodules

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Paragonimiasis is the infestation of lung flukes of the trematode genus Paragonimus. This disease is common in Asia, and the southern part of Korea has been known as one of the endemic areas of *Paragonimiasis westermanii* in Korea. Human infection is associated with specific dietary habits, such as eating freshwater crawfish or crabs. In a 6 1/2-year-old boy with pleural effusion and eosinophilia, paragonimiasis was diagnosed by skin test, serologic exam, and histologic identification of the parasites in a skin lesion. The same diagnosis was entertained in his elder sister with silent pleural effusion. We describe herewith these rare cases of paragonimiasis in two siblings who had a history of eating cooked freshwater crabs. **(Korean J Pediatr 2005;48:1385-1388)**

Key Words : Paragonimiasis, Eosinophilia, Pleural effusion, Sibling

Introduction

Paragonimiasis is the infestation of lung flukes of the trematode genus Paragonimus[1]. Species are distributed globally but the disease is common in certain areas of East and Southeast Asia, and the southern part of Korea has been known as one of the endemic areas of *Paragonimiasis westermanii* in Korea[2]. Human infection is associated with specific dietary habits, such as eating freshwater crawfish or crabs. Only a few cases of pediatric cases have been reported in the literature[3-5]. We report herewith two cases of paragonimiasis in siblings who had the history of freshwater crabs ingestion.

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Case Reports

Case 1

A 6 1/2-year-old boy visited a regional hospital because of dry cough for 10 days. He had leukocytosis with eosinophilia and bilateral pleural effusions on chest X-ray. He was referred to our hospital for further evaluation.

On admission, his vital signs were body temperature of 36.4℃, heart rate of 92/min, respiratory rate of 24/min and blood pressure of 100/70/mmHg. On examination, he had acutely ill appearance, and his breathing sounds were decreased in both lower chest. Mild hepatomegaly was found and diffuse swelling on anterior chest was noted. A complete blood count showed white blood cell, 33,800/mm^3 (neutrophils, 16.0%; lymphocytes, 13.9%; monocyte, 2.8%; eosinophils, 66.8%), hematocrit, 34.5% and platelets, 352,000/mm^3. The absolute eosinophil count was 22,600/mm^3. Total serum IgG and IgE were markedly elevated with 2,580 mg/dL and 4,360 IU/mL, respectively. Chest X-ray demonstrated bilateral pleural effusions with a consolidation in

the left lung (Fig. 1A).

Bone marrow aspiration smear showed increase of eosinophilic series (24%) with immature eosinophil precursors on differential count. Cytogenetics and fluorescence *in situ* hybridization for bcr/abl were negative. Reverse transcriptase-polymerase chain reaction (RT-PCR) for FIP1L1-PDGFRA, specific for the diagnosis of hypereosinophilic syndrome (HES)[6], was negative. In the pleural fluid obtained by thoracentesis, an increased percentage of neutrophils and eosinophils was found, but Gram stain and culture were negative.

He had positive skin intradermal test (papule of 10 mm) to *P. westermanii* and subsequent serum *P. westermanii*-specific IgG antibody was positive by enzyme linked immunosorbent assay (ELISA) (optical density value, 0.63), rendering the diagnosis of paragonimiasis. Computed tomography of the brain was normal. With thorough history taking, he and his family members had the history of ingesting cooked fresh water crabs. Several days after admission, a subcutaneous nodule developed on the left upper chest region (Fig. 2A). It was surgically excised and *P. westermanii* young adult fluke was found in the section (Fig. 2B). Praziquantel, 75 mg/kg/day in three divided doses for 2 days, was given and he was discharged with improvement of symptoms and signs. Eight months later, his chest radiography revealed a small amount of pleural effusion and thickening with normalization of pneumonic consolidations. The eosinophil count decreased to normal.

Case 2

A 12-year-old girl, who is an elder sister of case 1, visited a regional hospital because of a palpable, non-tender, subcutaneous nodule on the back. Because of her brother's illness and history of freshwater crabs ingestion, she also was evaluated for paragonimiasis. Physical examination was normal except for her skin nodule. A blood count showed white blood cell of 10,600/mm^3 with eosinophilia (absolute count of 3,180/mm^3). Chest radiography revealed

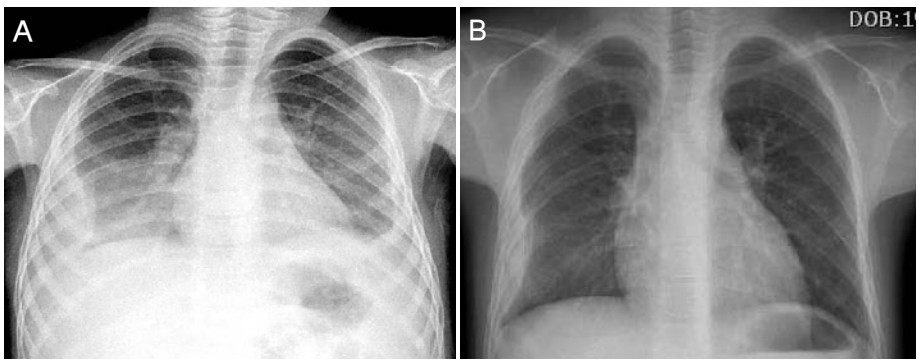

Fig. 1. (A) Initial chest radiograph shows bilateral pleural effusions and right lower lobe consolidation in Case 1. **(B)** After 8 months, follow-up chest radiograph shows remaining small amount of right-sided pleural effusion with thickening.

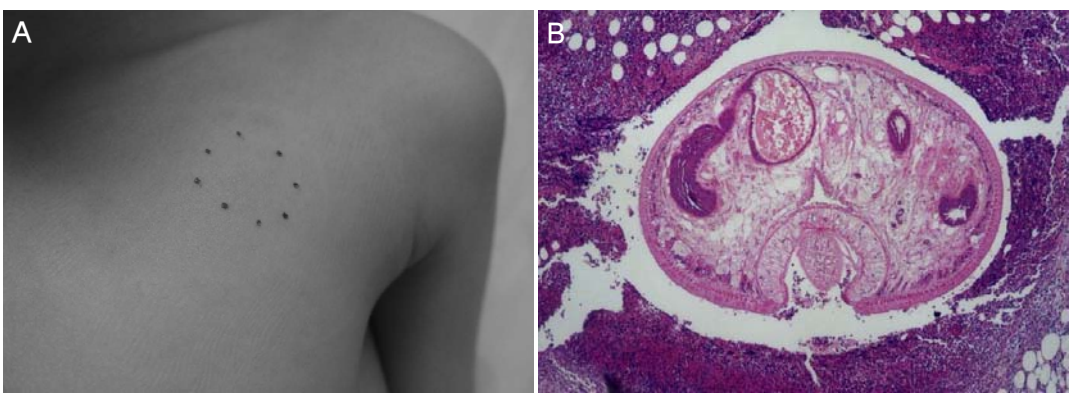

Fig. 2. (A) Subcutaneous nodule on the left upper chest region in Case 1. **(B)** Section of the subcutaneous nodule demonstrates adult fluke of *Paragonimiasis westermanii*(H&E stain, ×400).

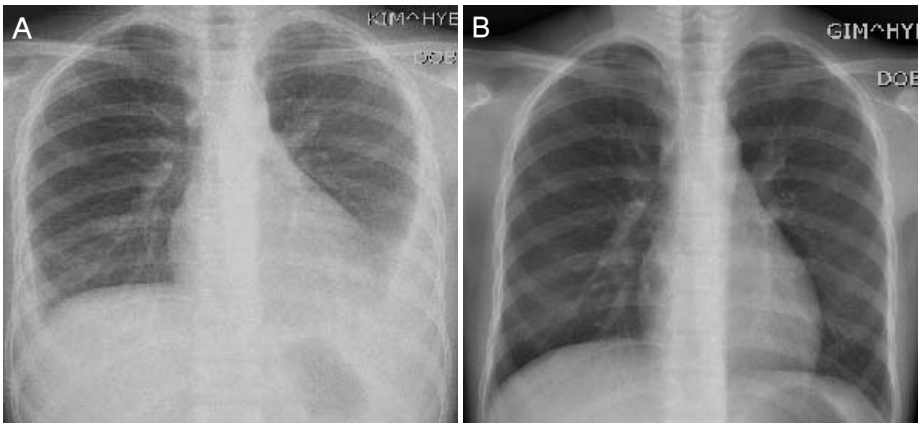

Fig. 3. (A) Initial chest radiograph shows bilateral pleural effusions and left lower lobe consolidation in Case 2. **(B)** After 4 months, follow up chest radiograph shows resolution of previous lesions.

asymptomatic pleural effusion in both and lower lobe consolidation in left (Fig. 3A). Skin test as well as ELISA for *P. westermanii* was positive. Praziquantel was prescribed. At 4 months after treatment, her chest radiography findings became normal (Fig. 3B).

Discussion

More than 10 species of paragonimus infect humans around the world. The most common species, *P. westermanii*, is distributed mainly in South Korea, Japan, India, and Southeast Asia[1, 3]. The eggs from the host's sputum or faeces, hatch in fresh water, enter snails and are released as cercariae. The cercariae penetrate the crustacean shell of the freshwater crab or crayfish (the second intermediate host) and development progresses to the metacercaria stage. The metacercariae ingested by humans or other mammals (the definitive host) excyst in the intestine, migrate to the pleural cavity, where they maturate into the adult worm capable of egg production in the lung during the ensuring 5 to 6 weeks[4]. Human infection generally occurs from specific dietary habits of eating raw or undercooked crabs or crayfish, a second intermediate host, or boar[7]. Metacercariae are sensitive to heat but can adhere to utensils, surface and human hands during meal preparation, causing disease despite adequate cooking of the meal itself[4]. This might explain the occurrence of paragonimiasis in our cases who had the history of ingesting cooked freshwater crabs. Paragonimiasis in the pediatric population was endemic in Korea in association with using raw crayfish juice for the treatment of measles in the past[4, 8]. Although the prevalence of this disease decreased in recent years, the level of infection in endemic areas such as the southern part of Korea may be still high with sporadic case reports[9-11].

The initial parasite migration is usually asymptomatic and symptoms may be acute or chronic[4]. During the acute phase, abdominal pain, diarrhea, and urticaria are followed a few days later by fever, cough, dyspnea, chest pain, and malaise. During the chronic phase, manifestation may be pulmonary or extrapulmonary. The onset of pulmonary symptoms is often months to years after the initial infection. The typical pulmonary symptoms consist of fever, cough, hemoptysis, pleuritic chest pain, dyspnea and wheezing. The physical examination in paragonimiasis is often unremarkable[8]. Chest radiographic findings include patchy infiltration, cavitation and pleural lesion such as pleural thickening, effusion and pneumothorax[8, 10]. These findings may mimick tuberculosis. But our patients had non-specific radiologic findings with pleural effusions. Extrapulmonary paragonimiasis can occur in the various sites such as brain, spinal cord, thoracic muscle, subcutaneous tissue, liver, and spleen, etc[7]. Migrating subcutaneous nodules are usually present in less than 20% of patients[8]. Subcutaneous nodules occur mostly between the abdomen and the knees, with rare presentation in upper chest or back, like in our cases[8].

Eosinophilia is common in parasitic diseases and most patients have an absolute level of more than 500 cells/mm^3 [11]. The degree of eosinophilia in paragonimiasis is usually 10% to 30% with a total WBC of 10,000 to 20,000/mm^3, although a case of extraordinary high eosinophilia (91%;

absolute eosinophil count, 84,000/mm^3) was reported[11]. IL-5 and parasite-derived eosinophil chemotactic factors were known to be important in the mediation of parasite-induced eosinophilia[5].

Patients with moderate to severe eosinophilia should be evaluated for the possibility of having HES[12]. Because absolute eosinophil count of more than 20,000 was present in case 1, diagnostic tests including bone marrow aspiration, cytogenetics, FISH for bcr/abl and RT-PCR for FIP1L1-PDGFRA were performed to exclude the possibility of having HES or clonal eosinophilic disorders.

In addition to positive skin test results, these cases were positive for *P. westermanii* specific immunoglobulin G ELISA, which is known to be very sensitive and specific[13]. Interestingly, the diagnosis in case 1 was confirmed histologically by identification of the young adult parasite in subcutaneous nodule. Radiographic abnormalities were known to gradually improve over months, with most resolving completely by 1 year[8]. Small pleural effusions generally clear with therapy, whereas larger effusions may require either thoracentesis or surgical decortication to prevent long-term pleural complications[3, 7]. In our case 1 small amount of pleural effusion and thickening persisted 8 months after initial treatment, while case 2 showed complete resorption of effusion 4 months after treatment. Praziquantel was given to patients without significant toxicities. Praziquantel was highly effective as the drug of choice with a cure rate greater than 85%[14].

In this report, we present two cases of typical cases of paragonimiasis in siblings who had pleural effusion and eosinophilia. A thorough history taking of ingesting fresh water crabs or crayfish, even after cooking, should be sought to identify these rare cases.

한 글 요 약

흉막삼출과 피부결절로 발현한 초등학생 남매의 폐흡충증 2례

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인체 감염된 폐흡충류가 폐, 복부장기, 뇌 등에 기생하는 경우를 폐흡충증이라 한다. 우리나라를 포함한 아시아의 특정지역에 유행하며 우리나라에서는 전남 및 경남 해안지방 등지에 유행지가 보고되어 왔다. 인체 감염은 민물 가재나 민물 게를 날 것으로 먹을 때 발생한다. 저자들은 과도한 호산구 증다증 소견과 함께 삼출성 흉막염과 피부 침범을 동반한 6세 남아에서 paragonimus 피부 반응검사와 ELISA 검사와 피부 결절의 병리조직학 검사를 통해 폐흡충증을 진단하였고, 동일한 증상을 보인 누나에서도 진단을 하여 praziquantel로 치료한 2례를 경험하였기에 보고하는 바이다.

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