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Radiological findings of alveolar hydatid disease of the lung caused by *Echinococcus multilocularis*.

Ohsaki Y, Sasaki T, Shibukawa K, Takahashi T, Osanai S.

**Radiological Findings of Alveolar Hydatid Disease of the Lung Caused by
*Echinococcus multilocularis***

**Yoshinobu Ohsaki¹, Takaaki Sasaki¹, Kiyoko Shibukawa¹, Toru Takahashi¹, Shinobu
Osanai¹**

¹Division of Respiratory Diseases, Department of Medicine, Asahikawa Medical College.

2-1-1-1 Midorigaoka Higashi, Asahikawa 078-8510, Japan.

Phone +81/166/68-2442

Facsimile +81/166/68-2449

Corresponding author

Yoshinobu Ohsaki, MD, PhD

e-mail yohsaki@asahikawa-med.ac.jp

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Running title: *Radiological findings of alveolar hydatid disease.*

Abstract

Alveolar hydatid disease is a highly malignant form of echinococcosis caused by the larvae of the cestode *Echinococcus multilocularis*. Alveolar hydatid disease always affects primarily to the livers, unlike cystic hydatid disease caused by *E. granulosus*. Occasionally, there are metastases to the lungs and brain from the primary lesions. There have been few reports on the radiological features of alveolar hydatid disease caused by *E. multilocularis*. In the present report, we describe the radiological features of alveolar hydatid disease of the lung caused by *E. multilocularis*. In the cases presented here, multiple nodules were noticed on the chest radiogram. These nodular shadows varied in size and shape. On computed tomographs, the majority of nodular shadows were lobulated and well circumscribed. These nodules were triangular, spherical, oval or linear in shape. We considered that multiple lobulated lesions located between two segments of the lung and showing various shapes were characteristic of pulmonary alveolar hydatid disease caused by *E. multilocularis*.

Introduction

Alveolar hydatid disease is a highly malignant form of echinococcosis caused by the larvae of the cestode *Echinococcus multilocularis*. *E. multilocularis* enters the human body via contaminated drinking water and infects the liver. Progressive larval spread within the liver in a cancer-like fashion leads to death from hepatic insufficiency. Alveolar hydatid disease always affects primarily the liver, unlike cystic hydatid disease caused by *E. granulosus*. Occasionally, there are metastases in the lungs and brain. Echinococcosis caused by *E. multilocularis* is an endemic disease in Hokkaido, Japan.

Four *Echinococcus* strains - *E. granulosus*, *E. multilocularis*, *E. vogeli* and *E. oligarthus* - have been reported to be pathogenic to humans. These four species of *Echinococcus* are morphologically distinct in their adult and metacestode stages, geographic distribution and host specificity (1). *E. granulosus* is widely distributed throughout the world because of its very low intermediate host specificity and its high prevalence in sheep and cattle herding locals. *E. multilocularis* is restricted to the northern hemisphere, with endemic areas extending from the European alpine countries, the former Soviet Union, China, Alaska, South-Central Canada, North of the United States, and Hokkaido where the fox appears to serve as the definitive host and certain wild rodents as intermediate hosts. The cycle involves these canids and rodents including voles, muskrats, meadow voles, and house

mice. In settlement areas, domestic dogs and cats may feed on these rodents, contaminate water and vegetation, and in this way, human beings become accidental intermediate hosts.

E. vogeli and *E. oligarthus* are restricted in their distribution to South and Central America.

The host assemblages include the bush-dog and paca and a wild feline cycle involving the puma/jaguar/Geoffroy's cat and agouti/paca (1).

Majority of *E. multilocularis* infections arise solely in the liver. Pulmonary involvement is seen in 5 to 10% of the cases with hepatic echinococcosis caused by *E. multilocularis*. Wilson *et al.* found only three cases of pulmonary involvement among 33 cases of alveolar hydatid disease (2). The radiological features of pulmonary hydatid disease caused by *E. granulosus* are well documented; multiple cysts are seen in 20% to 30% of the patients (3-6). Whereas there have been few reports on the radiological features of alveolar hydatid disease caused by *E. multilocularis*.

In the present report, we describe the radiological features of alveolar hydatid disease of the lung.

Cases of *E. multilocularis*

Case 1 is a 59-year-old woman. She underwent surgical resection of a large soft tissue tumor in the anterior chest wall ten years previously. The tumor was diagnosed as a granuloma caused by *E. multilocularis* based on pathological findings. Besides hepatic lesions, multiple pulmonary lesions were found on the chest radiogram and the patient was referred to the Division of Respiratory Diseases. Her chest lesions have been monitored for more than ten years.

Case 2 is a 51-year-old woman who has been treated for hepatic echinococcosis caused by *E. multilocularis* for 12 years already. Hepatic lesions were detected after surgical resection of the right iliac bone affected by *E. multilocularis*. Besides the hepatic lesions, multiple pulmonary lesions were detected on the chest radiogram and she was referred to the Division of Respiratory Diseases. She has been followed up for more than seven years. These pulmonary lesions have gradually increased in size during follow-up.

Serological examination was performed in Asahikawa Medical College and the results were positive for *E. multilocularis* in both cases (7).

Findings on the Chest Radiogram

Multiple nodules were detected on the chest radiogram of case 1. There were at least three nodules in the right lung field, and one in the left lung field. In case 2, more than

ten nodules including a 2.5 cm mass were noticed in both lung fields on the chest radiograph. These nodular shadows varied in size and shape. Both patients were treated using **albendazole**. Pulmonary lesions were decreased in size during the therapy and increased in size by termination of the therapy. Figure 1 shows chest radiogram of case 2.

Findings on the Chest Computed Tomography

The diameter of these pulmonary nodules varied from several mm to 2.5 cm. These lesions located between two segments of the lung, and pulmonary veins which distribute between these segments were involved. The majority of nodular shadows were lobulated and well circumscribed. Nodules were either triangular, spherical, oval or linear in shape. These findings seemed to be characteristic of pulmonary alveolar hydatid caused by *E. multilocularis* (figure 2).

Discussion

Alveolar hydatid disease is a consequence of metastasis of the larvae of the cestode *E. multilocularis* from the primary lesion. The parasitic cycle involves fox and rodents, which contaminate water and vegetation, and it occasionally involves domestic dogs. In Japan, systemic parasitic diseases are usually seen as imported cases from endemic regions (8) except for the disease caused by *E. multilocularis*.

In the present observations, we found that the radiological appearance of the pulmonary alveolar hydatid of *E. multilocularis* varied in size and shape. Each pulmonary lobe is divided into individual segments by inter segmental connective tissue and segmental veins. Segmental veins can be differentiated from segmental arteries by HRCT because these arteries locate at the center of the segment in parallel with the segmental bronchus. The characteristic radiological findings of pulmonary alveolar hydatid disease in the present cases included inter-segmental distribution involving these segmental veins. It has been documented that rarely, tumor emboli can be seen in segmental or larger arteries, and seldom, centrilobular nodules and branching linear opacities (tree-in-bud pattern) can be seen in metastatic pulmonary neoplasms (9). Probably, alveolar hydatid disease spreads in a manner similar to that of metastatic neoplasms. We consider this pathomechanism of alveolar hydatid disease as one of the reasons for the distribution of the lesions in this location. Nodules were

lobulated in shape, increased in density and well circumscribed. Also, absence of marked retraction of adjacent organs such as pleura, bronchus and pulmonary vessels seemed to be characteristic of pulmonary alveolar hydatid disease caused by *E. multilocularis*. Cavity formation was not seen in our cases.

Most pulmonary metastatic tumors have smooth margins, probably because they grow relatively uniformly in all directions from their original focus (9); although nodules with spicules or poorly defined margins can also be seen. Primary pulmonary adenocarcinoma is typically observed as a spherical or lobulated lesion, well circumscribed or showing margins retraction of the pleura into the tumor and a distinctive puckered appearance indicating retraction of adjacent organs (10). Usually this characteristic appearance is seen only in primary adenocarcinoma; metastases from the primary lesion lack these characteristics. These differences in radiological features differentiate pulmonary alveolar hydatid disease from metastatic tumors and primary adenocarcinomas of the lung. However, the radiological differentiation of alveolar hydatid disease of the lung from other multiple pulmonary parasitic diseases may be difficult.

Most commonly, pulmonary involvement is evident in the right lower lobe of the lung because of direct spread by the hematogenous route. Our cases lack such tendency of distribution, however, more lesions were seen in the right lung than in the left one. Treugut

et al. reported that radiological findings of alveolar hydatid disease of the lung can be classified into two types (11). Type 1 lesions denote hematogenous spread and result in multiple small, irregular, peripheral lesions. Type 2 manifestations are caused by local extension of the hepatic mass resulting in focal pulmonary involvement, which may be difficult to distinguish from a pulmonary abscess. According to this classification, pulmonary nodules in the present cases were type 1 lesions.

Compare to the development of unilocular hydatid cyst caused by *E. granulosus*, the alveolar hydatid caused by *E. multilocularis* develops very differently. The germinal layer of the metacestode proliferates both exogenously and endogenously. This form of development and infiltration of the host tissues is facilitated by the relative absence of the pericyst and a very thin laminated layer. The metacestode mass thus consists of vesicular tissue composed of 5- to 15-mm-size vesicles that progressively invade the host tissue. Cells from the germinal layer can detach and metastasize to distant organs (1). We speculate that these distinctive features in the pathogenesis of the alveolar hydatid caused by *E. multilocularis* are responsible for the radiological features of the disease in the lungs.

The symptoms of these pulmonary lesions are usually overshadowed by the manifestations of the primary infectious burden of the liver (2). None of our patients complained of respiratory symptoms. Alveolar echinococcosis by *E. multilocularis* caused

up to 100% lethality in untreated patients before the 1970s. Benzimidazole derivatives, mebendazole and albendazole, treatment have shown some promise in the treatment. However, a very high oral dose for three months to more than 17 years is needed to control the disease because of their variable absorptions.

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Figure legends

Figure 1. Chest radiogram of case 2. More than ten nodules including a 2.5 cm mass were noticed in both lung fields

Figure 2. Forty nodules were found in case 1 and case 2. Upper: 17 nodules were detected by high-resolution computed tomography scanning. Lower: 23 nodules were detected by conventional computed tomography scanning. Each figure is 3x3 cm in size.

Figure 1. *Radiological Findings of Alveolar Hydatid Disease by Y. Ohsaki et al.*

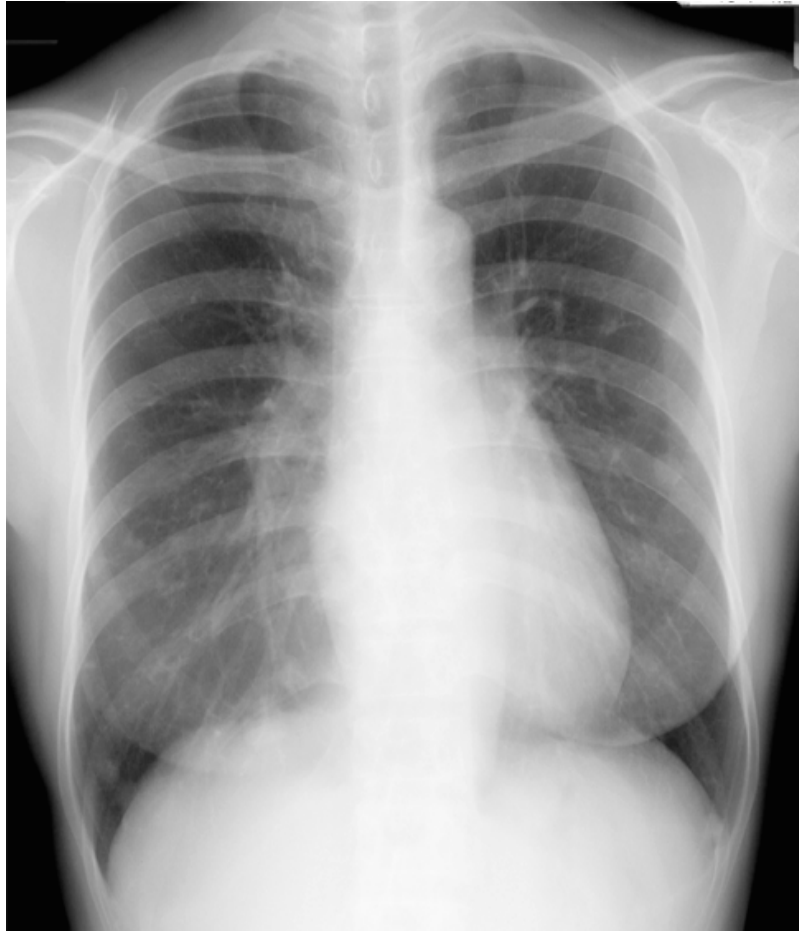


Figure 2. *Radiological Findings of Alveolar Hydatid Disease by Y. Ohsaki et al.*

