

Alveolar hydatid cyst with lung metastasis: A case report

Akciğer metastazı olan alveolar kist hidatikli bir olgu

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Alveolar hydatid disease is a serious and often fatal condition caused by infection with the metacestode form of echinococcus multilocularis. Liver involvement is seen in 90% of the cases. The parasite develops within the liver as a rapid invasive pseudomalignant growth and may make distant metastasis.

We present the case of a 37 year old woman admitted to our gastroenterology department with obstructive jaundice and then diagnosed as alveolar hydatid cyst with lung metastasis. This case is presented due to its rare prevalence.

Key words: Alveolar hydatid cyst, lung metastasis.

Alveolar kist hidatik; echinococcus multilocularisin oluşturduğu ciddi, çoğu zaman da ölümcül bir enfeksiyondur. Vakaların %90'ında karaciğer tutulumu görülür. Parazit karaciğeri hızla işgal edip, pseudomalign bir büyüme gösterebilir ve uzak metastaz yapabilir.

Bu makalede kliniğimize tıkanma sarılığı ile başvurup akciğer metastazlı alveolar kist hidatik tanısı alan 37 yaşında bir bayan hasta sunulmuştur. Hastalığın nadir görülmesi bu vakanın sunum nedenidir.

Anahtar kelimeler: Alveolar kist hidatik, akciğer metastazı.

INTRODUCTION

Alveolar echinococcosis is a parasitic disease caused by the larva form of the cestode *E. multilocularis*. While mice and other rodents are intermediate hosts, foxes are known to be the definite host (1). The disease is endemic in the northern hemisphere (North America, Northern Europe and Central Eurasia). Recent surveys in Central Europe have extended the known geographical occurrence of *E. multilocularis* in foxes from four countries at the end of the 1980 s to at least 11 countries in 1999 (2). Alveolar hydatid cyst is endemic in the Eastern Anatolia region of Turkey. A total of 207 cases were reported between 1962 and 1995 and most of these data depend on hospital records (3). The male/female ratio of the disease is 1.3/1 and it is generally diagnosed in the fourth decade (4). The liver is the primary and most commonly involved organ but cases without liver involvement have also been reported (5). Extrahepatic infection is thought to be the result of the secondary infection which spreads via

metastasis from the hepatic focus. Pulmonary metastasis is the most prevalent and is observed in 22% of cases. Other extrahepatic locations are rare.

In this study a case of alveolar hydatid cyst disease with lung metastasis is presented.

CASE

A 37 year-old female patient from a rural area was admitted to our gastroenterology department with a 20-day history of jaundice, nausea and vomiting. She also complained of itching and a weight loss of about 2 kg in the previous month. She had no respiratory symptoms. The vital signs on admission were: temperature: 36.5 °C, pulse rate: 86/min, respiratory rate: 18/min, blood pressure: 120/70 mmHg. Her physical examination revealed icteric skin and sclera. She also had right upper quadrant tenderness and hepatosplenomegaly. Physical examination

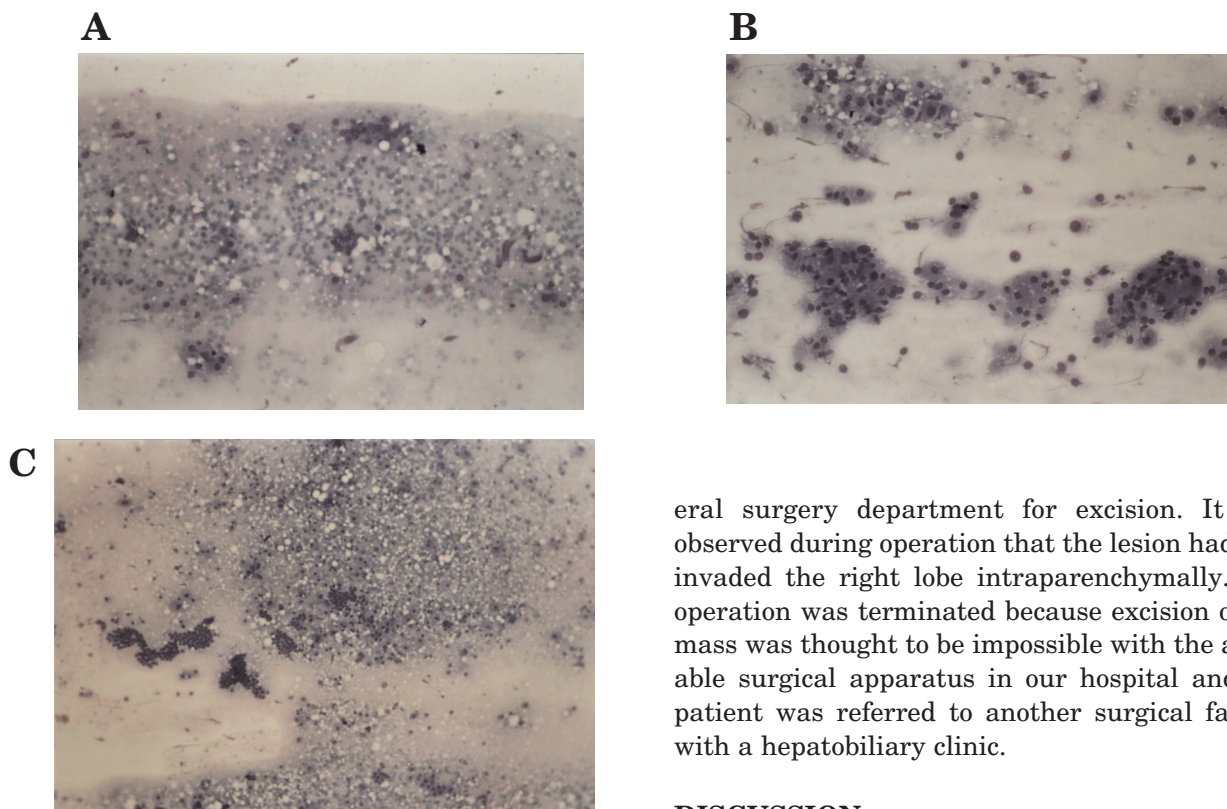


Figure 1. A, B, C Eosinophilic cyst cuticles and hepatocytes

results of other systems were normal and laboratory findings were as follows: positive urobilinogen and bilirubin in urine, AST:71U/L (15-48), ALT:67 U/L (13-40) indirect bilirubin=1.8 mg/dl, direct bilirubin : 4.7 mg/dl, ALP was six times and GGT four times higher than the upper limit of reference value. Her HBsAg, Anti HBc total and anti HBe was positive, HBV DNA: 8 pg/ml while her esophagogastroduodenoscopy was normal. She had a 7 cm x 7.5 cm isoechoic lesion on the left lobe of the liver in abdominal ultrasonography and her intrahepatic bile ducts were dilated at that location. The same lesion was observed to be hypodense at abdominal CT. A mass lesion, 4 cm x 4.5 cm in dimension at the left upper lobe of lung was seen in her chest X-ray. Her hepatic angiography and portal venography was normal. Ultrasonography guided fine needle aspiration biopsy was performed from the lesion in the liver. Histological examination of the aspiration material showed cuticle-like structures between normal hepatocytes suggesting alveolar hydatid cystosis (Figure 1,2,3). Albendazole treatment at a dose of 400 mg/BID was initiated after the establishment of diagnosis and the patient was referred to the gen-

eral surgery department for excision. It was observed during operation that the lesion had also invaded the right lobe intraparenchymally. The operation was terminated because excision of the mass was thought to be impossible with the available surgical apparatus in our hospital and the patient was referred to another surgical facility with a hepatobiliary clinic.

DISCUSSION

Alveolar hydatid cyst is a zoonotic disease caused by *E. multilocularis*. While the egg of the echinococcus is sensitive to heat and dryness, it is resistant to the chemical compounds. It can stay alive for one year at room temperature. The egg of the echinococcus is dissolved by the pancreatic juice in the alkaline medium of the duodenum after oral administration. The oncosphere then transforms into the free form where it penetrates intestinal mucosa, goes to the liver via the portal vein and causes hydatid cyst. If the oncosphere can not penetrate, it goes to the heart and causes hydatid cyst of the liver by passing the pulmonary artery or goes to the other organs by the systemic circulation and causes hydatid cyst there (6). Our case had lung metastasis of the disease but skin (7), cerebral (8), pancreas (9) and even foot and spine (10) metastasis have been reported in the literature. The macroscopic appearance of alveolar echinococcosis resembles a honeycomb or sponge. Microscopic findings reveal small cystic areas surrounded by membranes, granulomatous inflammatory reaction and necrosis (11). The duplication period of the cyst is slow, with symptoms presenting in 5-10 years. It can lead to right hypochondrial pain, nausea, vomiting and jaundice (6). Jiang et.al reported the clinicopathological findings of 70 alveolar hydatid cyst cases in China (12). They revealed that the cases were usu-

ally young and female. While the most common reason for presentation was due to liver mass (with a 56.4% ratio) the second reason was observed to be due to abdominal pain (28.2%). Our case was also a young female patient who presented at our clinic with jaundice. Her HbsAg, Anti Hbc total, Anti Hbe and HBVDNA were found to be positive at laboratory examination. Although hepatitis B virus infection is the most common etiological agent of chronic liver disease in Turkey, the coexistence of chronic hepatitis B virus infection and alveolar hydatid cyst was thought to be coincidental in this case because cholestasis parameters were elevated predominantly and a liver lesion was demonstrated by radiology. Fine needle aspiration of this lesion revealed alveolar hydatid cyst and the patient had lung metastasis of the disease. The diagnosis of *E. multilocularis* infection is dependant on radiological techniques, specific antibodies in serum and histological findings. In one study comparing the radiological methods of USG, CT and MRI, it was concluded that USG is the method of choice in screening but should be completed by CT, which shows more clearly the demarcation lines of characteristic calcifications and MRI which may facilitate the diagnosis in uncertain cases with non-calcified or partially calcified lesions showing the characteristic multivesicular structure, necrotic areas and proximity to vascular structures (13).

Because most of the cyst portors develop an immune response, serological tests can be helpful when they are positive. Lanier compared ELISA, İHA and double diffusion tests for this purpose and concluded that ELISA is more sensitive (14). The cyst was diagnosed by abdominal USG and CT in our case but her *Echinococcus* İHA test was negative. In recent years, the PCR method of detecting minute amounts of parasite DNA and mRNA has been used as a complimentary method to synergise immunodiagnostic techniques (15), but a definite diagnosis is only possible by histopathological evaluation.

The treatment of alveolar hydatid cyst depends on surgical extraction of the parasite and chemotherapy. Transplantation is needed only in cases with chronic liver failure (16). In an open labelled prospective study, the high therapeutic efficacy of both mebendazole and albendazole with similar response rates in the treatment of alveolar echinococcosis was demonstrated (17). Our case was prescribed albendazole therapy and then referred to the surgery department for mass excision.

In conclusion, alveolar hydatid cyst is a fatal infection with poor prognosis which should be diagnosed promptly and be considered in the differential diagnosis of liver mass lesions, even though it is rare.

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