BAOJ Cancer Research & Therapy

Metin MR, et al., BAOJ Cancer Res Ther 2015, 1:1
1: 004

Case report

Alveolar echinococcus: A rare case with atypical localization

Metin MR[1], Aydin H[2], Ates OF[1*], Cetin H[1] and Karaoglanoglu M[1]

[1]*Ankara Ataturk Education and Research Hospital / Radiology Department*

[2]*Onkoloji Education and Research Hospital / Radiology Department*

Abstract

Alveolar echinococcus is a chronic parasitic infection, especially hepatobiliary system is involved, portal and biliary tract invasions are frequent, they have indistinct borders can behave just like malignant masses, may invade adjacent organs or show distant metastases. Liver, lungs, brain, heart and bone involvement of echinococcus were reported but however, cases from aorto-caval region, main abdominal vascular structures and the main portal vein has not been reported up to now. In this case, localization and infiltrative character of an atypical alveolar echinococcus, para-aortacaval echinococcus was presented with MDCT, MR imaging, and CT angiography findings.

Keywords: Alveol; Echinococcus; Aorta-caval-parasitic.

Introduction

Alveolar echinococcus, is a chronic parasitic infestation which is endemic in some countries of Asia, central and western europe, middle east countries [1]. All of the organs especially hepatobiliary systems may be involved, may remain asymptomatic for a long time, then when becomes symptomatic, usually has a poor prognosis [1,2]. When localized in the liver, portal and biliary tract invasions are frequent and is really difficult to make a differential diagnosis from malignant masses as they have indistinct borders, in addition, it can behave just like a malignant mass, may invade adjacent organs or show distant metastases [1-3].

Alveolar echinococcus which is caused by 'Echinococcus multilocularis' is a rare parasitic disease, very rare in contrast to Echinococcus granularis and is endemic in central and Western Europe, China, and in Middle Eastern countries [1]. Its metasestod form is transmitted to humans, rodents are the intermediate hosts of adult form of the parasite in fox and in dog-like animals [1-3]. In humans, these parasites are very random settled, mostly to the liver and manifest as a liver disease. Extrahepatic spread is quite rare, but however in the literature; Lungs, brain, heart and bone involvement of echinococcus were reported but, cases in the aorto-caval region, the aorta, vena cava and the main portal vein had not been reported up to now [2-4].

In this report, localization and infiltrative character of the mass with diagnosis of an atypical alveolar echinococcus was presented. A rare case of echinococcus was reported here with Multidedector (MD) CT, MR imaging, and CT angiography findings. To our experience, para-aortocaval Echinococcus with atypical localization without any hepatic involvement, is the first case in the literature.

Case Report

35 year old male patient with diffuse abdominal pain, occasionally with complaints of high fever and weight loss was referred to a medical center for abdominal ultrasound (US) examination and US stated a presumptive diagnosis of solid mass in the epigastric region, then he was sent to our hospital for further examinations. The patient was admitted to General Surgery Department in our hospital and surgeons found a vague palpable mass on physical examination with high fever. In terms of laboratory results for a complete blood count and biochemistry; Direct biluribin: 0.39, Hgb: 12.8, Hct: 38.1 MCV: 76.8 MCH 25.8, CRP: 2.93, Sedimentation: 54/hour and mild leukocytosis were indicated. Patient was referred to our clinic for abdominal MDCT examination with a preliminary diagnosis of a palpable mass in the epigastrium. Abdominal MDCT examination revealed an approximately 110 x 90 mm sized multilobulated semisolid mass lesion which was located at hepatoduodenal ligament level and bilateral para-aortic, paracaval area, seperated from each other with indistinguishable boundaries , presented an irregular calcific foci, surrounding an approximately 10 cm segments of the abdominal aorta via luminal narrowing and causing irregularities in the arterial wall that might displace Vena cava inferior (VCI) to the right lateral and predict unclear boundaries from VCI, also surrounded portal vein confluence involving hypodense necrotic areas. Celiac trunk, Superior Mesenteric artery, left renal vein and right main renal artery could not be distinguished from the mass lesion with clear boundaries (Figure 1).

***Corresponding author:** Aydin H, Onkoloji Education and Research Hospital /Ankara-Turkey, Tel: 0090-312 5962616;
E-mail: dr.hasanaydin@hotmail.com

Rec Date: July 2, 2015, **Acc Date:** August 3, 2015, **Pub Date**: August 5, 2015.

Citation: Metin MR, Aydin H, Ates OF, Cetin H and Karaoglanoglu M (2015) Alveolar echinococcus: A rare case with atypical localization. BAOJ Cancer Res Ther 1: 004.

Copyright: © 2015 Metin MR, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

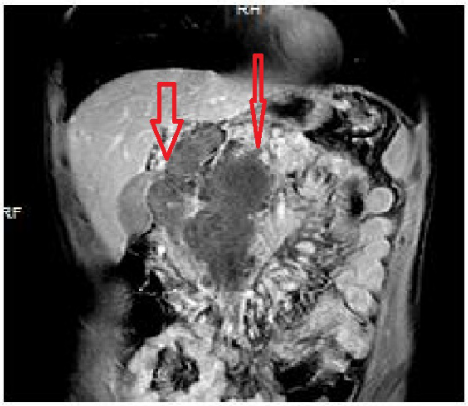

Figure 1: A multilobulated semisolid mass lesion located at hepatoduodenal ligament level,infiltrating bilateral para-aortic and paracaval areas , presenting irregular calcifications and indistinct borders.

Additionally, patient had maximum normal sized-spleen (Long axis of spleen was 135 mm) and an increase in left kidney size (140 mm). Venous collaterals were present at perigastric area and at the level of left renal hilum. There were no solid or cystic lesions in the liver. Patient informations were listed in Table 1.

Dynamic MR imaging of the abdomen was performed due to the suspicious diagnosis of infiltrative mesenchymal sarcomatous malignant mass lesion. In MR Imaging, that was contributing to the MDCT results, it was predicted that the mass also invaded the aorta, VCI, celiac artery, the right renal artery and the left renal vein, regarded a peripheral contrast enhancement in the late venous phase. In addition, the mass regarded significant but partial diffusion restrictions on diffusion weighted MR imaging (DWI) which was mainly observed in DWI and ADC mapping under b value 800s/mm^2 (ADC value: min: 0.25, max: 0.65, mean: 0.40 mm frames/sec) (Figure 2). In the differential diagnosis; A malignant sarcomatous mass or mesenchymal tumor and/or lymphoma was considered, the patient had no known cancer history and biopsy was recommended from the lesion .

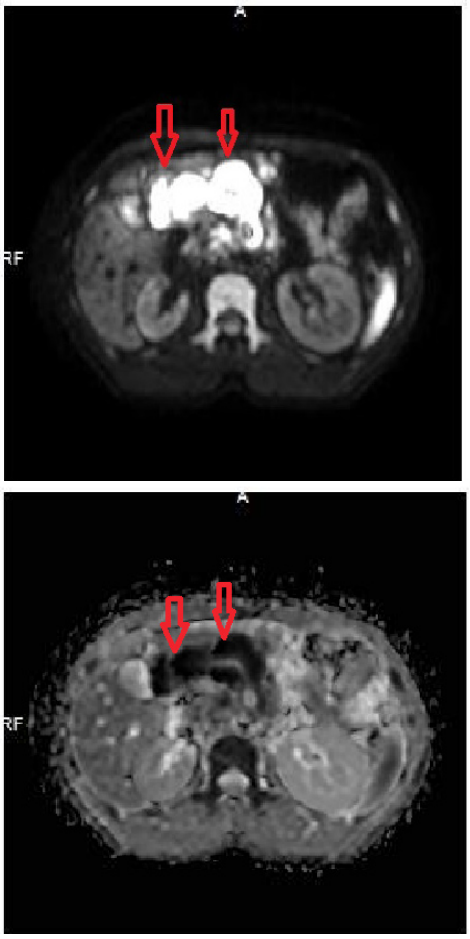

Figure 2a-b: The mass regarded a significant diffusion restrictions on DWI and ADC-maps.

An Interventional radiologist from our department removed a solid fragment from the lesion with a true-cut technique from the necrotic and/or from the diffusion restricted areas, the biopsy

	PATIENT DATAS
Lab.findings.	*Directbiluribin: 0.39, Hgb: 12.8, Hct: 38.1 MCV: 76.8 MCH 25.8, CRP: 2.93, Sedimentation:54/hourandmildleukocytosis.*
Abd.CTresults.	*11 x 9 cm sizedmulti-lobulatedsemisolidmasslesion,located at hepatoduodenalligamentandbilateral para-aortic, paracavalarea, presented an irregularcalcificfoci, surrounding an approximately 10 cm segments of theabdominal aortaleadingtonarrowedlumenandcausingirregularities in thearterialwallalsosurrounded portal veinconfluence. MassalsosurroundedCeliactrunk, SuperiorMesentericartery, leftrenalveinandright main renalartery*
Dynamic MRI findings.	*Themassinvadedthe aorta, VCI, celiacartery, therightrenalarteryandtheleftrenalvein, regarded a peripheralcontrastenhancement in thelatevenousphase.In DWI,themassregardedsignificant but partialdiffusionrestrictionsunder b value 800s/mm2(ADC value: min: 0.25, max: 0.65, mean: 0.40 mm frames/sec).*

Table1: Patient'sLab.andimaginginformations

specimen was sent to clinical pathology and pathologic evaluation proved a diagnosis of a compatible Echinococcus multilocularis enfection with granulomatous inflammation, containing cuticular membranes. With regard to this diagnosis, in order to analyse adjacent arterial structures and to assess the details of aortic invasion, CT angiography was performed, in the Angiography, 50-60% stenosis and vascular irregularities in the aortic-wall were detected (Figure 3).

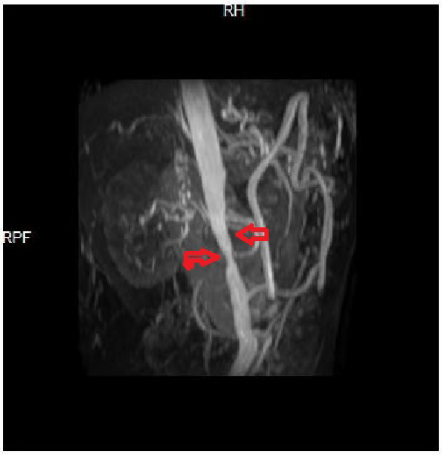

Figure 3: CT angiography revealed 50-60% stenosis in the aortic-wall with irregularities.

Patient was considered to be inoperable and informed about the status of the lesion. 6 months follow-up was recommended and was discharged with advises for the aortic graft implantation but he refused to have the vascular graft operation. Oral albendazole as an anti-infective medical drug was administered in this period and the patient is still being under medical conservative therapy. After 6 months, he will be referred to CT and CT-angiography in order to search the effectivity and outcome of medical treatment. As our hospital was a generalized research hospital, including special oncologists-thorax surgeons and radiologists, the patient will continue to his follow-up and treatment in our hospital.

Discussion

Parasite reaches to the gastrointestinal tract via faecal-oral route, destroys the intestinal wall and localizes through the liver by portal vein or lymphatic routes [2,3]. It forms a granulomatous area by stimulating the host immunity, also triggers the fibrosis and develops in acellular laminar membrane and cellular germinal membrane inside which is surrounded by fluid-filled cysts [1,3,5]. Daughter cysts can involve thousands of protoscolex and every one of these protoscolex is capable of reaching to adult form. Thus, the lesion grows either by the parasite or by secondary to the host immunity [2-4]. In contrary to Echinococcus granularis, the entire lesion can not be limited by the host, It grows faster and its course is infiltrative, as the lesion grows peripherally, central parts of it can be accompanied by necrosis [5].

Alveolar echinococcosis, initially often remains asymptomatic for many years and later becomes symptomatic through the neighbourhood spread or by causing liver failure, or both as a result of hematogenous spread to other organs, clinical symptoms vary according to the organs involved, abdominal pain and cholestatic symptoms are the most common symptoms [6]. Hepatic involvement can lead to complications such as invasion of the biliary ducts, cholangitis, portal hypertension, biliary cirrhosis, lung or brain involvement may cause chest pain, hemoptysis, cough, dyspnea, epilepsy, increased intracranial pressure and neurological deficits [5-7]. Factors determining the prognosis of the disease are namely the dissemination of the disease, causing the systemic or local invasion, If the parasite is not diagnosed early or not treated earlier, the prognosis is generally poor [6-8]. Liver failure and/or cirrhosis, vascular invasion or biliary system invasion, may limit the treatment facilities and can lead to inoperable cases. The World Health Organization Informal Working Group on Echinococcosis published a classification as "PNM" system that leads the evaluation of the parasite in terms of diagnosis, treatment and its performance (7.8)

To our belief, a detailed diagnosis of the disease can be supplied by specific serological tests, radiological imaging and histopathological examination together. For the radiological diagnosis, the first reference method is US; Irregular, multilocular-solid components and areas containing calcifications are the classic sonographic appearance and cholangiocellular cancer plus liver cysts are the other possible masses for differential diagnosis, but rarely can be seen as hyperechoic nodules and in such cases may be confused with hemangioma or metastasis [8,9]. Tomographic views are more useful than sonography due to its better presentation for anatomical appereance, characterization of the lesion and relationship with neighboring structures, also indicate calcification which is a frequent finding of alveolar echinococcus. In post-contrast studies, except for a slight peripheral enhancement, it can be differentiated from solid mass lesions without any obvious enhancement [7-10]. MR imaging is the best method for its higher resolution for characterization of soft tissue, although it is insufficient to show calcification, is the most accurate method of predicting perilesional propagation and invasion, additionally, the relationship of the lesion with biliary tree can be demonstrated by performing MR cholangiography [10].

Primary therapy of the resectable diseases is the radical surgery with administration of benzimidazole derivative drugs, however, curative resection is possible only in a small group of early-stage hepatic diseases, chemotherapy can mostly be applied as the treatment of choice. In extrahepatic manifestations, the treatment is controversial, its response to chemotherapy is often not sufficient [8-10].

In the previous reports, Senturk et al [11] described a case of a hydatid cyst that completely filled the left main pulmonary artery and its distal part without any cardiac involvement by thoracic CT and Endobronchial US. Tercan et al [12] presented a case of hydatic cysts, located in bilateral pulmonary arteries and left ventricular wall by Thoracic CT.

In the presented case, hepatic involvement was not seen in contrast to the classical presentation of alveolar echinococcus. Intra-abdominal, extrahepatic involvement is available. Retroperitoneal spaces were invaded without any involvement of portal confluence level. Considering the propagation paths of the parasite, it can be thought that the disease might have been occured after bowel wall invasion and might spread to the portal vein via direct invasion without forming any liver diseases, Direct spread through the bowel wall invasion can also be considered, however, imaging findings suggestive of bowel wall invasion are not available.

Both clinicians and we were quite surprised due to its presentation without any liver involvement and chronic course of the disease with patients weight loss, encouraged us to determine a diagnosis of malignancy. In addition, we encountered infiltrative imaging findings, with areas peripherally enhanced and diffusion restriction provoked us to misdiagnose this case as a cancer.

Multicystic appearance, areas of calcification and infiltrative pattern of classical findings of alveolar echinococcosis were also present in our patient. In addition, unseldom in alveolar Echinococcus , our patient regarded some areas in which there were obvious limitations of diffusion. There was a limited information of echinococcosis for restricted diffusion in the literature. In a recently published study,it was presented that alveolar echinococcus revealed lower ADC values than the cystic and benign solid lesions of liver [9]. However, this article also reported that the differential diagnosis of echinococcosis was dependent upon the less diffusion restriction of alveolar echinococcus than the malignant lesions. In our case, the average ADC values of the mass was 0.40. In other words, the diffusion properties predicted like the malignant solid lesions did . Abscess development in the necrotic areas of the mass due to superinfection was also considered in the differential diagnosis which was not confirmed with immunohistochemistry.

As a result, alveolar echinococcus is an infiltrating parasitic disease which often presents in the liver. It should be considered in atypical clinical findings without enhancing multicystic , infiltrative liver lesions. A quite rare and atypical alveolar echinococcus case was presented above with para-aortocaval localization without any liver involvement. The diagnosis of alveolar echinococcus can be quite difficult when classic symptoms and imaging findings of the liver is not present.

References

1. Vuitton DA, Zhou H, Bresson-Hadni S, Wang Q, Piarroux M, et al. (2003) Epidemiology of alveolar echinococcosis with particular reference to China and Europe. Parasitology 27: 87-107.
2. Nourrisson C, Mathieu S, Beytout J, Cambon M, Poirier P (2014) Osteolytic bone lesion: alveolar echinococcosis in a patient with vertebral splenectomy. La Revue de medecine interne 35: 399-402.
3. Scherer U, Weinzierl M, Sturm R, Schildberg, Friedrich-Wilhelm, Marlene Z, et al. (1978) Computed Tomography in Hydatid Disease of the Liver: A Report on 13 Cases. J Comput Assist Tomo 2: 612-617.
4. Algros MP, Majo F, Bresson-Hadna S, Majo F, Bresson-Hadni S, et al. (2003) Intra-cerebral alveolar echinococcosis. Infection 31: 63-5.
5. McManus DP, Zhang W, Li J, Bartley PB (2003) Echinococcosis. Lancet 362: 1295-1304.
6. Pawlowski ZS, Eckert J, Vuitton, et al. (2001) Echinococcosis in humans: clinical aspects, diagnosis and treatment. In: Eckert J, Gemmell MA, Meslin FX, Pawlowski ZS, eds. WHO / OIE manual on Echino-coccosis in humans and animals: a public health problem of global concern. Paris, France: WHO / OIE. 20-72.
7. Bresson-Hadna S, Vuitton DA, Bartholomot B, Heyd B, Godart D, et al. (2000) A twenty-year history of alveolar echinococcosis: analysis of a series of 117 Patients from eastern France. Eur J Gastroenterol Hepatol 12: 327-36.
8. Kern P, Wen H, Sato N, Vuitton DA, Gruener B, et al. (2006) WHO classification of alveolar echinococcosis: principles and application. Parasitol Int 55: 283-287.
9. Kantarci M, Bayraktutan U, Karabulut N, Aydinli B, Ogul H, et al. (2012) Alveolar echinococcosis: spectrum of findings at cross-sectional imaging. Radiographics 32: 2053-2070.
10. Bresson-Hadni S, Delabrousse E, Blagosklonov O, Bartholomot B, Koch S, et al. (2006) Imaging aspects and non-surgical interventional treatment in human alveolar echinococcosis. Parasitol Int 55: 267-272.
11. Senturk A , Er M, Karalezli A et al**. (**2015**)** A Case of Pulmonary Artery Hydatid Cyst Observed on Endobronchial Ultrasound. Iranian Journal of Radiology; 12(1): e15995 , DOI: 10.5812/iranjradiol.15995
12. Tercan F; Kacar N, Kilic D. et al. **(**2005) Hydatid Cysts of the Bilateral Pulmonary Arteries and Left Ventricle Wall: Computed Tomography and Magnetic Resonance Imaging Findings. Journal of Computer Assisted Tomography 29: 31-3.