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Case Report

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Diagnosis of Hepatic Alveolar Echinococcosis with Massive Peritoneal Seeding Treated Surgically- A Case Report

Barraco Elisa[1*], Lazarenko Irina[1], Trotta Nicola[1], Pezzullo Martina[2], Lucidi Valerio[3], Mathey Céline[1]

[1]*Department of Nuclear Medicine*

[2]*Department of Radiology*

[3]*Department of Digestive Surgery*

[1,2,3]*Université Libre de Bruxelles (ULB), Hôpital Universitaire de Bruxelles (HUB), CUB Hôpital Erasme, Brussels, Belgium*

[*]Corresponding Author
Barraco Elisa, Department of Nuclear Medicine, Belgium.

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Abstract

The diagnosis of Alveolar Echinococcosis is often a difficult diagnosis due to its low incidence and a long asymptomatic incubation period. We report a case of a 32-year-old man with an uncommon location of Alveolar Echinococcosis.

He presented abdominal pain, conjunctival icterus and dark urine. Abdominal ultrasonography, CT scan and MRI demonstrated the presence of a hepatic lesion and a pararectal mass associated with peritoneal and omental implants. The lesions showed perilesional hypermetabolism on 18F-fluorodeoxiglucose (FDG) positron emission tomography magnetic resonance imaging (PET/MRI). Para-rectal biopsy, serological test, and sample taken during exploratory laparoscopy all suggested Echinococcus multilocularis infection that was later confirmed by PCR test. The patient was treated with Albendazole and underwent curative-intent two-step surgery by right-extended hepatectomy, resection of all peritoneal implants and total mesorectal excision. Thirty months after initial surgery, the patient is doing well and is followed up with MRI and 18F-FDG-PET without morphological signs of recurrence.

This case illustrates the capacity of alveolar echinococcosis to affect different organs by infiltration or metastatic dissemination and highlights the role of ^{18}F-FDG-PET/MRI in diagnosis and disease monitoring.

Keywords: Alveolar Echinococcosis, Echinococcosis, Echinococcus Multilocularis, 18F-Fluorodeoxiglucose Positron Emission Tomography Magnetic Resonance Imaging (FDG-PET/MR).

1. Introduction

Human echinococcosis is an infectious disease caused by larval forms (metacestodes) of Echinococcus (E.) tapeworms found in the small intestine of carnivores. The infection is acquired through the fecal-oral route. Among the recognized species, two are of medical importance – E. granulosus and E. multilocularis – causing respectively cystic echinococcosis (CE) and alveolar echinococcosis (AE) in humans [1].

AE is endemic in some regions of the northern hemisphere, such as Western China, Western Europe, Turkey, Eastern Russia, North America and Japan [2]. Until recently, Belgium was considered a low-risk country for AE. However, in 2008, Hanosset et al. demonstrated a prevalence of AE at up to 60% in red foxes (Vulpes vulpes) in some parts of Southern Belgium [3].

In alveolar echinococcosis, initially, metacestodes of E. multilocularis develop almost exclusively in the liver, predominantly in the right lobe. From the liver, the larvae spread to other organs by contiguous infiltration or distant metastasis formation. Primary extra-hepatic locations of E. multilocularis are rare.

The prognosis is poor with a 10-year mortality approaching 90-100%. Therefore, an early diagnosis and radical surgery followed by anti-infective prophylaxis with benzimidazole (BZM) remains crucial [4].The diagnosis is based on clinical findings, epidemiological data, imaging techniques, histopathology and/or nucleic acid detection, and serology.

Computed tomography (CT) and magnetic resonance imaging

(MRI) provide useful morphologic information for the accurate diagnosis of hepatic alveolar echinococcosis (HAE), typically revealing a heterogeneous mass with central necrosis and peripheral contrast enhancement.

Thus, the European Network for Concerted Surveillance of Alveolar Echinococcosis and the World Health Organization-Informal Working Groups on Echinococcosis (WHO-IWGE) developed a classification based on imaging findings for standardized evaluation of diagnostic and therapeutic measures. It denotes the extension of the parasitic mass in the liver (P), the involvement of neighbouring organs (N), and metastases (M) [5].

In 2003, Kodama [6] defined five types of HAE lesions depending on their morphological characteristics on MRI. This classification illustrates the biological course of the disease. The initial stage is mostly represented by very small cystoid lesions (Type 1 and Type 2). Advancements during the course of untreated disease are often manifested by lesion with a larger central cystic lesion (Type 3), with a continuous transition to regressive stages represented by a sole larger solid component (Type 4) then a large pure cystic lesion (Type 5).

^{18}F-fluorodeoxyglucose- positron emission tomography/computed tomography or magnetic resonance imaging (^{18}F-FDG-PET/CT or PET/MRI) can provide additional information on disease activity [2].HAE is often presented with a peripheral hypermetabolic rim and a central hypometabolic region. This appearance was described by Shen et al. as the « doughnut sign » [7].The increased peripheral FDG uptake is related to inflammation around the parasite, therefore FDG-PET could evaluate the inflammatory response and assess the parasite viability indirectly [8,9].

Different studies have investigated the relationship between FDG-uptake and morphological characteristics of AE lesions on MRI. Two of them [10,11] demonstrated that the majority of lesions composed of small cysts (initial stage) presented a significantly increased FDG uptake at the periphery of the lesions, while all lesions composed of a large cyst without a solid component were completely photopenic on FDG-PET/CT.

The combination of different types of imaging techniques is essential for diagnosis, therapeutic decision-making, follow-up, and decision on BZM treatment withdrawal in patients with AE.

2. Patient History/Investigations

A 32-years-old man presented with right upper quadrant abdominal pain, conjunctival icterus, dark brown urine and a notion of loss of appetite and weight.

The biological work-up showed cytolysis and cholestasis with negative viral serologies (HAV, HBV, HCV, HIV, EBV and CMV).

Successive abdominal ultrasound, CT scan and hepatic MRI revealed a large hypovascular hepatic mass of 12.6 cm with intracystic calcification, occupying most of the right hepatic lobeinfiltrating the liver hilum, causing dilatation of the intrahepatic bile ducts, associated with multiple diffuse peritoneal implants with similar morphological characteristics.

An abdominopelvic MRI was performed to further investigate the peritoneal extension and showed the presence of a para-rectal mass.

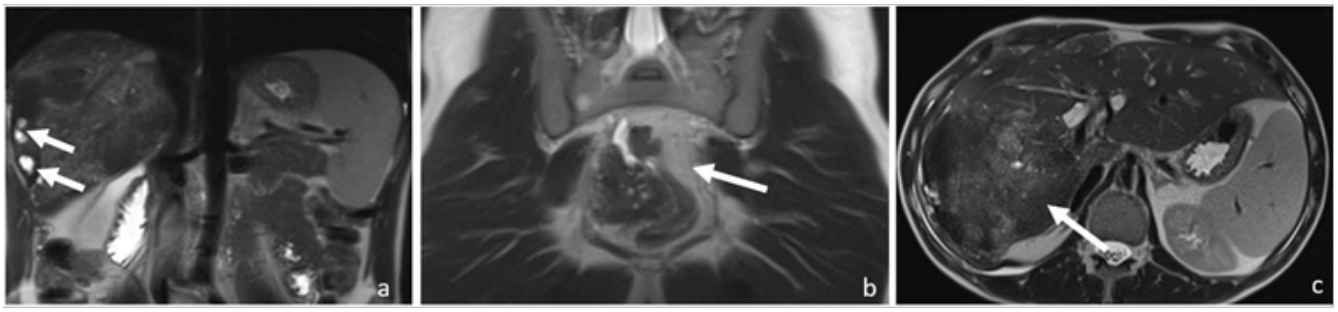

Figure 1: A and B: coronal T2w images at the level of the liver (a) and pelvis (b) showing two masses with similar texture, mostly solid with peripheral cystic components (white arrows); C: axial T2 image showing the heterogeneous mass in the right liver (white arrows) with extracapsular involvement and biliary dilatation secondary to infiltration of biliary confluence.

The diagnosis of echinococcosis was suspected and treatment with Albendazole was started. Moreover, an endoscopic biliary drainage was performed in the context of jaundice and intrahepatic bile ducts dilatation caused by the liver mass.

The antibody test was positive for E. multilocularis and granulosus, but the specific Enzyme-linked immunosorbent assay (ELISA) test was strongly positive for E. multilocularis, so the biologist concluded a high immunological suspicion of AE. The transrectal fine needle biopsy also showed cytological images compatible with AE.

The assessment was completed by an ^{18}F-FDG-PET/MRI investigation, with early (1h post injection) and late (3h post injection) acquisition. It demonstrated that the liver lesion, different implants (peri-hepatic, mesenteric, epiploic, in the right flank and in the right iliac fossa) and especially the right pararectal mass had increased perilesional FDG uptake both on early and late images.

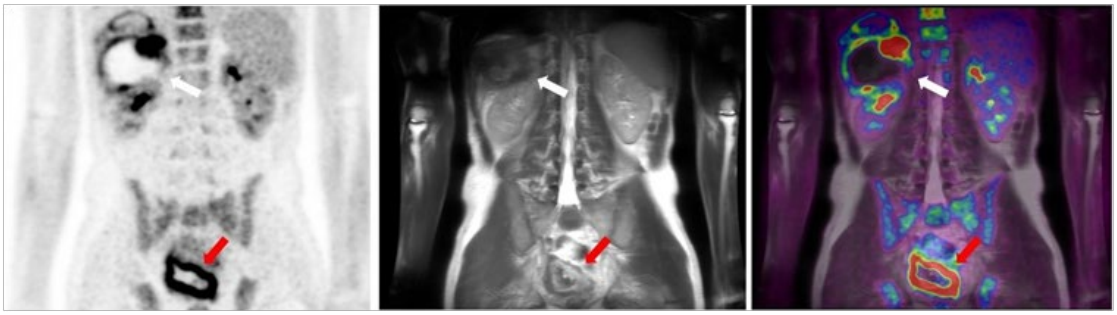

Figure 2: Coronal images of Fluorine-18-fluorodeoxyglucose positron emission tomography/Magnetic resonance imaging (18F-FDG PET/MRI) demonstrating hepatic (white arrows) and mesorectal (red arrows) lesions with a peripheral hypermetabolic rim and a central hypometabolic region.

An exploratory laparoscopy performed to verify the intra-abdominal extension of the parasitic disease revealed the primary right liver mass infiltrating the diaphragm associated with peritoneal nodules in the right paracolic gutter, the rectovesical pouch, the anterior parietal peritoneum and in the great epiploon. A biopsy specimen acquired during the laparoscopy showed fibro-inflammatory, calcified, and cystic material compatible with alveolar echinococcal infection.

3. Treatment

The curative surgery was postponed several times due to the COVID-19 pandemic but above all due to episodes of superinfection marked by pyrexia and increased abdominal pain due to cholangitis with the appearance of air bubbles in the hepatic lesion on CT scan and Enterobacter cloacae in the analysis of biliary fluid.

Subsequently, the patient underwent initially a right extended hepatectomy with hepatojejunal anastomosis, diaphragmatic resection with patching, epiploic and peritoneal resection. Ten weeks after the first surgery, he underwent a total mesorectal excision with coloanal anastomosis and discharge ileostomy that was closed surgically 2 months later.

The PCR analysis performed on the surgical specimens confirmed the presence of echinococcus.

4. Outcome and Follow-Up

At present, thirty months after first surgery, the patient is still on Albendazole, and the follow-up by MRI and 18F-FDG-PET/MRI has not shown any recurrence.

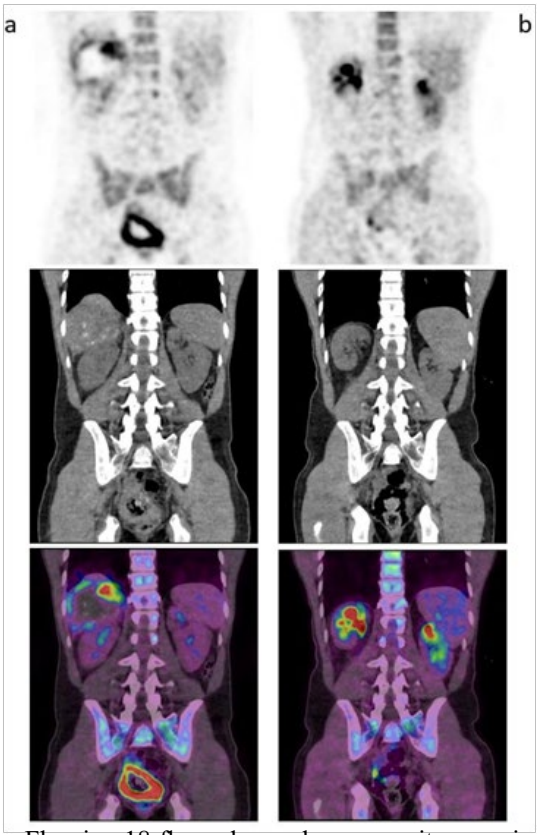

Figure 3A: Coronal images of baseline Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) showing the same hepatic and mesorectal lesions as on MRI; B: Coronal images of 18F-FDG PET/CT thirty months postsurgery without recurrence.

5. Discussion

Echinococcal infection is an important public health concern, especially as there has been an increase in the vulpine populations in Europe and in Belgium in recent years, where the prevalence of alveolar echinococcosis in foxes increases [12].

Without timely diagnosis and therapy, the prognosis of alveolar echinococcosis is dismal and life-threatening. Diagnosis is based on four diagnostic criteria: typical lesions on imaging (ultrasonography, CT, MRI), detection of echinococcus antibodies by a serological test of high sensitivity and confirmed by a serological test of high specificity, histopathology consistent with echinococcosis and PCR.

On ultrasonography, alveolar echinococcosis appears as an expansive process with irregular contours, infiltrating type, with a tendency to extend towards the hepatic hilum. The lesion is often dotted with calcifications and formed by multiple cysts. On CT, it also presents as an infiltrating lesion without clear boundaries, of low density, with little contrast enhancement. MRI provides additional and important pre-surgical information concerning the extension to the vascular, biliary, and extra-hepatic structures [12].These imaging techniques allow to determine the PNM (P = parasitic mass in the liver, N = involvement of neighbouring organs, and M = metastasis) staging, which is important for the therapeutic management of the patient [5].

In our case, the diagnosis of AE was suspected based on imaging features and confirmed by serological and histopathological findings. Indeed, the patient's ultrasonography revealed a right intrahepatic mass causing dilatation of the intrahepatic bile ducts. The CT scan showed a hypovascular infiltrating process of the right hepatic lobe with calcifications, subcapsular cystic patches and paracolic and umbilical peritoneal. The MRI demonstrated a hepatic mass with heterogeneous content, mostly solid, responsible for dilatation of the left intrahepatic bile ducts, an infiltration of the right portal vein branch and presence of implants with similar characteristics in the liver capsule, omentum and mesorectal space.

18F-FDG PET/CT is the imaging technique of choice for follow-up and therapeutic monitoring of patients with AE. Conventional imaging modalities, such as ultrasound or CT, have limited role for follow-up because neither the reduction of lesion size nor the presence of calcifications within the lesions accurately reflects parasitic disease activity [13].As reported in several previous studies, AE lesions often had a peripherical hypermetabolic rim and a central hypometabolic region visible on PET/CT images [7].As in the case of malignant liver tumours, a delayed PET image (3h post-injection) improves the lesion-to-background ratio, provides better visualisation of the lesion, and consequently increases the sensitivity of this technique. FDGPET also represents a sensitive and specific added value in the differential diagnosis of alveolar and cysticechinococcosis by demonstrating a perilesional hypermetabolism in AE, as opposed to EC [13].Thus, FDG-PET/CT allows the monitoring of disease activity indirectly, via the metabolic activity of peri parasitic inflammatory cells.

In our case, we decided to combine PET with MRI as well as a CT acquisition for its better characterisation of lesions and its ability to differentiate the diagnosis of AE and CE, which was fundamental since the serological analysis was positive for both of them. Indeed, MRI shows heterogeneous mass with irregular margins and necrotic center in AE contrary to CE where it shows well defined mass composed by cyst or multicyst [14].

As mentioned above, FDG-PET imaging is also useful for the follow-up of patients treated with Benzimidazole. While it is generally accepted that BZM are parasitostatic, some authors suggest that they could sometimes be parasitocidal (curative) and could therefore be safely discontinued after years of treatment in some patients. However, lack of detectable metabolic activity does not mean parasite death, but indicates suppressed periparasitic inflammatory activity, it would therefore be unwise to discontinue treatment based on the disappearance of metabolic activity alone [15].

That is the main reason why we chose to combine PET with MRI, as MRI is the most adapted technique for morphological characterization (and staging) of parasitic lesions. A study by Caoduro et al. established a strong correlation between MRI and PET findings in AE lesions, namely that the presence of intralesional microcysts correlates with perilesional hypermetabolism. These two aspects (morphological and metabolic) seem to reflect a parasite activity. Therefore, the simultaneous disappearance of microcysts on MRI and of metabolic activity on FDG PET/CT together with negative serological results might help to distinguish inactive from active AE lesions and select patients for which BZM could be discontinued [10].

Patients with echinococcosis are asymptomatic for a long time and symptoms, when they appear, are non-specific (asthenia, weight loss) or related to liver involvement (right hypochondrial pain, icterus), meaning that the diagnosis is often late and the prognosis can be fatal. Treatment should be initiated as soon as the diagnosis is made. In addition to the mortality related to the hepatic and extra-hepatic involvement, complications such as bacterial or mycotic superinfection of the lesions have been reported and can lead to abscess and sepsis, which are potentially fatal [14].Our patient also had two infectious episodes, documented by imaging and bile fluid microbiology, between the time of diagnosis and complete resection of the AE lesions.

The unusual feature of this case is represented by the pararectal location of AE lesion. It is well known that the AE larvae spread from the liver to other organs by infiltration or metastasis formation. Primary extrahepatic localizations of E. multilocularis are rare [4]. From the liver, the parasite can disseminate to the lungs (7-20%), brain (1-3%), bone (1%), heart, peritoneal and mesenteric tissues, and rarely to visceral organs such as the stomach, spleen, kidneys, and adrenal glands. Symptoms depend on the affected organs and the size of the lesion. The diagnosis is based on imaging findings and serology [14].To date, no case of echinococcosis with rectal involvement has been reported in the literature. It is important to remember that a complete imaging work-up should be performed in cases of suspected alveolar echinococcosis to establish the PNM stage and improve the management and survival rate of such patient.

6. Conclusion

Echinococcosis remains a difficult disease to diagnose with a poor prognosis. Imaging techniques, in particular MRI and 18F-FDG-PET, have a fundamental role in the diagnosis and follow-up under treatment of the patients with AE, providing additional information on the parasite activity. This case represents a new illustration that AE has the capacity to invade different organs, including visceral organs, which are rarely affected.

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Conflict of Interest

Author's do not have any conflict of interest to declare.

Author Contributions

E. Barraco participated in the design of the manuscript, PET-CT and PET/MR images analysis and interpretation, clinical data collection, and drafting of the manuscript. I. Lazarenko participated in the design of the manuscript and drafting of the manuscript. N. Trotta participated in PET/CT and PET/MR acquisitions and drafting of the manuscript. M. Pezzullo participated in MR and PET/MR images analysis and interpretation and drafting of the manuscript. V. Lucidi participated in the clinical data collection and drafting of the manuscript. C. Mathey participated in the design of the manuscript, PET-CT and PET/MR images analysis and interpretation and drafting of the manuscript.

Ethical S Tatement

The retrospective analysis of data acquired on the PET/CT (Vereos, Philips Medical Systems, Cleveland, Ohio, USA) and PET/MRI (Signa 3T, GE Healthcare, Milwaukee, Wisconsin, USA) systems has been approved by the Ethics Committee of the institution. Published with the written consent of the patient to use data and images for writing this article.

Data Availability S Tatement

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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