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Case Report: Role of Interventional Radiology in the Management of Patients with Alveolar Echinococcus: Successful Management of Three Cases

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Abstract. Alveolar echinococcus (AE) is an infestation by *Echinococcus multilocularis*. Partial hepatectomy or liver transplantation is the first choice of treatment. However, the disease is usually diagnosed at an unresectable stage. In those cases, invasion of the bile ducts and vessels, and necrosis in the center of the lesion lead to severe complications, such as cholangitis and liver abscesses. Palliative surgery has been reported to not offer advantages in management, and percutaneous and endoscopic interventions have become more prominent in management. In this case series, outcomes in three cases with unresectable AE were reported. In one of the cases, interventional procedures were used to manage the complications after surgery. In the second case, the cystic component was aspirated to decrease the size before the surgery and in the third case, it was used to drain biliary tree and no surgery was done.

INTRODUCTION

Alveolar echinococcus (AE) is an infestation by *Echinococcus multilocularis*. The parasite found in the intestinal mucosa of the definitive host, which is red fox 60% of the time, produces eggs dispersed through feces. The oncospheres in the eggs ingested by the intermediate hosts penetrate the intestinal wall and develop into multilocular cysts. The intermediate hosts are rodents most of the time, but it can occasionally be humans as well.[1] The liver is the most commonly affected organ. Biliary tracts, portal vein, and hepatic veins are usually involved.[2–4] The disease is clinically silent for many years and the parasite grows infiltratively as a malignant tumor.[3,5,6] In untreated or inadequately treated AE, mortality is more than 90% within 10–15 years.[7] Partial hepatectomy or liver transplantation is the first choice of treatment.[8–10] However, because of the silent growth pattern, the disease is usually diagnosed at an unresectable stage. In those cases, invasion of the bile ducts and vessels and necrosis in the center of the lesion lead to severe complications, such as cholangitis, liver abscesses, portal hypertension, Budd–Chiari syndrome, biliary cirrhosis, or superimposed infections affecting morbidity or mortality.[11] Mass resection, formerly recommended to decompress the biliary tree,[12] later was reported not to offer any advantages while the success of chemotherapeutics and percutaneous and endoscopic interventions was demonstrated.[11,13–20]

In 2010, an expert consensus was published in which interventional procedures were favored over palliative surgery. The main indications for percutaneous bile and abscess drainage were stated as the liver abscess formation secondary to infection of necrotic lesions, jaundice due to bile duct obstruction with or without cholangitis, and bilioma formation.[21]

Herein, we present the outcomes in three cases that have been managed and followed up for various indications in our interventional unit.

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CASE 1

A 22-year-old female presented with jaundice and acholic stool. The bilirubin and the liver enzymes were elevated. Ultrasonography (US) revealed cholecystitis and an avascular liver mass. On computed tomography (CT), a 9.9 cm unenhancing necrotic mass with indistinctive borders, occupying the right lobe of the liver, was observed (Figure 1A–C). Accompanying satellite nodules were detected (Figure 2). The left intrahepatic bile ducts were dilated (Figure 2). The core biopsy revealed *Echinococcus alveolaris*. An extended right lobectomy with Roux-Y choledochojejunostomy was performed. After surgery, an abscess was detected in the resection site (Figure 3). A 12 Fr catheter was placed (Figure 4). It was withdrawn after 6 months when the abscess was completely resorbed.

Ten months after the surgery, the patient presented with jaundice. Laboratory tests revealed high bilirubin levels. Percutaneous transhepatic cholangiography (PTC) showed stenosis at the level of hepaticojejunostomy. An 8 Fr internal–external percutaneous transhepatic biliary drainage catheter (PTBDC) reaching distal to the stenosis was inserted (Figure 5) and stenosis was dilated with a balloon of size 8 mm × 2 cm. After repeating PTC on the seventh day, the PTBDC was withdrawn. Six months later when stenosis recurred in the anastomosis site, balloon dilatation was done. A PTBDC was held in place closed. After 3 months, balloon dilatation was repeated and the catheter was withdrawn when no elevation in the bilirubin levels had been detected. The patient was followed up at 3-month intervals in the first year and 6-month intervals in the years later. The patient is still being followed up after 42 months.

CASE 2

A 60-year-old male with complaints of right upper quadrant pain and jaundice was referred to our center with a clinical diagnosis of biliary obstruction and high bilirubin levels. Ultrasonography demonstrated a mass with a diameter of 20 cm in the left lobe involving parts of the segments 5 and 8. Computed tomography examination showed an unenhanced necrotic mass and dilatation in right intrahepatic biliary tracts. Histopathologic diagnosis was that of *E. multilocularis*. A 10 Fr

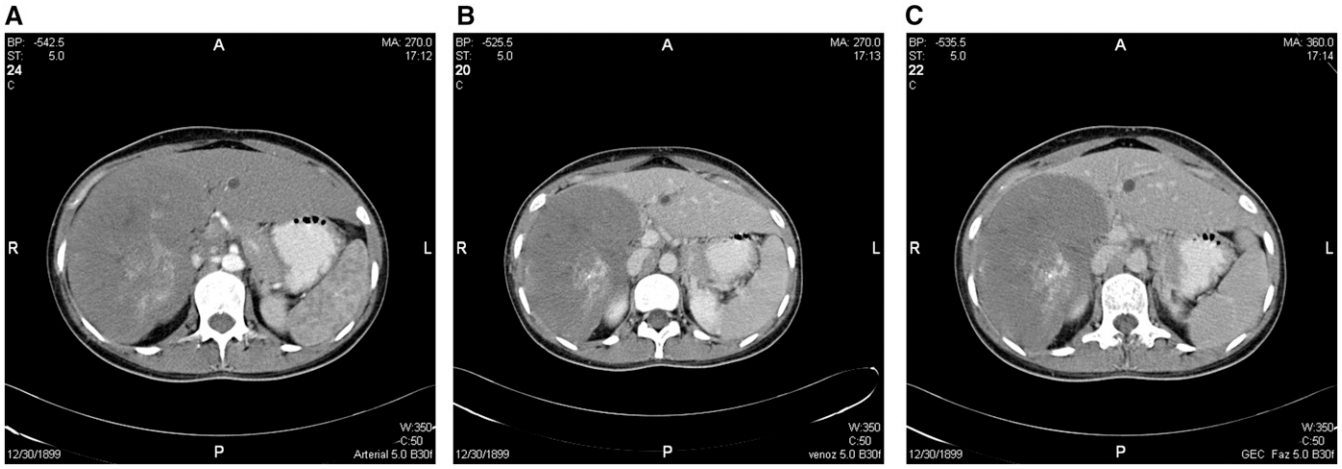

FIGURE 1. (**A**–**C**) Axial plane computed tomograpy images in arterial (**A**), portal (**B**), and venous (**C**) phases show a hypodense mass which is subtle on arterial phase (**A**) and better depicted in portal (**B**) and venous (**C**) phase images because of contrast enhancement of normal liver parenchyma. The lesion itself does not enhance in the later phases and is better appreciated because of increased contrast between lesion and liver parenchyma. Also, low density calcification is noted within the lesion which is typical of *Echinococcosis*.

internal–external PTBDC was placed via biliary tract draining segment 6 into the duodenum. The cystic mass was aspirated in multiple sessions. When the decrease in the size of the mass was confirmed by CT, the patient was operated.

Two months after surgery, the patient, who had been followed up with PTBDC for the stable dilatations in the biliary tracts, presented with jaundice and high bilirubin levels. Computed tomography demonstrated an abscess in the resection site (Figure 6). A 12 Fr catheter was placed (Figure 7). Six months later the abscess was resorbed. However, there was an increase in the dilatation of the biliary tracts. A second catheter with a size of 10 Fr was placed. The patient was followed up for 3 years with regular intervals of 3 months. At the end of 3 years, the patient was admitted with jaundice and high bilirubin levels. Because the biliary tracts were dilated, a third catheter of 8 Fr was inserted and was withdrawn when the dilatation had regressed. The patient has been followed up for 8 years by now.

CASE 3

A 32-year-old male was admitted to an outside center with complaints of abdominal pain, and changes in stool and urine color. The bilirubin and liver enzymes were elevated. Ultrasonography showed an abscess in the right lobe of the liver. In endoscopic retrograde cholangiopancreaticography, a leak

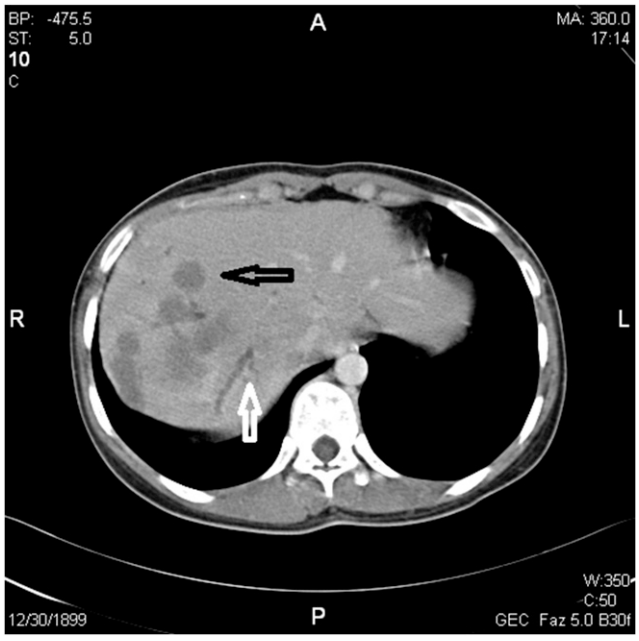

FIGURE 2. Axial plane computed tomography image in venous phase obtained cranial to Figure 1C demonstrates multiple nonenhancing nodules consistent with satellite nodules.

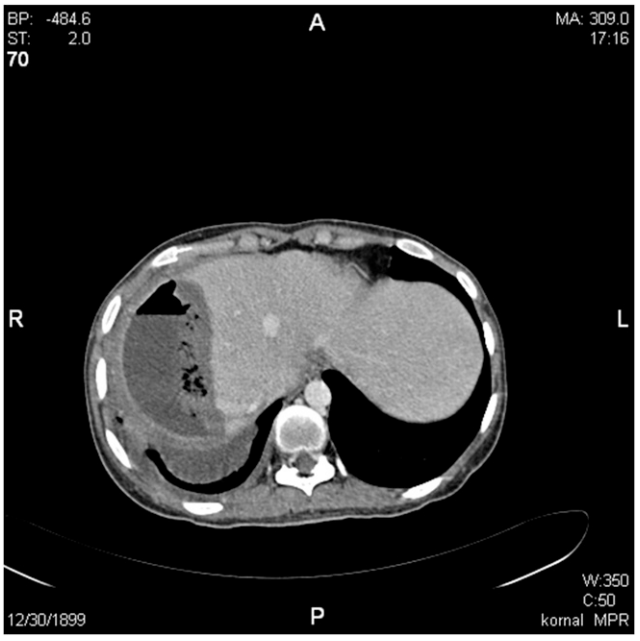

FIGURE 3. Axial plane computed tomography image obtained after surgery demonstrates fluid collection at the site of surgery, and air within the collection indicates infection.

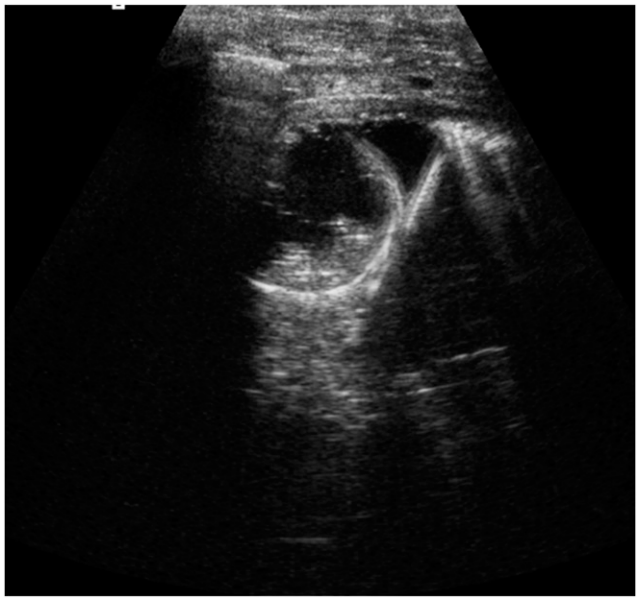

FIGURE 4. Axial plane ultrasonography (US) image obtained when the drainage catheter is inserted and confirmed with US that the catheter is within the collection.

FIGURE 6. Axial plane computed tomography image obtained after surgery shows fluid collection with water density at the resection site. Catheter that was placed in the biliary tract is appreciated as a high-density image passing through hilum.

was detected in the intrahepatic biliary tract and nasobiliary drainage was placed. Biopsy revealed *Echinococcus alveolaris*. The patient was referred to our clinic. Computed tomography showed several loculated abscess collections with internal calcifications, the greatest of which measured 5 cm within the right hepatic lobe and caudate lobe.

Fluid collections in the right lobe and in the caudate lobe were drained with 12 Fr and 8 Fr catheters, respectively. An internal–external PTBDC of 10 Fr was inserted in the right intrahepatic biliary tracts (Figure 8A). After 1 month, the bilirubin levels were within the normal limits and the follow-up CT confirmed that the collections were resorbed and dilatations in the intrahepatic biliary tracts of the right lobe were stable. The patient was followed up with PTBDC with 3-month intervals. After 7 years of follow-up, CT showed residual cavitary lesion in the right hepatic lobe. The biliary tracts were unchanged. The patient is still being followed up with PTBDC (Figure 8B).

DISCUSSION

Diagnosis of AE includes epidemiological data, imaging, and serology. Diagnosis of the infestation can be a challenge, leading to delays in treatment. In a case series, misdiagnosis was reported to lead a mean delay time of 50 months.[22] The primary manifestations have been given as epigastric pain and obstructive jaundice.[23] Similarly, in the current report, all of the patients presented with symptoms of cholestasis such as jaundice, acholic stool, changes in urine color, and pain as a result of biliary involvement.

In cases 1 and 2, radiologic imaging demonstrated an avascular necrotic mass with internal calcifications and in case 3 cystic masses with necrosis were described, consistent with the literature.[3,24,25] Imaging-guided core or fine needle biopsy is the gold standard to confirm the diagnosis of the hepatic masses.[26,27] The major drawbacks of these techniques are inadequate material because of massive necrosis and fibrosis and the risk of seeding.[22,26] In all of our

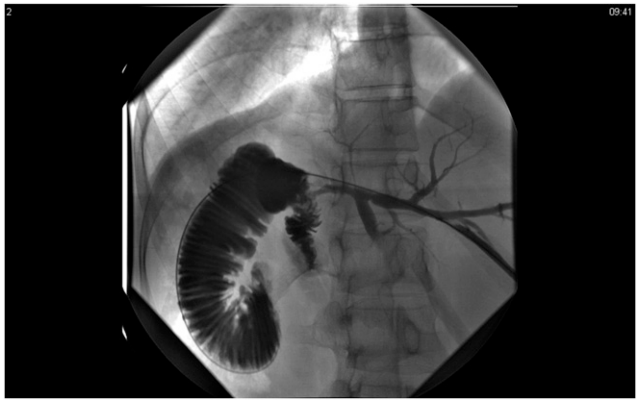

FIGURE 5. Flouroscopy image obtained after the catheter was placed distal to the hepaticojejunostomy anastomosis. Ten days later, stenosis at the site of anastomosis was dilated with a cutting balloon.

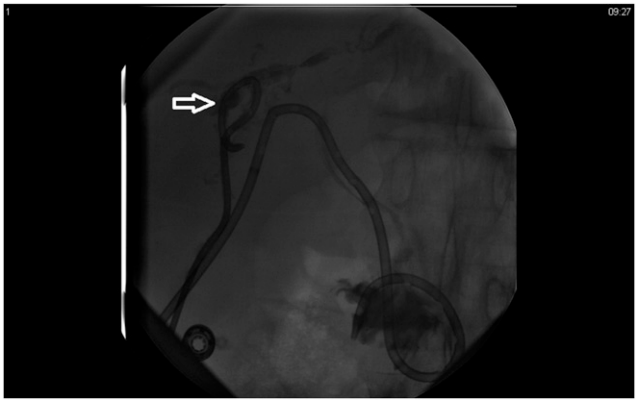

FIGURE 7. Flouroscopy image shows a catheter in the abscess formation (arrow), the catheter seen on the medial aspect is the biliary catheter draining biliary tract of segment 6 and passes into the duodenum.

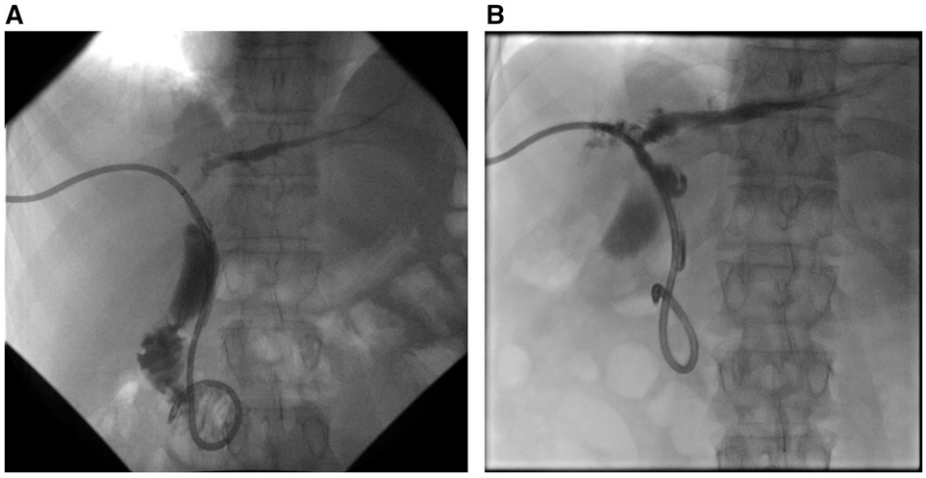

FIGURE 8. (**A**, **B**) Flouroscopy image (**A**) demonstrates biliary catheter passing through the duodenum draining right lobe biliary tract. (**B**) Flouroscopy image obtained after 7 years during routine follow-up demonstrates the catheter in place and stable dilatation in the biliary tract.

patients, the final diagnosis was confirmed by image-guided core biopsy without any complications.

Radiologic interventions are particularly useful for the treatment of infections in the necrotic portion of the mass and cholangitis. The percutaneous abscess drainage significantly contributes to patients' status.[15] Biliary complications are mainly due to the invasion of the biliary tract or severe inflammation and fibrosis.[21] They were reported to occur in 1/3 of the patients and survival was 3 years after the onset of complications, whereas in a more recent study the survival was 8.8 years.[28,29]

Percutaneous transhepatic biliary drainage catheter is particularly useful in hilar involvement. The catheter is placed either in the dilated bile duct or can be extended distal to the obstruction site as an external–internal biliary drainage. The first case treated with PTBDC was in 1994.[30] Then, PTBDC was reported to avoid palliative surgery or be a bridge to curative surgical treatment in 13% of patients.[15]

Despite the fact that interventional procedures are used palliatively, there is one case presenting a patient with an unresectable AE followed up with PTBDC. At the end of 20 months, the lesion was found out to be resolved.[31]

In these three cases, interventional procedures were performed for several different indications. In case 1, they were used in the management of complications after surgery which included abscess drainage and balloon dilatation of anastomotic stenosis. In case 2, the drainage of cystic component preoperatively reduced the cavity size and has demonstrated an example of how interventional procedures may be a bridge for the surgery. After surgery, biliary dilatations were managed by PTBDC. In case 3, an irresectable AE was managed by drainage instead of palliative surgery. However, in all three of the patients, interventions had to be performed repeatedly, if not permanently, because of iterative recurrences. Long-term regular follow-up and regular exchanges were required.

In conclusion, radiological interventions are palliative but efficient methods, particularly in draining necrotic parts and managing obstructions in biliary tree and complications after surgery. However, there is no sufficient data to recommend a protocol that would prevent recurrence. Therefore, patients should be managed individually and followed up cautiously which makes patient compliance essential.

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