CASE REPORT

Uncommon cause of acute encephalopathy in liver cirrhosis

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SUMMARY

A 49-year-old woman with a medical history of alcoholic cirrhosis status post-transjugular intrahepatic portosystemic shunt (post-TIPS) in 2012, and ongoing alcohol abuse, presented to the hospital, with haematuria. CT intravenous pyelogram (IVP) was normal except for 'a large intrahepatic cystic mass adjacent to the TIPS, causing intrahepatic biliary duct dilation'. The patient also presented with acute encephalopathy, jaundice, right upper quadrant abdominal pain and hyperbilirubinaemia (total bilirubin of 8.1 mg/dL with direct bilirubin of 3.0 mg/dL). She remained encephalopathic despite adequate treatment for alcohol withdrawal, hepatic encephalopathy and enterococcus urinary tract infection. MRI of the abdomen later confirmed presence of an obstructing biloma. The biloma, drained by CT-guided percutaneous drains, demonstrated an *Escherichia coli* and ESBL *Klebsiella* infection. The patient's encephalopathy completely resolved after treatment of the infected biloma. With adequate drainage, her hyperbilirubinaemia resolved to her post-TIPS baseline (total bilirubin of 3.7 mg/dL with direct bilirubin of 3.3 mg/dL).

BACKGROUND

Bilomas, extrabiliary encapsulated collections of bile, are reported to occur as a result of biliary surgery, endoscopic retrograde cholangiopancreatography (ERCP), laparoscopic cholecystectomy, trauma and spontaneously in association with certain conditions, primarily choledocholithiasis.^{1–3}

We describe an uncommon cause of biloma, transjugular intrahepatic portosystemic shunt (TIPS), which, to the best of our knowledge, has not yet been described as a cause of biloma in the literature (in the absence of liver infarction). In addition, acute encephalopathy has not previously been described to occur secondary to a partially obstructing infected biloma. Acute encephalopathy is a common problem with many different aetiologies in the differential diagnosis. An obstructing and/or infected biloma should be considered as a possible cause of acute encephalopathy and infection, especially in a patient with liver disease, where it may be under-recognised and attributed to hepatic encephalopathy or other causes.

CASE PRESENTATION

A 49-year-old woman with a medical history of alcoholic cirrhosis status post-TIPS in 2012, with ongoing alcohol abuse (20 years), initially presented to the hospital with gross haematuria. She was also

noted to have jaundice, right upper quadrant abdominal pain and elevated total bilirubin of 8.1 mg/dL with direct bilirubin of 3.0 mg/dL. No ascites was present on examination. Urology was consulted and the patient was adequately treated for haematuria with bladder irrigation. CT intravenous pyelogram (IVP) was normal except for a 'large intrahepatic cystic mass adjacent to the TIPS, causing intrahepatic biliary duct dilation' (figure 1). The patient was placed on a protocol for alcohol withdrawal given her long-standing history of alcohol abuse.

The patient developed progressively worsening confusion and agitation initially thought to be secondary to alcohol withdrawal, and was placed on an Ativan infusion for 9 days. She remained severely encephalopathic despite adequate treatment for alcohol withdrawal. There was also concern that her encephalopathy was caused by hepatic encephalopathy, but it continued to worsen despite adequate treatment with lactulose and rifaximin (with normal ammonia level of 35 µmol/L). Other causes of acute encephalopathy were pursued. Re-evaluation for infectious aetiologies demonstrated a new enterococcus urinary tract infection on urine culture (hospital day 15), but encephalopathy persisted despite appropriate treatment with amoxicillin (according to urine culture and antibiotic sensitivity testing). After this was treated, the patient developed a fever up to 102°F with chills.

INVESTIGATIONS

Initial evaluation for her acute encephalopathy was negative: chest X-ray was performed, which was normal, without evidence of pneumonia. Numerous sets of blood cultures demonstrated no growth. Head CT was normal. Repeat urine culture showed resolution of previous enterococcus urinary tract infection after treatment.

Ultrasound was performed to evaluate TIPS flow given the patient's hyperbilirubinaemia, and this raised concern for possible partial biliary obstruction; MRI confirmed presence of a partially obstructing biloma (hospital day 30). ERCP was performed to attempt internal drainage of the biloma, but was unsuccessful at decompressing the biloma. The biloma was subsequently drained by placement of two CT-guided percutaneous drains. Biloma fluid culture and analysis demonstrated infection (*Escherichia coli* and ESBL *Klebsiella*) with an elevated white cell count of 34 mm³ with 66% neutrophils. Percutaneous cholangiogram was subsequently performed by interventional

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Figure 1 CT of the abdomen/pelvis after transjugular intrahepatic portosystemic shunt (TIPS) procedure with biloma present adjacent to TIPS.

radiology, which demonstrated communication of the biloma with the biliary tree.

CT of the abdomen/pelvis that had been performed shortly after the TIPS procedure (2012) demonstrated a small biloma, which had not been recognised initially but was now visualised to be present on re-evaluation of this imaging. The biloma was not present on similar imaging performed before the TIPS procedure.

Prior to drainage of her biloma, the patient's Child-Pugh score (Child class C) was 13. This score decreased (improved) to 11 after drainage of the biloma. The only scoring component that changed was an improvement of her encephalopathy, but otherwise all other scoring components were similar before and after biloma drainage: her total bilirubin remained greater than 3 mg/dL (although it improved from 8.1 mg/dL back down to her baseline value of 3.7 mg/dL), her albumin remained <2.8 g/dL, her international normalised ratio (INR) remained greater than 2.2, and she continued to have absence of ascites s/p TIPS clinically and on CT of the abdomen/pelvis.

DIFFERENTIAL DIAGNOSIS

Acute encephalopathy can be caused by a large number of metabolic, infectious and neurological aetiologies. In a patient with liver cirrhosis and ongoing alcohol abuse, common aetiologies include alcohol withdrawal, hepatic encephalopathy and routine infections (ie, aspiration pneumonia is common in patients with ongoing alcohol abuse, urinary tract infections are common in women and spontaneous bacterial peritonitis (SBP) can occur in patients with cirrhosis who have ascites). This patient did not have improvement of her acute encephalopathy despite adequate treatment for alcohol withdrawal, and had no other signs nor symptoms to suggest alcohol withdrawal was a persistent concern (ie, no hypertension, no tachycardia and no tremor). Furthermore, she did not have improvement despite adequate treatment for hepatic encephalopathy with both rifaximin and lactulose. She was having frequent bowel movements with lactulose and had a normal ammonia level of 35 μ mol/L. In addition, after drainage of the biloma, her hyperbilirubinaemia immediately improved to her baseline levels (total bilirubin/direct bilirubin was 8.1/3 mg/dL on admission and improved to her baseline level of 3.7/3.3 mg/dL after drainage), but her encephalopathy did not improve for another 5 days, until she was treated with antibiotics after culture results returned. Child-Pugh scoring was

similar before and after biloma drainage, with the exception of improvement of her encephalopathy (which improved the score from 13 to 11). Thus, improvement of her hyperbilirubinaemia (relief of partial biliary obstruction) was not felt to have improved her encephalopathy. She was found to have a new urinary tract infection but her encephalopathy did not improve with adequate treatment and resolution of this. She did not have ascites on examination nor on CT of the abdomen and pelvis, so there was no concern for SBP. There was no pneumonia on several chest X-rays, no bacteraemia on blood cultures and no metabolic aetiology for her encephalopathy on bloodwork.

TREATMENT

When this patient was treated for biloma infection with cefepime for 7 days and drainage, her acute encephalopathy completely resolved. With adequate drainage of her partial biliary obstruction, her hyperbilirubinaemia also improved back to her baseline level after her TIPS (total bilirubin of 3.7 mg/dL and direct bilirubin of 3.3 mg/dL), but this alone did not improve her encephalopathy until antibiotics were administered.

Sclerosing therapy of the biloma after treatment of the infection was not an option since it communicated with the biliary system as confirmed on percutaneous cholangiogram.

OUTCOME AND FOLLOW-UP

According to hepatology and interventional radiology, this patient would likely need life-long percutaneous drainage of the biloma since she was not felt to be a good surgical candidate for a Roux-en-Y procedure (hepatobiliary surgery felt she had a high mortality risk since she would not have viable liver after resection and her Model for End-stage Liver Disease (MELD) score was high at 21). Hepatology determined that she would need to be sober from alcohol for at least 3–6 months to be considered for liver transplantation, but there were other psychosocial factors that made it less likely she would be a good transplant candidate. She had some problems with leaks around the biliary drains and ultimately developed progressively worsening decompensated liver cirrhosis over time and opted to pursue comfort care. She died 2 months later, but after having had an opportunity to spend additional time with family and friends.

DISCUSSION

Coined by Dr Gouda in 1979, A 'biloma' is a rare abnormal accumulation of intrahepatic or extrahepatic bile caused by traumatic or spontaneous rupture of the biliary tree. In the literature, bilomas have been found to be encapsulated and caused by surgery, percutaneous transhepatic cholangiography (PTC), percutaneous transhepatic biliary damage, abdominal trauma and spontaneous rupture of the biliary tree primarily associated with choledocholithiasis (16 cases).^{1–3} On CT of the abdomen/pelvis, shortly after this patient's TIPS procedure, a small biloma had developed that was not present prior to the procedure. After gradual enlargement of the biloma, it caused partial biliary

Patient's perspective

'I do not remember the past couple of weeks. I am glad you figured out what was going on. I just want to get out of here (the hospital)'.

Learning points

- ▶ Common causes of acute encephalopathy in patients with liver cirrhosis include hepatic encephalopathy from hyperammonaemia, alcohol withdrawal in the setting of alcohol abuse and common infections (ie, urinary tract infections and aspiration pneumonia).
- ▶ A rare, but important, cause of acute encephalopathy in patients with liver or biliary problems and/or procedures is an infected biloma.
- ▶ Bilomas can cause obstruction of the biliary system with hyperbilirubinaemia.
- ▶ When an infected, obstructing biloma is adequately treated with antibiotics and is adequately drained, acute encephalopathy and direct hyperbilirubinaemia improve.

obstruction and became infected, causing acute encephalopathy. In the past 37 years since bilomas were initially described, there have been approximately 30 published case reports demonstrating bilomas related to a variety of the aforementioned causes, but to our knowledge, this is the only case reported to occur after TIPS (without liver infarction) and only the third case of

an infected biloma.^{2 4} In the other two reported cases of infected biloma, the patients presented with septic shock and a very large liver infarct after liver transplantation, whereas our patient had a much more subtle presentation with acute encephalopathy that could easily be overlooked.² This emphasises the need to consider this as an aetiology of acute encephalopathy and infection, especially in patients with liver problems; this may be under-recognised and underdiagnosed.

Competing interests None declared.

Patient consent Obtained.

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