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A Local Experience in the Management of Recurrent Pyogenic Cholangitis (Oriental Cholangitis)

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Objective: Recurrent pyogenic cholangitis (RPC), though endemic in most parts of Southeast Asia, is only occasionally seen in Saudi Arabia among expatriate workers from those areas. We present our experience in the management of RPC with a view to raise the awareness of this condition among surgeons and physicians, practicing in this area.

Method: This is a retrospective study conducted between 1994 – 1995 on five patients admitted with RPC to Riyadh medical Complex (RMC). Presenting features, investigations and management for all patients were revised.

Results: All patients presented with abdominal pain, fever, rigors and jaundice. Ultrasonography and endoscopic retrograde cholangiopancreatogram (ERCP) were the main tools of investigation. Intrahepatic and extrahepatic calculi were present in 4 patients whereas one patient had extrahepatic calculi. ERCP successfully removed all stones from the biliary tree in 3 patients. Two patients required surgery for complete removal of stones from the bile ducts.

Conclusion: Patients of RPC are seriously ill on presentation and need aggressive resuscitation and early decompression of biliary tree. ERCP plays a major role in their management. Surgery may be needed in selected patients.

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Recurrent pyogenic cholangitis (RPC), oriental cholangitis, cholangiohepatitis, infestational cholangitis are different names for one clinical syndrome[1,2]. It is endemic in most parts of Southeast Asia, although sporadic reports have appeared from western countries among immigrants from endemic areas[3]. The features of the disease includes bouts of cholangitis, pigment stone formation within intrahepatic and extrahepatic bile ducts, varying combinations of biliary duct stricture development and biliary duct dilatation. Biliary cirrhosis, portal hypertension, bleeding esophageal varices and cholangiocarcinoma are terminal manifestations

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of chronic cases[4]. The disease more commonly affects left hepatic duct and left lobe liver[5]. The etiology of this disease remains obscure. However, the most likely cause is the establishment of bowel organisms in the liver and biliary tract of a compromised host[6]. Association of the diseases with low protein diet and Clonorchis or other parasitic infestations has been documented[7]. Prompt diagnosis is essential, as untreated exacerbation can be fatal[8]. There is a large expatriate population in Saudi Arabia, many coming from endemic areas, this should alert physicians and surgeons in our region to consider this differential diagnosis. We describe here our experience in managing five cases of recurrent pyogenic cholangitis at Riyadh Medical Complex over 2 years (1994-1995). The aim of this presentation is to discuss the presenting features, the role of different diagnostic imaging techniques, and especially endoscopic retrograde cholangiopancreatogram (ERCP) and surgery in the management of this disease.

CASE 1

Thirty five year-old Indonesian woman presented with one-month history of epigastric pain, fever, rigors, itching and jaundice. The patient was afebrile at the time of admission, deeply jaundiced with a tender hepatomegaly. Blood investigations showed anemia, leucocytosis, elevated liver enzymes, normal serum amylase, and negative hepatitis profile. Ultrasound (U/S) of abdomen showed stones in intrahepatic ducts and ductal dilatation. She was initially treated with IV Fluids, 2 units of packed RBC and antibiotics (cefuroxime, Metronidazole). CT scan showed grossly enlarged intrahepatic biliary radicals with multiple stones. Gallbladder was not visualized and common bile duct was clear of stones. Next day ERCP was performed which showed choledochoduodenal fistula, dilated common bile duct (CBD), a stenosed common hepatic duct (CHD) with multiple areas of intrahepatic dilatations and stricture formation and stones (Fig1). Partial extraction of the stones was done. Attempt to insert a nasobiliary drain failed. The patient showed some improvement. Bile culture showed E.Coli and Proteus species, which were sensitive to antibiotics. ERCP was repeated few days later and more stones were removed however a few larger stones were left in the left intrahepatic ducts. Nasobiliary drain was inserted. Following that the patient underwent a few sessions of extracorporeal shock wave lithotripsy (ESWL) using the nasobiliary drain as a guide. At a third session of ERCP, all stones were removed. The patient was discharged after 6 weeks with clear biliary system and a normal liver function test. She was asymptomatic with a normal liver function test when seen in the clinic six weeks later. Subsequently, she failed to appear for follow-up.

Figure 1. ERCP showing choledochoduodenal fistula, dilated CBD, stenosed CHD with multiple areas of intrahepatic dilatations and stricture formation and stones.

CASE 2:

Figure 2. ERCP showing hugely
Dilated CBD above an impacted stone

Twenty two years-old Indonesian female presented with severe epigastric pain, fever, vomiting, and jaundice. On examination she was febrile (39 C), confused, jaundiced, and dehydrated. She had upper abdominal tenderness and guarding. On admission her Hb was 11.9 gm/dl, WBCs 8100/ dl with elevated liver enzymes and bilirubin. An upper abdominal ultrasound showed a dilated CBD and intrahepatic bile ducts with multiple stones and an empty gallbladder. She received IV fluids and IV Cefuroxime and Metronidazole. Later, she developed hypotension (BP 70/40) and leucocytosis (20,500/dL). Following resuscitation with IV fluids, an urgent ERCP showed hugely dilated C.B.D. (3.3cm) above an impacted stone (Fig 2). Papillotomy was done and the stone was extracted. A nasobiliary drain was left in the C.B.D. The patient showed marked improvement. Blood culture and pus from C.B.D. grew E. Coli and enterococci species. A week later ERCP was repeated and the rest of extrahepatic and intrahepatic ducts were cleared completely of stones. (Fig 3) The patient fully recovered and was discharged from hospital. She failed to attend the follow-up-clinic.

Figure 3. ERCP showing clear extra and intrahepatic ducts from stones.

CASE 3

Twenty seven years-old Indonesian woman admitted with right upper quadrant pain, fever,

vomiting, and jaundice of one week duration. On examination she was dehydrated, febrile and deeply jaundiced with a palpable liver. Blood tests revealed anemia, leucocytosis, and mild elevation of liver enzymes and bilirubin. An abdominal ultrasound (U/S) showed multiple stones in a dilated CBD (2.3 cm) and intrahepatic ducts. The gallbladder was contracted and echo-free.

Following conservative management with IV fluids and antibiotics, she underwent ERCP which showed choledochoduodenal fistula and marked dilatation of CBD, CHD and intrahepatic ducts with multiple big stones (Fig 4). She underwent extension papillotomy and insertion of a nasobiliary drain. The patient improved significantly after nasobiliary drain. Bile grew E. Coli and group D Streptococci which were sensitive to Vancomycin, and Piperacillin. A week later she underwent repeat ERCP and Laser lithotripsy. Only partial fragmentation and extraction of stones were possible at this attempt. Nasobiliary drain was left in situ.Following that she underwent few ESWL sessions. A third, ERCP failed to clear stones from bile ducts. She was operated, and open cholecystectomy, and choledochoduodenostomy and clearance of all stones was achieved. She progressed well except for wound infection. She was discharged after complete recovery. Unfortunately, the patient failed to appear for follow-up.

Figure 4. ERCP showing choledochoduodenal fistula and marked dilatation of CBD, CHD and intrahepatic duets with multiple big stones.

CASE 4

Thirty two years-old Indonesian female presented with epigastric pain and vomiting of two days duration. On examination she was pale, dehydrated, jaundiced with tenderness and guarding in epigastric region. Her blood tests showed anemia, leucocytosis, elevated serum amylase (4237 U/L), elevated liver enzymes and bilirubin. Ultrasound examination showed swollen pancreas, hepatomegaly, and intrahepatic bile ducts dilatations with stones. She was treated with IV fluids and antibiotics. Next day, the ERCP showed a patulous ampula, smooth stenosis of distal CBD, dilated CHD and intrahepatic bile ducts, with intrahepatic duct stones (Fig 5). Papillotomy was done with reasonable drainage. A week later when ERCP was repeated only partial extraction of stones from intrahepatic bile ducts was possible. CT scan of the upper abdomen showed strictures and dilatations with multiple stones in left main duct, and a single stone in the right main hepatic duct . She underwent cholecystectomy, CBD exploration with the help of choledochosocope, and resection of involved liver segment. A complete clearance of ductal stones was achieved. One week later T-tube cholangiography showed dilated intrahepatic ducts with strictures but no stones. The patient did well and the T-tube was removed. She was

asymptomatic after discharge when seen on follow-up after 4 weeks.

Figure 5. ERCP showing patulous ampula, smooth stenosis of distal CBD, dilated CHD and intrahepatic bile duets, with intrahepatic duet stones.

CASE 5

Thirty seven year-old Filipino male presented with epigastric pain, fever, chills, and jaundice of 3 days duration. He was having intermittent abdominal pain for the last one year. On examination he was febrile, jaundiced, dehydrated and tender in the epigastric region. Blood tests showed leucocytosis, elevated liver enzymes and bilirubin. Hepatitis profile was negative. Abdominal ultrasound showed multiple gall stones and CBD stones. He was treated with IV fluids and antibiotics (Cefuroxime and Metronidazole).

Two days later he underwent ERCP which showed oesophageal varices (Grade 1), gastritis, duodenitis, periampullary diverticulum, choledocholithiasis, and suppurative cholangitis. Papillotomy and complete CBD stone extraction was done. Post ERCP the patient developed meleana with a drop in haemoglobin. Repeat ERCP showed bleeding point in periampullary region which was coagulated. His clinical improvement was not satisfactory. A repeated ultrasound showed gall bladder stones, clear biliary ducts and a liver abscess in right lobe which was aspirated under ultrasound guidance. Following which he improved significantly and was discharged. Six weeks later, laparoscopic cholecystectomy was successfully done. Patient remained asymptomatic since then.

DISCUSSION

Recurrent pyogenic cholangitis (RPC) is an acquired disease which characteristically presents with recurrent bouts of abdominal pain, fever, chills and jaundice. Patients are usually in their 3rd- 4th decades, from lower socioeconomic class and both sexes are equally affected. It is important to differentiate between RPC, where biliary infection is the primary pathology and leads to the changes in the bile ducts which may lead pigment stones and cholangitis which is secondary to gall stones. Our patients were all young expatriates (27-37 years) from southeast Asia, presenting with the characteristics features which made the diagnosis easier. The only difference in the patient's profile from endemic area was the predominance of females (4:1). This could be due to larger number of female domestic workers coming from these endemic areas.

Modern imaging helps in the diagnosis, evaluation of the extent of disease and planning the management of RPC. Ultrasound examination is the primary investigation. Diagnosis can be successfully established in 97% of patient having biliary pathology[9]. Ultrasound may also

detect liver parenchymal disease. Liver abscess developed as a complication of RPC in one of our patients which was diagnosed on follow-up ultrasound examination. However, ultrasound examination has some limitations in these patients. The presence of pneumobilia may hamper the examination and soft pigment stones which are usually iso-echoic and can be missed by ultrasound[8] Endoscopic retrograde cholangiopancreatogram (ERCP) plays a major role in the management of RPC. Radiological features on ERCP includes extrahepatic and intrahepatic bile ducts dilatations showing the arrow head configuration and lack of branching in intrahepatic ducts[8], irregular contour and stricture formation in bile ducts[10]. These radiological features were demonstrated on ERCP in all our patients.

Management of this disease involves control of acute attack, complete removal of extrahepatic and intrahepatic stones and establishment of satisfactory drainage of biliary tree. A multidisciplinary team approach involving surgeons and gastroenterologists is required to provide an adequate management.

An excellent outcome in patients with only extrahepatic calculi can be achieved, however, the management of patient with intrahepatic calculi is more problematic[3]. An ERCP may fail to remove intrahepatic stones in up to 50% of patients[3], whereas, the stone removal may be incomplete in up to one third of patients undergoing surgery[6,11]. Only one of our patients (Case 5) had extrahepatic bile duct stone which was removed by ERCP. The remaining 4 patients had both extrahepatic and intrahepatic duct stones. Repeated ERCP sessions (minimum 2) in all four patients successfully removed all stones in only two patients (Case 1 & 2). Other two patients (Case 3 & 4) required surgery to remove these stones. Extracorporeal shockwave lithotripsy (ESWL) was given to two patients but it seemed to fragment stones in only one patient. Surgery was indicated in our patients when ERCP failed to remove all bile duct stones or when patients had gallstone disease. Other indications for surgery in RPC is to bypass strictures[11] or resection for focal hepatic lesion[12]. In the absence of stricture, common bile duct exploration with the help of choledochoscope will be successful in removing all stones. When there is a stricture in the main bile duct, a bypass above the stricture by constructing a Roux-en-Y hepaticojejunostomy with subcutaneous implantation of long lateral limb for subsequent access to biliary tree is recommended[12]. We have done choledochoduodenostomy in one of our patients (Case 3) which would allow subsequent access to bile ducts if needed. If the intrahepatic disease is confirmed to a localized area of liver, affected segment can be resected[12]. Intrahepatic duct stricture can be managed by repeated balloon dilatation[14]. Metallic stent placement across the stricture was found to be ineffective in long term due to blockage by recurrent stone or epithelial hyperplasia[15] Despite the aggressive intervention, both stones and strictures have high recurrence rate[13].

CONCLUSION

Patients with RPC on presentation are seriously ill and need aggressive resuscitation, wide spectrum antibiotics with early decompression of the biliary tree. ERCP plays a major role in the diagnosis and therapy. Surgery may be needed in selected patients. As stone formation and stricture have a high recurrence rate, these patients need a long-term follow-up. Unfortunately, this was not possible in our patients as they returned to their country after completing initial treatment.

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