CASE REPORT

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Rare primary intrahepatic lithiasis in a young patient

Retka primarna intrahepatična litijaza kod mladog bolesnika

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Abstract

Introduction. Intrahepatic lithiasis (IHL) is a disease that occurs in middle-aged and elderly people. Presentations of IHL in the young are rare, and considerations in the differential diagnosis include primary sclerosing cholangitis, recurrent pyogenic cholangitis, bile acid transporter defect, Caroli's disease, and other known genetic diseases. Treatment is often complex, all in order to prevent complications. In this report, we describe the diagnosis and treatment, with the application of a flexible ureteroscope of 4 Fr, of a younger patient with intrahepatic lithiasis. **Case report.** A 25-year-old man appeared with a known diagnosis of IHL and a recurrent attack of abdominal pain that required medical treatment. Magnetic resonance imaging of the abdomen showed segmental stenosis of the left bile duct and segmental bile duct for the lateral section with intraductal calculi and its proximal dilatation and mild dilatation of the bile ducts for liver segments II and III. During surgery, a cholangiography and ultrasonography of the liver were performed. Through choledochotomy, the bile ducts were flushed, and extirpation of the several calculi was performed. The bile ducts were examined with a choledochscope, and the remaining concrements were removed with a flexible ureteroscope. **Conclusion.** Segmental liver bile ducts may be explored with a flexible ureteroscope without bile duct injury or trauma. In selected cases, with isolated lithiasis in one liver lobe and the absence of concomitant diseases, IHL can be treated surgically without liver resection. This case is unique because we did not perform liver resection but duct stone extraction, which was an appropriate treatment since there was no recurrence during the two-year follow-ups.

Key words:
bile ducts, intrahepatic; choledocholithiasis; diagnosis; surgical procedures, operative; ureteroscopes; treatment outcome.

Apstrakt

Uvod. Intrahepatična litijaza (IHL) je bolest koja se javlja kod osoba srednjeg i starijeg životnog doba. Pojava IHL kod mladih osoba je retka, a diferencijalna dijagnoza uključuje primarni sklerozirajući holangitis, rekurentni piogeni holangitis, defekt transportera žučne kiseline, Karolijevu bolest i druge poznate genetske bolesti. Lečenje je često složeno, sa ciljem da se spreče komplikacije. U ovom radu prikazujemo dijagnozu i lečenje bolesnika mlađeg životnog doba sa IHL korišćenjem fleksibilnog ureteroskopa promera 4 Fr. **Prikaz bolesnika.** Muškarac star 25 godina, sa ranije poznatom dijagnozom IHL i ponavljajućim napadima bolova u stomaku koji su zahtevali medicinski tretman, javio se na pregled. Magnetna rezonanca abdomena pokazala je segmentnu stenozu levog žučnog kanala i segmentnog žučnog kanala za lateralnu sekciju sa intraduktalnim konkrementima, kao i proksimalnom dilatacijom, te blagu dilataciju žučnih puteva za II i III segment jetre. Tokom operacije urađeni su holangiografija i ultrazvuk jetre. Žučni kanali su isprani kroz holedohotomiju i izvršena je ekstirpacija nekoliko konkremenata. Pregled žučnih puteva urađen je holedoskopom, a preostali konkrementi su uklonjeni fleksibilnim ureteroskopom. **Zaključak.** Segmentni žučni kanali jetre mogu se eksplorisati bez traume fleksibilnim ureteroskopom. U odabranim slučajevima, kod izolovane litijaze u jednom režnju jetre i odsustva pratećih bolesti, IHL se može lečiti hirurški, bez resekcije jetre. Prikazani slučaj je jedinstven jer nismo uradili resekciju jetre, već ekstrakciju konkremenata iz žučnih kanala, što je bio odgovarajući tretman, imajući u vidu odsustvo recidiva tokom dvogodišnjeg praćenja.

Ključne reči:
žučni putevi, intrahepatički; holedoholitijaza; dijagnoza; hirurgija, operativne procedure; ureteroskopi; lečenje, ishod.

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Introduction

Intrahepatic lithiasis (IHL) is defined as the presence of gallstones in the bile ducts located proximally from the junction of the main hepatic ducts (left or right hepatic duct, sectional and segmental ducts, and their branches) [1]. Primary IHL is rare in Europe, and difficulties encountered in the etiological classification have led to the absence of generally accepted standard treatment [2]. Therefore, there is a high risk of residual and/or recurrent stones.

The highest incidence of primary IHL is present in Asia, predominantly in China, Japan, and South Korea, while the relative incidence in the Western world is about 1%. The disease occurs in people with lower socioeconomic status [3]. Primary IHL occurs more often in the 5th and 6th decades of life without gender preference. However, concomitant intrahepatic and extrahepatic stones are present in the older age groups (7th and 8th decades) and are found in approximately 70% of all cases of hepatic lithiasis [4]. However, it is not the same entity, and only several cases have been reported of primary IHL patients under 30 years of age.

In this paper, we present steps in diagnosis and our own experience in treatment using a flexible ureteroscope of 4 Fr for successful sparing surgical treatment of primary IHL in a young patient.

Case report

A 25-year-old man was admitted to our hospital for the treatment of previously diagnosed IHL. The patient had abdominal pain, nausea, and vomiting a month prior to hospitalization. Gallstones in the common biliary duct and IHL were diagnosed on abdominal ultrasound. The patient reported no chronic diseases, denied previous surgeries, and suggested allergies to penicillin, metronidazole, and ciprofloxacin. His mother suffered from gallbladder stones, and his father had chronic renal failure. The patient had no surgical interventions in the past.

On admission, he had light tenderness in the upper right quadrant of the abdomen. Increased serum values of direct bilirubin of 15 mmol/L [normal range (NR) 0–5 mmol/L] and indirect bilirubin of 34 mmol/L (NR 2–15 mmol/L) were found in the laboratory findings on admission. Findings of other laboratory parameters were presented in normal ranges. Magnetic resonance (MR) imaging of the abdomen showed segmental stenosis of the left bile duct and segmental bile duct for the lateral section with intraductal calculi and its proximal dilatation and mild dilatation of the bile ducts for liver segments II and III. The gallbladder calculi were also found, as well as the direct confluence of the right anterior sectional duct into the common hepatic bile duct. Extrahepatic bile ducts were of regular contour, width, and lumen without defined calculi.

Upper endoscopy showed no pathologic findings, while endoscopic ultrasonography (US) indicated multiple calculi of the left bile duct and one calculus in the right bile duct.

After preoperative counseling, an open surgery was performed. Intraoperatively, the gallbladder had a thickened wall, and a 10 mm stone diameter was found in its lumen. Following choledohotomy and extrahepatic and intrahepatic bile duct exploration with biliary forceps, a stenosis of the left hepatic duct was found. During surgery, a cholangiography and US of the liver were performed. An IHL has been verified for stenosis of the left bile duct and segmental distal dilatation (Figures 1 and 2). The variation of intrahepatic bile ducts was found with a separate drainage of the anterior right segmental branch into the common hepatic bile duct.

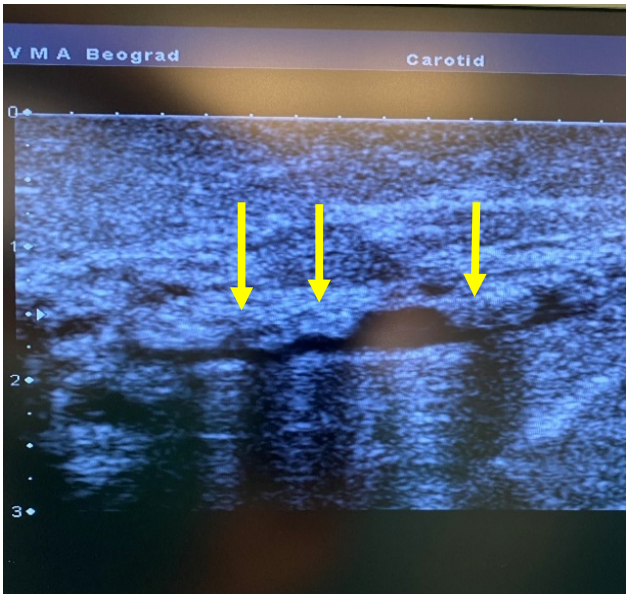

Fig. 1 – Intraoperative ultrasound with visible calculi in the intrahepatic bile ducts.

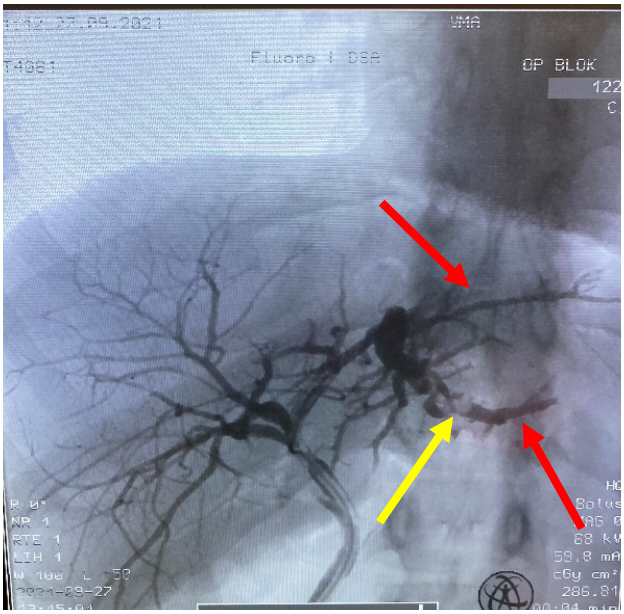

Fig. 2 – Intraoperative cholangiography with intrahepatic calculi in liver segments II and III (yellow arrow) and proximal dilatation of intrahepatic bile duct (red arrows).

The gallbladder was removed first. Through choledochotomy, the bile ducts were flushed with saline, and extirpation of the several calculi was performed (Figure 3). An Olympus (Melville, NY, USA) choledochoscope (URF-P2; outer diameter, 2.8 mm; channel, 1.2 mm) was then introduced into the biliary tree through choledochotomy; however, segmental ducts could not be explored due to the smaller diameter of the ducts themselves than the choledochoscope. A 0 [Flex X2 (FO) (Karl Storz®)] with a diameter of 4 Fr was used to confirm duct clearance (Figure 4). The remaining calculi were extracted subsequently. At the end of the operation, a biliary T-drain was placed.

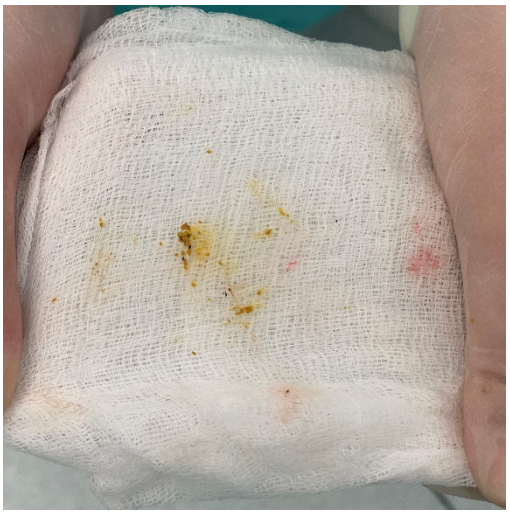

Fig. 3 – Removed bile duct calculi from the left biliary tree.

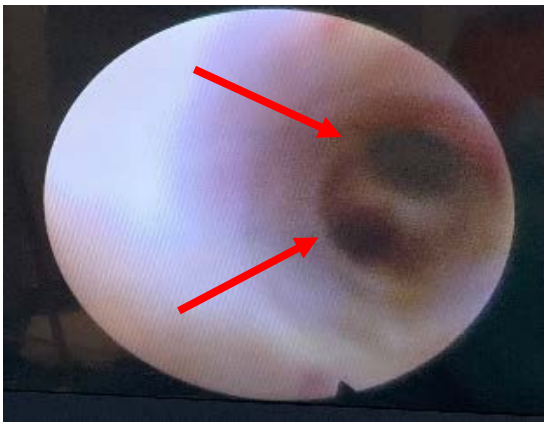

Fig. 4 – Confirmation of bile ducts clearance bile with flexible ureterorenoscope.

The postoperative course was uneventful. Our patient was taken care of for one day in the Intensive Care Unit and was, after that, transferred to the Department of Abdominal Surgery. On the sixth postoperative day, the patient was discharged from the hospital. Biliary T-drain was removed after six weeks following normal cholangiography and a normal level of bilirubin in the blood. During 12 and 24-month follow-ups, the patient was in good condition without rest IHL on repeated MR cholangiopancreatography (MRCP).

Discussion

The highest incidence of IHL occurs between the 5th and 6th decades of life and is usually distributed between 30 and 70 years of age [4]. IHL is more common in younger ages, while both IHL and calculi in the extrahepatic biliary tree occur in older groups. Usually, the first symptoms of IHL occur between the ages of 30 and 50, but sometimes earlier [5].

The differential diagnosis of IHL includes primary sclerosing cholangitis (PSC), recurrent pyogenic cholangitis, bile acid transporter defect, and Caroli's disease [1, 3, 6]. PSC is characterized by biochemical cholestasis, it generally affects the entire biliary tract and is not isolated on one side of the liver [7]. Recurrent pyogenic cholangitis is a chronic infectious process characterized by recurrent attacks of acute bacterial cholangitis in the environment of intrahepatic bile duct dilatation and strictures, which is endemic in Southeast Asia [8]. Bile acid transporter defects, such as ABCB4/MDR3, may occur with IHL at a younger age, but the absence of disease in the right hepatic duct system disputes this as a cause in our patient [6, 9]. Caroli's disease has been described as a congenital malformation of the intrahepatic bile ducts, characterized by the following: segmental cystic dilatation of the intrahepatic bile ducts; increased incidence of biliary lithiasis, cholangitis, and liver abscess; absence of cirrhosis and portal hypertension; association of renal tubular ectasia or similar renal cystic disease. The mode of inheritance is still unclear, but in most cases, it is transmitted in an autosomal recessive manner [10]. However, the MR in our patient and intraoperative US and cholangiography did not support this diagnosis. Finally, the cause of IHL in our patient remains unknown. A cause may be an undiscovered genetic factor due to the positive family history [6].

It is reported that a variation of segmental bile duct drainage may be associated with IHL [11]. Cranial shifting of the right or left sectorial ducts proximal to the hepatic confluence can cause bile stasis and enhance the formation of IHL. Our patient had a separate drainage of the anterior right segmental branch into the common hepatic bile duct. Therefore, the possible mechanism in our patient may be the bile stasis in the left-sided bile ducts; however, the MRCP on follow-up examination 12 months after surgery does not justify this hypothesis.

Primary IHL may be managed surgically and by using nonsurgical alternatives. These conservative or minimally invasive approaches include medical therapy, extracorporeal shock wave lithotripsy, electrohydraulic lithotripsy, transhepatic approach, and endoscopy, but do not eliminate the risk of recurrence neither provides complete clearance of bile ducts [2, 12–14]. On the other hand, endoscopic sphincterotomy and stone removal using endoscopic retrograde cholangiopancreatography may be associated with low rate success and relatively high morbidity, especially in cases with calculi in segmental bile ducts [15, 16]. Alternative to this minimally invasive technique is open surgery or laparoscopy. The main challenge with bile duct exploration is the large impacted stones that cannot be managed using a flexible choledochoscope as it has a narrow working channel and instruments

like graspers cannot be forced through it. In the right or left main bile ducts, a flexible choledochoscope may enter; however, to break stones, it is necessary to apply a holmium laser or an electrohydraulic lithotripter, which are very expensive and not available in most of the developing world [17, 18].

The usage of nephroscope, ureteroscope, and ureterorenoscope in bile duct exploration and removal of bile duct stones has been described in only two reports [19, 20]. In addition, in those reports, the laparoscopic approach used a rigid ureteroscope. A probable reason why a rigid ureteroscope was used lies in the fact that a flexible fiberoptic instrument is composed of thousands of densely packed flexible glass fibers, which is liable to damage [19]. In open surgery, gentle movements allow the safe introduction of fiberoptic instruments into the segmental bile ducts, which is not possible in laparoscopy. In our case, we used a flexible ureterorenoscope of 4 Fr for the exploration of bile ducts for liver segments II and III. The scope could be maneuvered into the distal and proximal bile ducts easily. To our knowledge, this is the first exploration of bile ducts with a flexible ureterorenoscope.

It has been reported that liver resection is the only valid treatment of primary IHL since it removes both stones and the involved area, thus eliminating the risk of recurrence [11, 21, 22]. However, the majority of patients who underwent liver resection for IHL had some concomitant disease (Caroli's syndrome, cholangitis, and/or previous biliodigestive anastomosis) or had recurrent IHL [11, 21]. In such patients, performing a hepatic resection as a treatment of choice is justified. When the etiology of primary IHL is unknown or uncertain, and the absence of cholangitis, hepatectomy, as a first-line treatment, should be avoided whenever possible. In our patient, we performed successful bile duct stone extraction without liver resection, which was an appropriate treatment since there was no recurrence during two-year follow-ups.

Conclusion

Primary IHL is rare at a young age, and as such, it needs additional diagnostic tools to illuminate possible etiological factors. Segmental liver bile ducts may be explored successfully with a flexible ureteroscope, which provides adequate visualization and maneuvering without bile duct injury or trauma. In selected cases with unknown etiology of IHL, isolated lithiasis in one liver lobe, and absence of concomitant diseases, IHL can be treated surgically but without liver resections.

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