CASE REPORT

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Percutaneous biliary intervention for primary sclerosing cholangitis in a patient with situs inversus totalis

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Situs inversus totalis is a rare condition that may not cause symptoms and often occurs concomitantly with other congenital anomalies. Special attention must be given to these patients when invasive interventions are required. There are only a few reports concerning percutaneous transhepatic biliary interventions in situs inversus totalis, but technical details were not fully explained. In this report, we attempt to explain the technical details of percutaneous transhepatic cholangiography, percutaneous biliary drainage and percutaneous biliary balloon dilatation in a patient with known primary sclerosing cholangitis and situs inversus totalis. While performing percutaneous biliary interventions in patients with situs inversus totalis, the key point is to use the mirror image of the traditional technique in an imaginary line dissecting the midline of the abdomen.

Key words: Situs inversus, primary sclerosing cholangitis, percutaneous biliary intervention

Total situs inversuslu primer sklerozan kolanjiti olan hastada perkütan bilier girişim

Situs inversus totalis tek başına semptomlara neden olmayan ve sıklıkla diğer konjenital anomalilerle brlikte görülen nadir bir durumdur. İnvaziv girişim gerektiren durumlarda bu hastalara dikkat edilmelidir. Bu tür hastalarda perkütan transhepatik girişimlere ait oldukça nadir bildirim bulunmaktadır ve tekniğin ayrıntılarına ait veriler mevcut değildir. Primer sklerozan kolanjiti olan situs inversus totalisli 44 yaşındaki bir erkek hastada akut kolanjit atağının iyileştirilmesi için uygulanan perkütan bilier kolanjiyografi, perkütan bilier drenaj, perkütan bilier balon dilatasyon uygulamasını tanımladık. Situs inversusun, situs solitusun anteroposterior akstaki ayna hayali olduğu göz önüne alınırsa; perkütan bilier girişimlerde, ksifoid proses ve umbilikusu birleştiren bir düzlem referans alınarak geleneksel öğretide yapılan işlemlerin ayna hayali kullanılabilir.

Anahtar kelimeler: Situs inversus, primer sklerozan kolanjit, perkütan bilier girişim

INTRODUCTION

Situs inversus totalis (SIT) is characterized by the mirror image of normal situs. SIT is a rare condition that may not cause symptoms and often occurs concomitantly with other congenital anomalies. Special attention must be given to these patients when invasive interventions are required. There are only a few reports concerning percutaneous transhepatic biliary interventions in SIT, but technical details were not fully explained. In this report, we attempt to explain the technical details of percutaneous transhepatic cholangiography (PTC), percutaneous biliary drainage (PBD) and percutaneous biliary balloon dilatation (PBBD) in a patient with known primary sclerosing cholangitis (PSC) and SIT.

CASE REPORT

A 44-year-old male patient with SIT referred to our clinic for PBD and PBBD. The patient was followed for known PSC and ulcerative colitis. His

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past medical history revealed previous hepaticojejunostomy operation for benign biliary stenosis caused by choledocholithiasis. The patient was unsuitable for endoscopic retrograde cholangiopancreatography (ERCP). The main complaints of the patient were jaundice, pruritus and fever. Laboratory examination revealed a direct bilirubin level of 15.31 (normal: 0-0.3 mg/dl), total bilirubin level of 17.73 (normal: 0-1.10) and serum alkaline phosphatase level of 642 U/L (normal: 40-129 U/L). In addition to SIT findings, there were dilatations in the intrahepatic bile ducts and abrupt obstruction of the common hepatic duct on ultrasound (US) and computed tomography (CT) examination of the patient. After the detailed US examination of the bile ducts with a digital fluoroscopy unit, PBD and PBBD were planned (Figures 1, 2). The interventions were done via a left-sided approach. The movement of the left hemidiaphragm was observed under fluoroscopy. An imaginary line dissecting the abdomen from the xiphoid process was envisioned, and we imagined ourselves to be on the right side. A 21-G Chiba needle was introduced into the left 10^{th}-11^{th} intercostal space and PTC was done under local anesthesia and sedation (Figure 3). There was a high-grade stenosis extending from the equivalent common hepatic duct (the term "equivalent common hepatic duct" is used to describe the common hepatic duct in situs inversus) to the hepaticojejunostomy site. Proximal to the high-grade stenosis, the bile ducts were dilated. There were also local non-critical stenoses in the equivalent right and left hepatic ducts. Considering the equivalent ducts as the mirror image of the normal ducts, a 21-G Chiba needle was inserted into the equivalent right anterior hepatic duct. This duct was chosen becau-

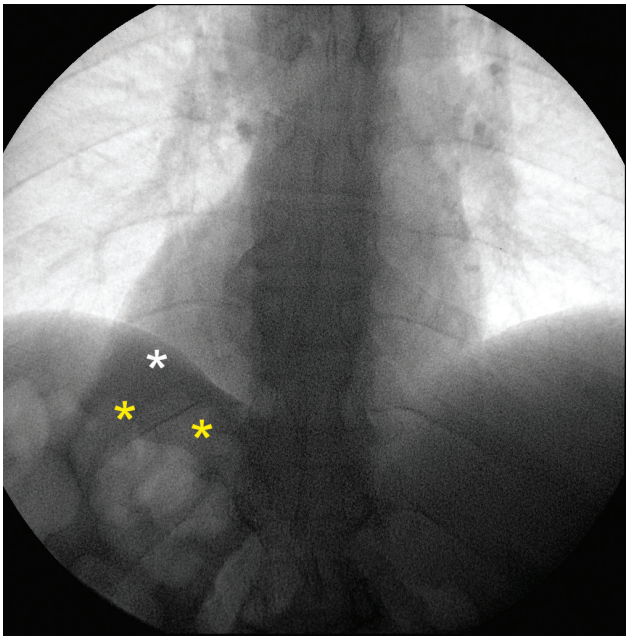

Figure 1. Dextrocardia (white asterisk) and right-sided air spaces of the stomach (yellow asterisks) in a patient with SIT.

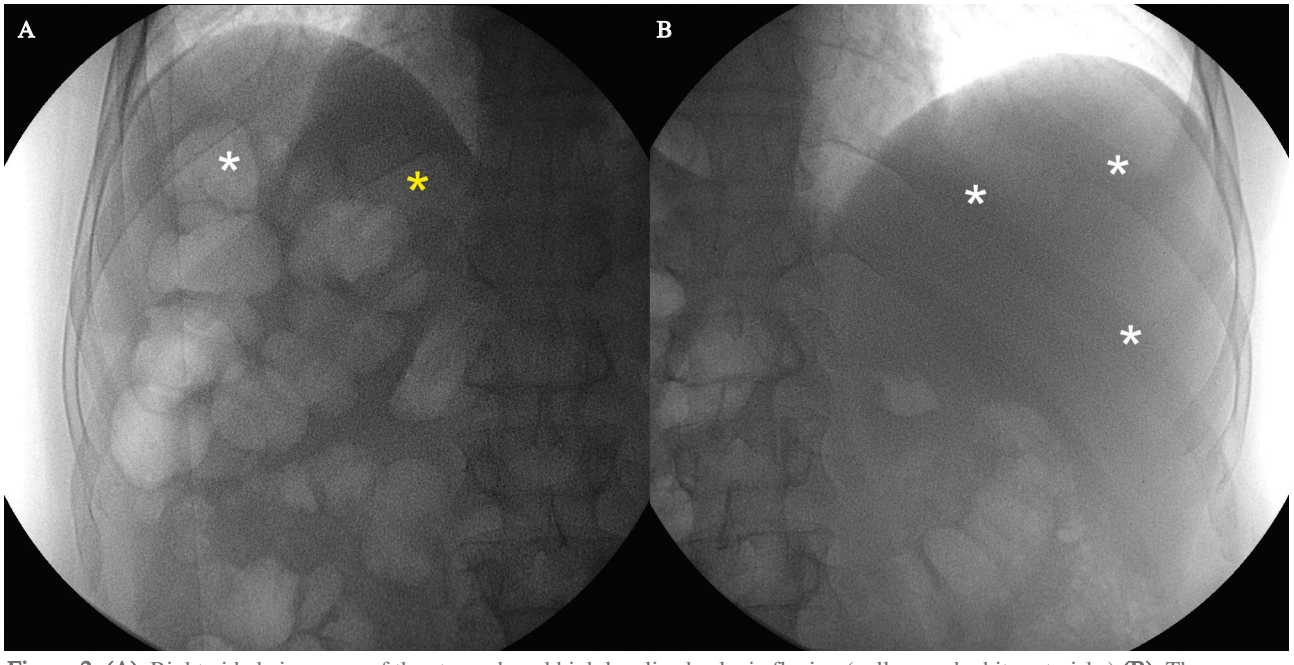

Figure 2. (A). Right-sided air spaces of the stomach and high localized splenic flexion (yellow and white asterisks) **(B).** There are no air spaces in the localization of the left-sided liver (white asterisks).

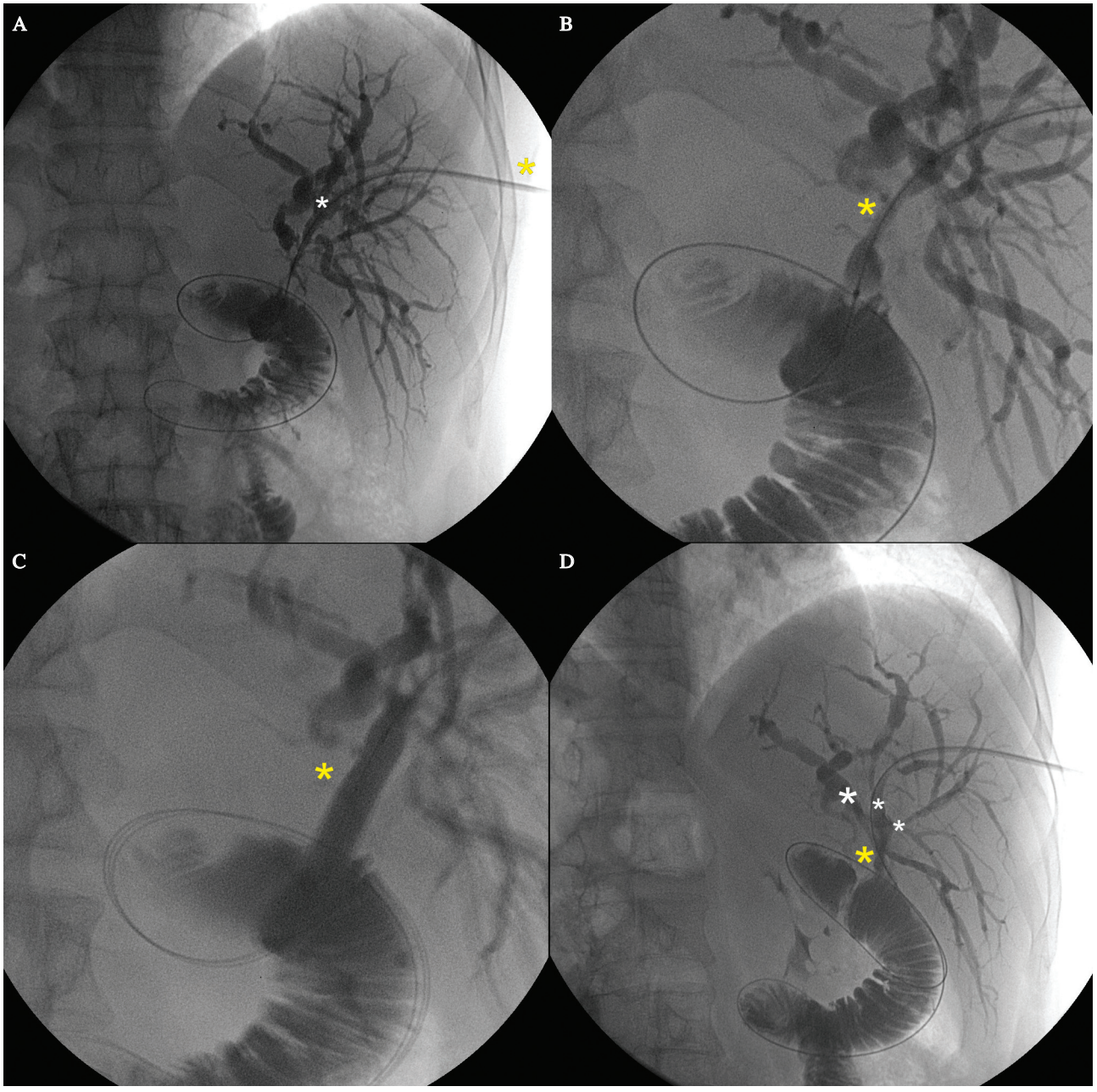

Figure 3. (A). Left-sided midaxillary approach (yellow asterisk) targeting equivalent right anterior hepatic duct (white asterisk). **(B-C).** PBBD to the equivalent common hepatic duct (yellow asterisk). **(D).** Satisfactory dilatation in the stenotic segment (yellow asterisk). Local non-critical stenotic segments in the equivalent right and left hepatic ducts (white asterisks).

se of its more vertical nature, which enabled crossing the stenosis by a guidewire. After guidewire insertion into the bile duct, coaxial system (Accu Stick II Introducer System, Boston Scientific, Natick, MA) was advanced into the bile duct and replaced with a hydrophilic catheter (Glidewire, Terumo Interventional Systems) and hydrophilic guidewire (Glidewire, Terumo Interventional Systems). The stenosis was traversed with the guidewire. After insertion of a 5F introducer sheath, dilatation using an 8 x 40 mm balloon (Opti-Plast Xtra Thin PTA Dilatation Catheter, Bard, USA) was performed for 3 minutes at 8 atm pressure to the stenotic segment. Satisfactory dilatation was obtained. The introducer sheath was removed and an internal-external drainage procedure was performed with polyethylene biliary drainage catheter (Angiotech, PBN Medicals, Denmark). Clinical and laboratory improvement was observed following the biliary drainage. After recovery from the

acute condition, the patient was evaluated for eventual surgical revision and no additional interventions were planned.

DISCUSSION

Situs solitus refers to the normal position of the thoracic and abdominal organs. Situs inversus is the mirror image of the normal situs in the anteroposterior axis of the body. Tri-lobed lung, liver, gallbladder, and other internal organs are on the right side in normal situs whereas they are on the left side in situs inversus. Situs inversus can be seen with levocardia or dextrocardia. Situs inversus with dextrocardia is defined as SIT. Situs ambiguus or heterotaxy is also an abnormal situs that falls between the normal situs and situs inversus and includes asymmetric distribution of the chest and abdominal organs, as well as conditions such as asplenia, polysplenia, intestinal malrotations, complex atrial morphology, and midline localized liver. SIT is a rare condition that may not cause symptoms and often occurs concomitantly with congenital anomalies. Special attention must be given in these patients when invasive interventions are required. In the case of acute abdomen or trauma, these patients must be examined in detail with CT prior to surgery (1-6).

Primary sclerosing cholangitis (PSC) is a chronic liver disease caused by progressive inflammation and scarring of the intrahepatic and extrahepatic bile ducts. In patients with PSC, during ursodeoxycholic acid treatment with 8.8–17.4 mg/kg, dominant stenoses of bile ducts were observed by Stiehl et al. (7). Endoscopic opening of these stenoses with balloon dilatation was found to improve survival -free of liver transplantation- significantly (7). After the first dilation of a dominant stenosis, Gotthardt et al. (8) observed a survival free of liver transplantation rate of 81% and 52% at 5 and 10 years, subsequently. Endoscopic therapy of symptomatic dominant strictures in PSC with balloon dilatations, bougie dilatations or stenting was found safe and effective by Parlak et al. (9). When endoscopic intervention fails, PBBD is an alternative method that improves the acute clinical condition of the patient (10,11). However, it is important to remember that the main treatment of this disease is liver transplantation.

There are reports about how surgery or ERCP procedures could be done in the patients with situs inversus (2,5,6). To our knowledge, there are only a few reports concerning percutaneous transhepatic biliary intervention in SIT, but technical details were not fully explained (6,12). Although very rare, percutaneous biliary interventions may be required in patients with SIT. In traditional thought, the right intercostal space is used to enter the bile duct with right midaxillary line approach, and the left subxiphoid approach is needed to enter the left bile ducts. This is not valid for the patients with situs inversus. While performing percutaneous biliary interventions in patients with SIT, the key point is to use the mirror image of the traditional technique in an imaginary line dissecting the midline of the abdomen. If difficulties are encountered in envisioning this imaginary line or in the technique, prone positioning of the patient and performing the procedures from the right side could be used, as is done in combined interventions with ERCP. Although the anatomy in situs inversus is equivalent to the anatomy in situs solitus, preprocedural US and CT evaluations of the patient may be needed. Performing the procedures under the guidance of both US and fluoroscopy may be more useful in these patients.

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