CASE REPORT

AGENESIS OF GALL BLADDER DIAGNOSED UNEXPECTEDLY DURING A LAPAROTOMY FOR CHOLECYSTECTOMY

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ABSTRACT

We report a case of a 55 year old female with suspected chronic cholecystitis due to cholelithiasis.She was operated on and found to have agenesis of the gall bladder, which is an extremely rare clinical condition. Standard investigative modalities currently used for chronic cholecystitis might be misleading. Agenesis of the gall bladder should be kept in mind whenever the gall bladder is improperly visualised in routine ultrasound methods. It is difficult to diagnose gall bladder agenesis preoperatively, as investigations tend to be misleading and therefore it is usually diagnosed intraoperatively.

Keywords: Gall bladder agenesis , Cholelithiasis , Cholecystectomy.

KOLESİSTEKTOMİ AMACIYLA YAPILAN LAPAROTOMİ ESNASINDA ŞAŞIRTICI ŞEKİLDE TANI KONULAN SAFRA KESESİ AGENEZİSİ

ÖZET

Safra kesesi taşına bağlı kronik kolesistit tanılı 55 yaşında kadın olguyu sunuyoruz. Yapılan ameliyatta oldukça nadir görülen safra kesesi agenezisi saptandı. Kronik kolesistit tanısında kullanılan standart tanısal yöntemler yanıltıcı ve şaşırtan sonuçlar verebilmektedir. Ultrasonografik tanı yönteminde safra kesesinin tam olarak tanımlanamadığı durumlarda safra kesesi agenezisi ihtimali düşünülmelidir. Kullanılan tanısal yöntemler yanıltıcı olabildiğinden safra kesesi agenezisi tanısı ameliyat öncesi değil ameliyat esnasında konulur.

Anahtar Kelimeler: Safra kesesi agenezisi , Kolelithiasis , Kolesistektomi.

INTRODUCTION

Gall bladder agenesis is a rare condition that results from the failure of the cystic bud to develop in the 4th week of the intrauterine life. It normally develops from the caudal part of the hepatic diverticulum[1]. Recently, a review of autopsies reported in the literature shows an incidence of about 1/6334 live births[2]. Although the female/male ratio in postmortem studies has been reported as equal[3] , in clinical studies there is a 3/1 predominance of females[4]. Agenesis is usually discovered during laparotomy or laparoscopy for cholecystectomy, since ultrasound examination of a patient with suggestive symptoms not visualizing the gall bladder is compatible with chronic cholecystitis (shrunken gallbladder). The surgeon must confirm agenesis by thoroughly examining the most common sites for an ectopic gall bladder and should perform an intraoperative cholangiogram and abdominal ultrasound if necessary. After the operation most of the

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patients become asymptomatic for unexplained reasons. In this report , we describe a case presented as cholelithiasis and found to have no gall bladder intraoperatively.

CASE REPORT

A 55-year old female patient was admitted electively to our clinic with cholelithiasis. She had a medical history of severe and colicy right upper abdomen pain accompanied by nausea, vomiting and intolerance to fatty foods for the last 2 years. Her physical examination was uneventful. All biochemical and hematological investigations were within normal limits. The workup before her admission included an abdominal ultrasonography examination. Ultrasonography revealed a gall bladder completely filled with bile stones with enlarged thickness of the wall and adhesions to the liver(Figure 1). Also visualization of the gall bladder was not easy because of the intestinal segments nearby. The biliary ducts were within normal limits. The patient underwent laparotomy for cholecystectomy and the gall bladder was not found. In the normal localization of the gall bladder there were adhesions of the liver with a transverse colon segment. A small green colored tissue, probably a remnant of the gall bladder and a lymph node adhered to it was resected for pathological investigation (Figure 2). Intraoperative cholangiography could not be performed. Instead a post-operation magnetic resonance cholangio pancreatography (MRCP) was planned. Despite a through examination of the abdominal cavity including the falciform ligament, lesser sac, retrohepatic and retroduodenal spaces, the gall bladder was not found. The diagnosis of gall bladder agenesis was made during the operation and the abdomen was closed. The patient was discharged from the hospital without having had any complaints. The MRCP was performed postoperatively and abnormality was detected. Also it was reported that the patient had had a cholecystectomy (Figure 3). The pathological report of the specimen revealed mucosal tissue of the gall bladder with omental tissue and a lymph node with reactive hyperplasia. The patient remains asymptomatic 3 months after the laparotomy.

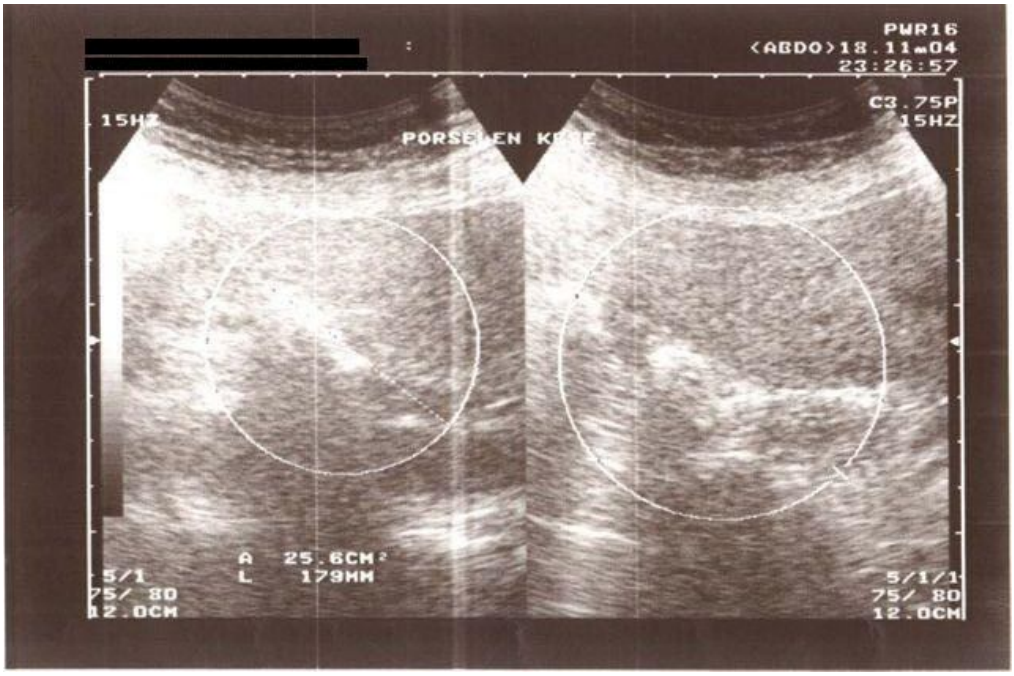

Figure 1: Gall bladder of the patient incorrectly investigated and diagnosed as cholelithiasis.

Figure 2: Resected remnant material reported as gall bladder mucosa and omental tissue.

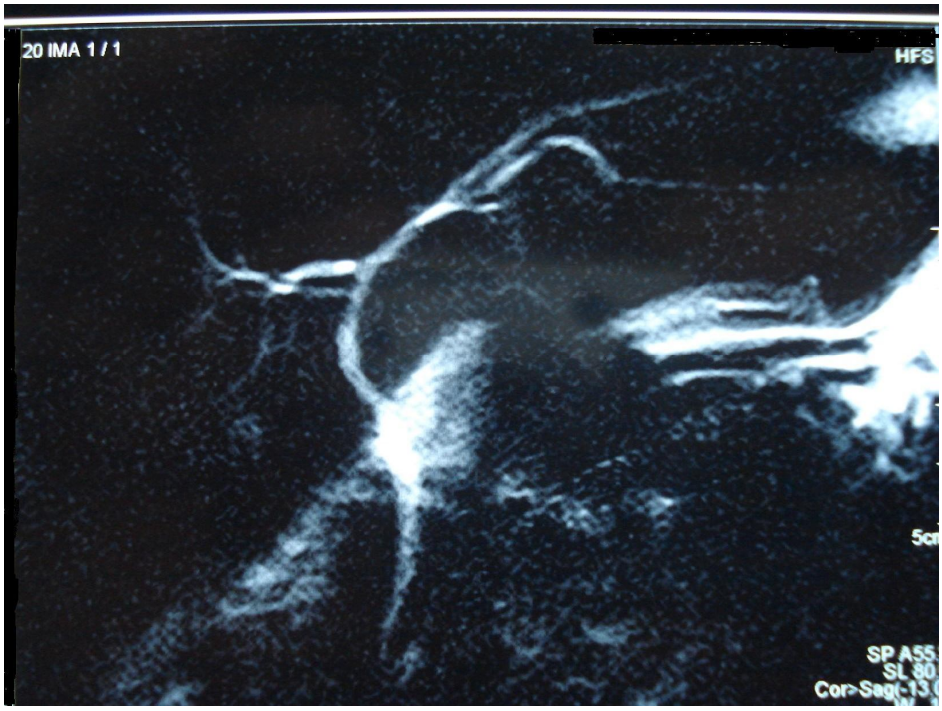

Figure 3: MRCP of the patient performed postoperatively.

DISCUSSION

Agenesis of gall bladder is an extremely rare condition with an incidence of 0,01-0.02% [2]. Several studies have revealed a strong familial association with this condition[2]. Any defect in the developmental process leads to the agenesis of gall bladder. This anomaly is transmitted as a non-sex linked trait with variable penetration. The actual incidence is not known[5]. After a review of 400 cases, three groups of presentations were noted: a) Asymptomatic patients(35%) who were diagnosed as incidental finding on abdominal exploration for some other reason ; b) Symptomatic agenesis(50%) : One third of these patients will have a dilated common bile duct(CBD) and another one third will have stones in CBD ; c) Children with congenital anomalies (15-16%) like agenesis of the lung, Tetralogy of Fallot, anomalous extremities, genitourinary and rarely gastrointestinal anomalies. The complexity of the situation makes such patients incompatible with survival (15-16 %)[6].

In symptomatic cholecystic agenesis, patients undergo surgery for right hypochondrial symptoms. Common symptoms include chronic right upper quadrant pain (90%), dyspepsia (30%), nausea and vomiting (66%), fatty food intolerance (37%), jaundice (35%). Possible mechanisms of symptoms include primary duct stones, biliary dyskinesia or non-biliary disorder. Usually the diagnosis is established during the operation , as in our patient. Commonly performed investigations tend to mislead the diagnosis of this clinical entity, as a result of which preoperative diagnosis is almost impossible. Ultrasonography is highly operator dependent; periportal tissues or subhepatic peritoneal folds are usually focused and interpreted as thick, contracted, shrunken or scarred gall bladder, so presents sensitivity of less than 100% for the identification of the organ[7]**.** In our case, the most likely cause of the false positive sonographic finding, was the visualization of a small or large bowel loop in the area of the gall bladder. Intraoperatively, if the gall bladder is not visualised in its normal anatomic position, a thorough search should be carried out in the ectopic locations intrahepatic, left sided, beneath the posteroinferior surface of the liver between the leaves of lesser omentum, retroperitoneal, retrohepatic, within the falciform ligament, retropancreatic and retroduodenal[6]**.** Any scar in porta hepatis and gall bladder fossa should be dissected. Complete exploration of the duodenum should be done. Also, CBD must be identified along its whole length from the confluence of the right and left hepatic ducts to the duodenum[7,8]. If all these manoeuvres fail to identify the gall bladder, peroperative cholangiography is mandatory in order to look for : a) intrahepatic gall bladder , b) ectopic gall bladder , c) stones in CBD (which is the commonest association). CBD exploration is carried out only if the cholangiogram shows calculi in CBD or CBD is dilated more than 20 mms[6]. If the cholangiogram is otherwise normal, nothing further needs to be done. There is a pathophysiological similarity between agenesis of the gall bladder and dilatation of the hepatic duct that may occur after cholecystectomy[8,9]. The hepatic bile duct may substitute for the absent gall bladder by becoming dilated and taking on the function of bile storage in agenesis[8,9]. In such a situation, the presence of dyskinesia of the bile tract, elevation of the basal pressure of the Oddi sphincter , cholestasis or infection of the bile ducts may provoke the onset of a clinical condition and/or lithiasis of the common bile duct, especially when two or more of these events occur in association[5,8,10]. These patients generally become asymptomatic in the postoperative period. A probable explanation that has been given is lysis of periportal and right hypochondrial adhesions at the time of surgery[2]. In conclusion, the diagnosis of gall bladder agenesis is probably impossible preoperatively and therefore remains a diagnostic dilemma. The surgeon should confirm agenesis of the

organ.Intraoperative cholangiography or ultrasonography must be performed.

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