Gall bladder hypoplasia with common bile duct stone: A rare case report

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Abstract

Gall bladder disease and cancer is a common finding and is highly prevalent along the plains of river Ganga in parts of northern India. Surgical intervention is routinely required in the management of many of such cases. So, the clinicians treating these patients must have a thorough knowledge of both the normal and aberrant anatomy of the gall bladder and surrounding extra biliary structures. Though very rare, aberrations in the embryonic development may present as an absent or hypoplastic non-functional gall bladder. These patients may become symptomatic and present with biliary colic. In the given case report, we present the case of a 52-year-old lady, who presented with pain abdomen and jaundice. Her pre-operative work up showed a severely contracted and dysmorphic gall bladder which during surgery was found to be a rudimentary hypoplastic gall bladder.

Keywords: Gall bladder, Biliary pain, Stones, Agenesis, Hypoplasia

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Introduction

Gall bladder disease and cancer is highly prevalent along the plains of river Ganga in northern India, which includes the state of Bihar.¹ Diseases mainly include cholecystitis (acute, chronic), gallstones (solitary, multiple) and gallbladder cancer.² A multi-institutional specialized field study by the GANGLA (Gallbladder Abnormalities in Northern Gangetic area) study group to estimate community based prevalence of gall bladder disease in these areas showed a prevalence rate of 4.55/100 persons in Patna, the capital city of Bihar and 7.96/100 persons in Vaishali, a district of Bihar on the banks of river Ganga.³ Surgery is the treatment of choice in most of these cases, which requires a thorough knowledge of the normal anatomy as well as the variants of gall bladder and other extra biliary structures. Normally, gallbladder is a pear shaped hollow viscous organ situated obliquely in a non-peritoneal fossa on the inferior surface of the right lobe of the liver. Anomalies include agenesis of the gall bladder, hypoplastic or rudimentary gall bladder, double gall bladder, short and long gall bladder, septate gall bladder, intra hepatic gall bladder, mobile or floating gallbladder and Phrygian cap.⁴ Gall bladder agenesis or presence of rudimentary hypoplastic gall bladder is a rare anomaly, with an estimated incidence with an estimated incidence 0.02% at birth.⁵ Hypoplasia is less frequent than agenesis of the gallbladder and can be associated with other gallbladder malformations, such as multi-septated gallbladder.⁶ The pathogenesis is related to embryonic development and is due to failure of the gallbladder and cystic duct to bud off from the common bile duct during the fifth week of gestation.⁷

Here we report a case of hypoplastic gall bladder presenting with symptoms similar to biliary colic and associated with common bile duct stone.

Case Report

A 52-year-old lady presented with complains of pain in upper abdomen for the last 1 month. The pain was located in her right upper quadrant, intermittent, sudden in onset, colicky in nature, and radiated to her right scapula. Associated symptoms included nausea, vomiting, and dyspepsia. These symptoms were worsened by meals and particularly by fatty food. Her past medical and surgical history was non-significant. Her physical examination showed icterus and tenderness in right upper quadrant of abdomen. The laboratory examinations showed a normal complete blood count, but a deranged liver function test. Her serum bilirubin was 2.38 mg/dl, SGPT was 48 U/L and serum alkaline phosphatase was 380 U/L. She had an ultrasound of her abdomen which showed that gall bladder was not well visualized and appeared dysmorphic and severely contracted. The common bile duct was dilated measuring 1.4 cm with two large calculi of size 1.3 cm each in the mid CBD lumen with few other tiny calculi in the distal CBD. An exploratory laparotomy was performed, which showed a rudimentary gall bladder on the inferior surface of the right lobe of the liver. There was no neck in the gall bladder and it was virtually merged with the common bile duct. [Fig. 1] There was no cystic artery or any prominent blood vessel coming out from the hepatic, superior mesenteric or right hepatic artery. The rudimentary gall bladder was supplied from multiple small blood vessels from the inferior surface of the liver. The gall bladder was dissected out and the small vessels in the operative bed were cauterised. This was
followed by cholecholithotomy to remove the stones present in the common bile duct. The gross pathological examination showed a grey brown to yellowish soft tissue piece measuring 1.5 x 1 x 0.6 cm. [Fig. 2 & 3] The cut surface was grey brown soft with no lumen. [Fig. 4] The microscopic examination confirmed it to be gall bladder with unremarkable fibromuscular tissue with adherent adipose tissue without any lining epithelium. There was no evidence of granuloma or malignancy. The patient recovered well and there were no post-operative complications.

**Discussion**

The liver, gall bladder and ductal system develops from the hepatic diverticulum of the foregut, at the beginning of the fourth week of development. This diverticulum rapidly proliferates into the septum transversum and divides into two parts; the cranial part is the primordium for the liver and the bile ducts while the caudal part gives rise to the gall bladder and the cystic duct. Initially the extra hepatic biliary system is occluded with epithelial cells, which later gets canalized because of subsequent degeneration of these cells. Any arrest or deviation from the normal embryological developmental process results in some malformation of the gall bladder and the biliary system. A hypoplastic gall bladder may occur when the caudal bud undergoes incomplete development or when the solid stage of the bud is not recanalized.\(^8\) Most cases are sporadic, but familial clustering phenomenon also exists, suggesting that a genetic factor may play a part in the pathogenesis of gallbladder agenesis or hypoplasia.\(^9\) However, no family history was found in our study.

In the literature, there are very few documented cases of gall bladder hypoplasia with symptomatic presentation in an adult patient as seen in our case report as this disease usually presents with symptoms and cholestasis in the first few months of life. It is usually found in association with congenital biliary atresia and in cystic fibrosis.\(^10\) The clinical presentation of gall bladder agenesis has been classified into three classes by Bennion et al which may be also seen in cases of gall bladder hypoplasia. The first group of patients have multiple congenital malformations like biliary atresia, ventricular septal defect, imperforate anus, duodenal atresia, etc. Most of these patients die shortly after their birth. In contrary to the first group, there is a second group of patients who remain asymptomatic and the malformation is discovered accidentally in autopsy or other open surgeries. The third group of patients are the symptomatic type, which may present with biliary symptoms consistent with an intact gallbladder, such as right-upper-quadrant pain, nausea, vomiting, and intolerance to fatty foods, dyspepsia, jaundice, and cholecholithiasis as was seen in our patient.\(^8\) The normal physiological collaboration of gallbladder, bile duct and the Oddi sphincter in bile reservation and discharge under neural and humoral control is interrupted, resulting in the Oddi sphincter dysfunction and bile duct dyskinesia. All these promote biliary stasis, infection, cholecholithiasis and corresponding biliary symptoms.\(^11\) The incidence of cholecholithiasis has been reported to range from 20% to 50% in such cases.\(^8,12\)

A pre-operative diagnosis of gall bladder hypoplasia remains a challenge. As in our patient, the radiologist may report a not well visualized, severely contracted or dysmorphic gall bladder. The differential diagnosis includes an acquired post-inflammatory,
fibrotic shrinkage of gall bladder.\(^{(13)}\) Advanced imaging techniques may be helpful in these cases. Magnetic Resonance Cholangiopancreatography (MRCP) is considered as the test of choice if there is suspicion. It is also helpful in demonstrating an ectopic gallbladder along with other possible anomalies of the biliary tract system, if present.\(^{(14)}\)

**Conclusion**

Though gall bladder hypoplasia is a rare clinical condition, it should be kept in mind whenever the gallbladder is improperly visualized in routine imaging methods in patients with biliary-type pain with or without calculi in the common bile duct. Other associated anomalies of the biliary ducts and vessels may be present which should be kept in mind while operating such patients to avoid complications.

**References**