First case report of gall bladder agenesis and absences of extrahepatic biliary system

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Abstract: extrahepatic biliary tree anomalies were an extremely rare anatomic anomaly, congenital absence of the gallbladder is commonest. Most of the cases are usually discovered intraoperative, because pre-operative diagnosis may be a challenges as patients can present with symptoms mimicking biliary colic. Ultrasonographic diagnosis in majority of cases may be misleading. We report a first case of agenesis of the gallbladder and absences of extrahepatic biliary system.

Introduction
Anomalies of the biliary system are not uncommon; theses may involve the biliary ducts or the gall bladder. Gall bladder anomalies may be in the form of abnormalities in the shape, position, or cystic duct. They are usually discovered during surgery for gall stone disease or other surgeries involving the biliary system of the liver. [1] Agenesis of the gallbladder, an extremely rare condition with incidence of 0.01%–0.02%, is characterized by the absence of the gallbladder without atresia of the extrahepatic biliary system (2) Nakamura K1, Mitsubuchi H, and his colleague report case of Complete absence of bile and pancreatic ducts in a newborn: a new entity of congenital anomaly in hepato-pancreatic development and she died shortly after birth.(3) First case of Gallbladder agenesis was described in 1701 by Lemery.(4)(5). Since then, there have been described about 500 similar clinical cases in the medical literature, most of them are presented with single clinical cases (4)(6). The disease is rare and congenital, with an incidence of 10–65 per 100,000(7)(8). In 50% of the patients with gallbladder agenesis, who complained of the similar symptoms to biliary colic, correct diagnosis: gallbladder agenesis is diagnosed intraoperatively (4)(6),(9),(10),(11),(12). Preoperative diagnosis is often complicated. The main reason of diagnostic difficulties is a “constricted, shrunken gallbladder” and sometimes hyperechogenic shadows seen by radiologist under ultrasonography. Those shadows are considered as gallbladder stones and patient is mistakenly diagnosed with cholelithiasis and later is operated too (4)(10), (12), (13), (14), (15).

Literature Search
A systematic review of the literature, using the, common bile duct, extrahepatic system anomalies, and gall bladder agenesis, no case reported in the literature found of gall bladder agenesis combined with absences of extrahepatic biliary system.

Case Report
A 40-year-old female presented with recurrent attacks of right hypochondrial pain for the last 3 months, dull, radiate to shoulder, associated with nausea, no history of upper gastrointestinal symptoms. No jaundice or history of jaundice. The patient admitted to casualty many time with similar condition. Ultrasound of the abdomen done which showed distended gall bladder with thick wall but no stone. After the initial management, laparoscopic cholecystectomy was considered but the patient insist to do it open. During surgery gallbladder could not be found and there was no extrahepatic biliary system and the duodenum completely attach to the liver. No gall bladder and no extrahepatic biliary system so gall bladder agenesis and absence of extrahepatic biliary system was made initially. Postoperative treatment included analgesics and...
antispasmodics. Later magnetic resonance cholangiopancreatography (MRCP) done which confirm the introperative diagnosis Figure (1). The patient was reassured and counseling about her congenital condition.

Discussion

In earlier studies of the literature, although the male/female ratio has been equal in postmortem studies, most clinical reports are of female patients [16]. Some reports suggest that gallbladder agenesis is familial [2–17]. Kobacker [18] reported agenesis of the gallbladder in two sisters. Investigation of that family revealed three more sisters with nonfunctioning oral cholecystograms. Sugrue et al. [16] reported ectopia and agenesis of the gallbladder in two sets of twins. These literatures suggested that occurrence of agenesis of the gallbladder might have a genetic connection. Most cases are sporadic (around 70%), and there is very little literature on any familial links [19]. The pathogenesis of this abnormality of embryological development is as of yet unknown. This is the first case female of gall bladder agenesis combined with absences of extrahepatic biliary system, the patient did not had family history of biliary anomalies or even gall bladder agenesis.

Most cases remain asymptomatic and are discovered at autopsy; however, approximately 50% of these present with biliary colic mimicking a clinical picture of acute cholecystitis or choledocholithiasis [19,20]. During Gallbladder agenesis the mechanism of biliary colic is unknown, but greater part of authors consider that it is determined by dysfunction of sphincter oddi and biliary dyskinesia. The fact that pain is relieved after antispasmodic treatment strengthens this theory [7,8,9]. This case was presented with feature not away from which was already mentioned in the literature, not just typical presentation but also the correct diagnosis is established intra-operatively.

Ultrasound is investigation of choice for gallbladder and hepatobiliary diseases, this may come true for normal presentation of biliary system, but in cases of biliary anomalies, things are totally different as mentioned in the literature, most of the cases diagnosed intraoperatively (4),(6),(9),(10),(11),(12).

MacDonald FR and his colleagues state that, Standard imaging modalities like ultrasound and nuclear studies cannot accurately diagnose this condition [21,22]; hence, patients are subjected to unnecessary surgical interventions [23]. With advances in imaging technology, a preoperative diagnosis of GA can be made when the gall bladder is not visualized by standard diagnostic tests, avoiding the need for surgery [24]. There exists one more debatable question: should the laparoscopic operation be converted into the
open surgery if no gallbladder could be found during laparoscopy? According to the opinion of quite big amount of surgeons no conversion is needed, because laparoscopy allows performing a complete and high-quality visualization of the abdominal cavity [4], [8], [6], [10, 12, 15]. In this case open surgery was performed due to patient wish, so is it a matter of luck or not that open surgery done in this case, laparoscopy allows performing a complete and high-quality visualization of the abdominal cavity, this could be true for normal anatomy or rare presentations like gall bladder agenesis, but in this case I do not expect to find complete absences of extrahepatic biliary system with gall bladder agenesis and the duodenum was attached and open directly to the liver. At the beginning I think that this is small contracted gall bladder and because of inflammation the duodenum adherent to the gall bladder. To make things clear I make use of tactile sensation (which lost in laparoscopy) by introduce small size nasogastric tube and directed toward the liver, surprise enough it was inside the liver. So this first case of gall bladder agenesis and complete absences of extrahepatic biliary system.

Conclusion

This is first case report of gall bladder agenesis and absences of extrahepatic biliary system. With the increase numbers of biliary anomalies in the literature, and with the difficulties in diagnosing most of the biliary anomalies with ultrasound, we recommend preoperative MRCP as gold standard investigation for gall bladder diseases to unnecessary surgical interventions.

Conflict of Interest

No conflict of interest

References

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