Congenital duplex gallbladder anomaly presenting as gangrenous perforated intrahepatic cholecystitis mimicking a gas forming liver abscess: A case report and literature review

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Abstract

Background: Ectopic intrahepatic gallbladder is a rare phenomenon. Gallbladder duplication is an even rarer phenomenon. Pathological processes are more common in congenital anomalies of the gall bladder than normal gallbladders due to poor drainage.

Case Report: We present a case of duplex gallbladder with one component intrahepatic and the other extra-hepatic, the duo draining via a common cystic duct into the common bile duct. Both gallbladder moieties were diseased. The intrahepatic moiety was gangrenous and perforated thus mimicking an intrahepatic abscess by a gas forming organism .The extra-hepatic moiety was chronically inflamed and packed with gallstones of the same physical and biochemical characteristics as the intrahepatic moiety. The definitive diagnosis was only made at emergency laparotomy. Stone gathering and debridement of the ruptured, gangrenous intrahepatic moiety and cholecystectomy for the extra-hepatic moiety was done. On table cholangiography, though desired, was not available. The patient fully recovered after post-operative intensive care.

Conclusion: An extensive internet literature search did not reveal any previously described case. This could be the first such case described in the world literature. Though rare, congenital anomalies of the gallbladder must be known to surgeons as they can present unexpectedly and pose diagnostic and operative surgical challenges with serious clinical implications. The management challenges experienced and literature review is presented.

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Introduction

Anomalies of the gallbladder are rare.[1] They may be discovered incidentally or when complications or some other pathological processes arise. In such situations they cause diagnostic problems and surgical challenges if they have not been thought of. On table cholangiography if available is indispensable for elucidating the biliary tree anatomy prior to definitive surgery.[2-4] Anomalies of the gallbladder are divided into three major groups; namely anomalies of formation, anomalies of number and anomalies of position.[5]

Anomalies of number range from agenesis to duplication of the gallbladder. Duplication is a rare gallbladder anomaly occurring in 1 in 4000 cases.[1] The first documented case of double- gallbladder was in a human sacrificial victim of the Emperor Augustus on the day of the battle of Actium in 31 B.C.[6] Since then cases of duplicated gallbladder have been described by Blasius in 1674 and Boyden in 1926.[3] Gross reported comprehensively on gallbladder anomalies in man and animals in 1936.[1] Gallbladder anomalies, though rare, are often associated with pathology that makes them clinically and anatomically important to the practicing surgeon. There is no record of gallbladder anomalies in Zimbabwe. The current classification of duplicated gallbladder is the work of Boyden.[5]

During gallbladder surgery, failure to detect an accessory gallbladder can result in repeated episodes of cholecystitis in the remaining gallbladder after cholecystectomy. The only case of a ruptured duplex gallbladder in the literature to date was described by AL Moores and SP Gregory in a cat. In this feline case, both gallbladders were extrahepatic and the ruptured moiety was repaired surgically (J Small Animal Practice 2007 Jul; 48(7):404-9).[5] The cat remained ill and was euthanized 72 hours postoperatively at owner's request. The surgical management of repair of a ruptured inflamed gall bladder is of doubtful value in our view.

The intrahepatic position is the second most frequent ectopic location of the gall bladder.[3] It results from failure of the gallbladder to migrate from its embryonic

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intrahepatic location. In adults approximately 60% of intrahepatic gallbladders are associated with gallstones.[7] Intrahepatic ectopic location of the gallbladder makes cholecystectomy a hazardous procedure as a hepatotomy must be done in order to access the gallbladder.[7]

We report a case of duplicated gallbladder with the two components draining through a common cystic duct, one completely intrahepatic and the other completely extrahepatic. The intrahepatic moiety developed gangrenous cholecystitis complicated by rupture and intrahepatic abscess formation. The gangrenous state of the intrahepatic gallbladder with free rupture precluded any consideration of a formal cholecystectomy which would have been necessary had the patient presented earlier during the stage of acute cholecystitis. The extrahepatic component had chronic calculous cholecystitis for which open cholecystectomy was performed.

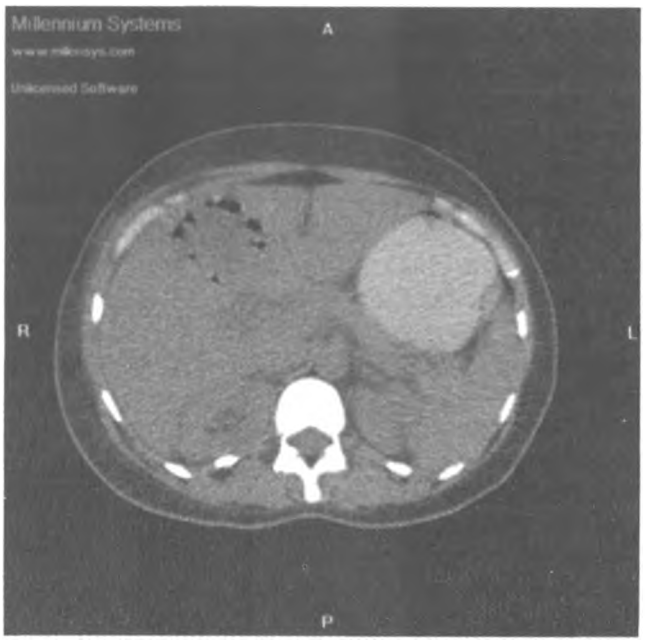
Case Report

A 43 year old female patient, Mrs M.S, presented with a three week history of epigastric pain, nausea, vomiting, and revulsion to fatty foods and generalized body weakness. The epigastric pain was sharp and stabbing and radiated to the back between shoulder blades. The pain worsened over the preceeding six days. There was no history of jaundice, pruritus, pale stools or passage of dark urine. She was hypertensive, well controlled on hydrochlorothiazide 25mg daily and nifedipine 20mg both orally. Clinical evaluation revealed a very ill looking patient with exquisite right upper quadrant tenderness, rebound tenderness and guarding. She was pyrexial, temperature 39 degrees Celcius. Blood pressure 90/60 mmHg, pulse 112 beats per minute. Investigations done comprised full blood count showed neutrophillic leukocytosis. Urea and electrolytes and liver function tests were normal excerpt for a mild ureamia. Plain abdominal X-rays showed a non-specific bowel gas pattern with no pnuemoperitonium. The results of abdominal ultrasound and computerized tomography (CT) are described below.

Radiological findings

Abdominal ultrasound scan done by a radiographer at the referring hospital 5 days prior to admission to our unit was reported thus "liver is of normal size and echotexture, no focal lesions or dilated intrahepatic ducts seen. The gallbladder is not well distended but contains bright echogenic structures? stones...cholelithiasis". A second ultrasound scan done by a Consultant radiologist 5 days later at our request, was reported thus "there was what appeared to be a mass within the liver which contained gas. We could not distinguish whether this was an overlying loop of bowel or within the liver itself hence a CT was recommended". The Radiologist elected to do a computerized axial tomographicstudy with a Multislice Helical scanner with a four phase study (i.e.) non-contrast, arterial and porto-venous phases and delayed phases using Omnipaque 300 intravenous radio-contrast material, 100ml intravenously. This showed "a mass lesion evident in the right lobe of the liver measuring 50mm by 70mm containing gas within it. Pockets of gas seen in the intra-hepatic biliary system especially in the right lobe of the liver but also in the left lobe of the liver. The gallbladder was clearly seen as separate from this mass which on the post venous phase still shows lack of enhancement but a septum appears to be around it. There is gas in the cystic duct and common bile duct is dilated measuring 12mm. The appearance of the mass within the liver is suggestive of a neoplasm but an abscess is also likely. Necrosis and gas within it is more suggestive of an abscess rather than neoplasm. I think a percutaneous aspiration of this abscess can be carried out easily".

Figure I: CT scan showing a gas filled "abscess cavity "in the right lobe of the liver lying quite superficially.



Operative findings

The patient was very sick and heamodynamically unstable and was deemed to be a candidate for percutaneous drainage. This was not available in our hospital, just like the CT scan. The patient was subjected to open drainage since the " hepatic abscess" was quite accessible and contained gas and therefore deemed of a highly virulent gas forming organism. Under general anaesthesia, a right subcostal (Kocher's) incision was made. Foul smelling pus and multiple free floating gall stones were encountered immediately on entering the peritoneal cavity as shown in figure II below.

Figure II: Intraoperative digital photograph of the ruptured intrahepatic gangrenous gallbladder with scattered free floating gallstones in the free peritoneal cavity.

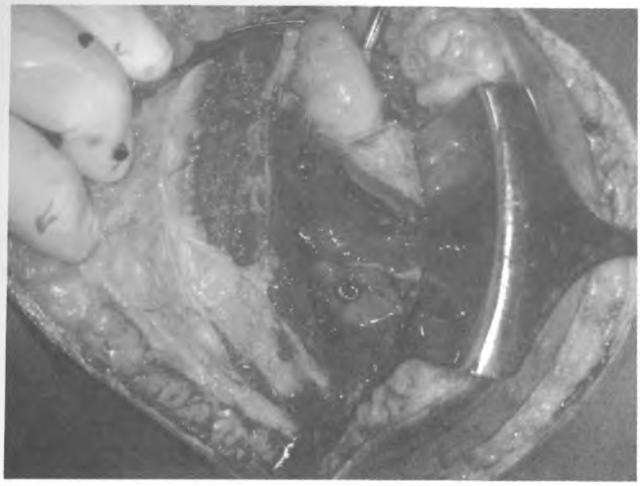
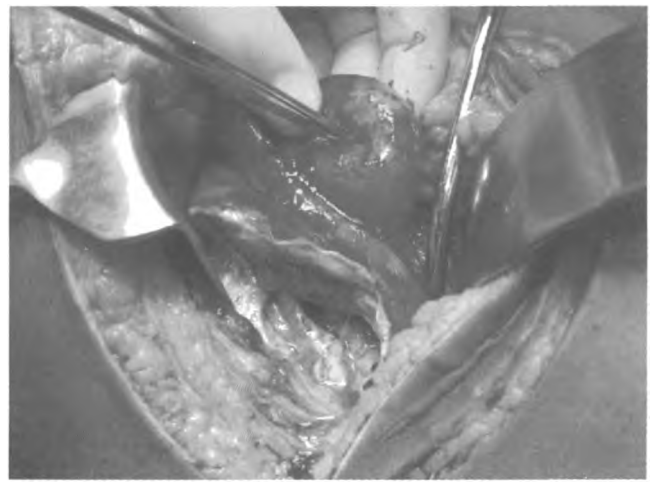


Figure III: Intraoperative digital image showing the gangrenous ruptured intrahepatic gallbladder moiety separated from the stone packed extrahepatic gallbladder by a rind of normal looking liver tissue.



Omentum and small bowel loops were congregated around a ruptured right lobe liver abscess cavity containing multiple mixed gall stones over segment IV. The intrahepatic gallbladder was separated by a 1cm rind of healthy looking liver tissue, from a thick walled extrahepatic gallbladder packed with stones as shown in Figure III below. There was a direct communication between the deep part of the cavity and a duct which met with another duct from the extrahepatic gall bladder to form a common cystic duct outside the parenchyma of the liver. Though strongly desired and indicated, an ontable cholangiogram could not be performed for logistical reasons. A retrograde cholecystectomy was performed. A single cystic artery distally dividing into two terminal branches, one for each gallbladder was identified, suture ligated and divided. The intrahepatic gallbladder being gangrenous, was subjected to debridement and by curettage, an incisional biopsy of parts of its wall was made for histological evaluation, pus swabs taken for microbiological investigations. The abscess cavity/intrahepatic gallbladder bed was loosely packed with a loop of omentum passing through the defect through which the duct draining the intrahepatic moiety to the common cystic duct traversed the hepatic parenchyma. All free floating gall stones were collected from the peritoneal cavity. The peritoneal cavity was thoroughly washed with warm sormal saline solution. A Latex tube drain was inserted in the subhepatic space and exteriorized via a separate stab incision in the right lumbar area. The wound was closed by the mass closure technique using 1PDS continuous sutures following the achievement of heamostasis. The skin was closed with 2/O nylon vertical mattress sutures.

Postoperatively the patient was admitted to the intensive care unit for heamodynamic monitoring, stabilization, fluid resuscitation, ventilation and continuation of intravenous antibiotic administration and analgesia. Histology from the abscess wall confirmed gangrenous cholecystitis and that from the extrahepatic gallbladder moiety confirmed chronic cholecystitis with acute inflammation of the serosa. No organisms were cultured from the peritoneal swabs.

Literature Review

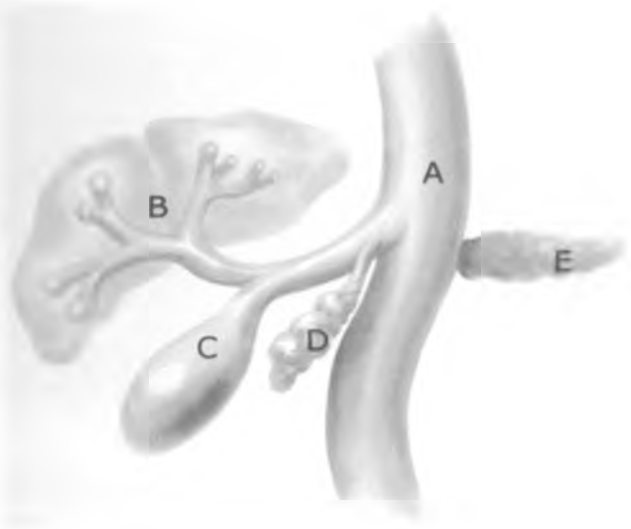
1.0 Epidemiology

Gallbladder anomalies are rare.[1] The occurrence of pathology within anomalous gallbladder(s) is an even rarer event. The anomalies of the gallbladder are of three types.[3] To understand these congenital abnormalities it is prudent that the embryology of the gallbladder be revisited first.

1.1 Embryology of the gallbladder

The hepatobiliary system develops from the ventral bud or the hepatic diverticulum from the caudal foregut during the fourth week of embryonic life. The hepatic diverticulum divides into two parts, the pars hepatica and the pars cystic. The pars hepatica gives rise to the liver, common hepatic ducts and intrahepatic ducts. The pars cystic gives rise to cystic diverticulum, which gives rise to the gallbladder and the cystic duct. The hepatic diverticulum then elongates to form the common bile duct. The structures are initially solid and become canalized by the eighth week of gestation. The embryology of the gallbladder, liver and biliary system is illustrated diagrammatically below.

Figure IV: Embryology of the gallbladder. The figure illustrates the normal embryologic development of the gallbladder and bile ducts. Foregut (A), the cranial end of the hepatic diverticulum, which represents pars hepatica (B) and the cystic diverticulum (C). The ventral (D) and dorsal (E) pancreas are also shown.[10]



During the early embryonic life, the gallbladder is initially intrahepatic in location. It then moves from its intrahepatic position in the second month of gestation to its normal superficial extrahepatic position.

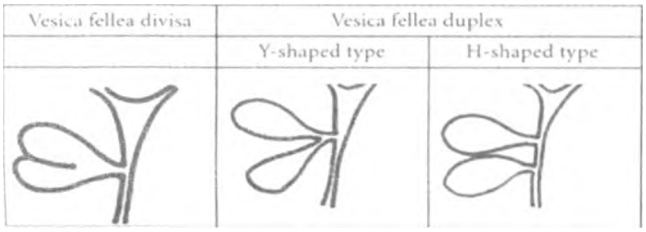
1.2 Classification of Gallbladder Anomalies

Three major types of gallbladder anomalies have been described.

1.2.2 Anomalies of number of the gallbladder

Anomalies of number of the gallbladder include agenesis. Congenital absence of the gallbladder is rare and an incidence of 0.003% has been reported. Duplication of the gallbladder is a rare congenital anomaly, occurring in about one per 4000 births.[5] It is thought to be due to exuberant budding of the developing biliary tree when the caudal bud of the hepatic diverticulum divides.[9,11] The first reported human case was noted in a sacrificial human victim of ancient Roman Emperor Augustus in 31 BC.[10] Anatomic variants of gallbladder duplication are differentiated according to Boyden's classification.[5,9]

Figure V: Boyden's classification of gallbladder duplication.



In vesica fellea divisa there is a bilobed or bifid gallbladder or double gallbladder with a common neck. In vesica fellea duplex there is a double gallbladder with two cystic ducts), either (i) Y-shaped type (the two cystic ducts uniting before entering the common bile duct), or (ii) H-shaped type (ductular type, the two cystic ducts entering separately into the biliary tree).Triple gallbladder has also been described with successfullaparoscopic cholecystectomy for the triplicate.[8]

1.2.3. Anomalies of gallbladder formation

Anomalies of gallbladder formation include the Phrygian cap, bilobed, hourglass, diverticulum, and rudimentary gallbladder.

2. Complications associated with gallbladder anomalies

An intrahepatic gallbladder usually has impaired function because it does not empty completely. This may result in gallstone formation due to stasis. In adults approximately 60% of intrahepatic gallbladders are associated with gallstones.[7] They may also cause diagnostic challenges especially if they develop gangrenous calculus cholecystitis as they may simulate a liver abscess in their presentation. Laparoscopic cholecystectomy can be a great challenge in these cases consequent upon the need for hepatotomy. Anomalies in position or formation can result in abnormal anatomy which must be appreciated during surgery. On table cholangiography is highly desirable to elucidate the anatomy in cases of doubt even with singleton gallbladder and becomes even more indispensable in cases of duplication. Repeated subsequent unexpected episodes of acute cholecystitis can follow cholecystectomy if only one of the duplex gallbladder complexes has been removed.

3. Diagnosis and Management of gallbladder anomalies

Most gallbladder anomalies are asymptomatic and are discovered incidentally during imaging of the abdomen. Congenital anomalies of the gallbladder and anatomical variations of their positions are associated with an increased risk of complications after laparoscopic cholecystectomy.[6,12] Preoperative imaging should be helpful for diagnosis .Those that present with complications can be diagnosed by imaging. Intrahepatic gallbladders can be readily imaged by either ultrasonography or CT scan. The condition may be suspected, if the cholecystogram, ultrasound or a CT scan reveals a gallbladder in an unusually high location. Diagnosis of a duplex gallbladder with ultrasound can be challenging. Magnetic resonance cholangiography is superior to ultrasound scan in diagnosing gallbladder anomalies.[13]

Most intrahepatic gallbladders are only partially embedded within the hepatic parenchyma andwhen can usually be easily identified at the time of cholecystectomy. Those that are completely buried

within the liver may be a challenge for the general surgeon. A completely embedded gallbladder is best approached by first identifying the cystic duct where it joins the common hepatic duct and then following the cystic duct back to the gallbladder. An open cholecystectomy is a safer than a laparoscopic cholecystectomy procedure for an intrahepatic gallbladder.[14] Laparoscopic cholecystectomy with intraoperative cholangiography seems to be the appropriate treatment for duplicated gallbladder provided both moieties are extrahepatic.[14]

Discussion

A fortuitous encounter with a congenital gallbladder anomaly comprising of a duplex gallbladder malformation of the Boyden Y type complicated by acute ruptured gangrenous calculous cholecystitis of the intrahepatic moiety of the duplex malformation masquerading as a liver abscess by a gas forming bacterial organism and pneumobilia is presented. This was not a classical Boyden Y type since the second gallbladder was embedded in the liver parenchyma. The intrahepatic location of the second gallbladder causes significant difficulty during surgery mainly because the organ was already ruptured. The diagnostic dilemma during the preoperative period was occasioned by the difficulty in interpreting the radiological findings, no doubt due to the rarity of the condition. The definitive diagnosis was only made upon thorough scrutiny at laparotomy. The intraoperative findings came as a total surprise to the surgical team. A significant amount of time was spent trying to figure out the anatomy. An on-table cholangiogram would have gone a long way in elucidating the anomalous anatomy. Unfortunately due to logistical problems, this could not be done. Fortunately the intraoperative images presented here are more eloquent than any narrative description as they clearly show the ruptured gangrenous intrahepatic moiety separated from its extrahepatic fellow by a rind of relatively normal looking liver parenchyma. This makes for an incontrovertible illustration of the pathological findings. The diagnosis is further supported by the finding of stones within the cavity and the histological confirmation of the intrahepatic gallbladder wall. Had the ultrasound been performed by a specialist radiologist earlier in the course of the disease, it is likely that a preoperative definitive diagnosis would have been made prior to rupture of the gangrenous intrahepatic component .The fact of an ectopically positioned intrahepatic gallbladder per se is not surprising as the embryological origins, though rare, are well described .The surgical decision making was not difficult once the anatomy and the pathology had been appreciated. Cholecystectomy was performed on the gallstone studded extrahepatic gallbladder moiety because there is well documented incidence of cholecystitis in the post-operative period if one moiety is left in situ.[15] This will necessitate a re-operation. What posed difficulty was how to deal with the ruptured gangrenous intrahepatic moiety? We elected to be less aggressive by simply debriding the gall bladder wall, making no attempt at comprehensively removing the whole wall for fear of causing bleeding in an already inflamed liver. In cases where the intrahepatic gallbladder is diagnosed unruptured pre-operatively, open cholecystectomy as opposed to laparoscopic is the procedure of choice. The use of omentum was prudent as this has been shown to be beneficial in cases of residual liver cavities following abscess or cyst drainage. The patient recovered well and was discharged.

It is likely that the intrahepatic gallbladder would have been missed, with disastrous consequences had it not undergone intrahepatic abscess formation secondary to inflammation, gangrene and perforation.

This case serves as an interesting reminder of the need for surgeons and radiologists to always remember the reality and complexity of congenital malformations of the biliary tree. The literature is awash with anecdotal cases of duplex gallbladder patients having cholecystectomy of only one gall bladder, the second one being missed, continuing to cause symptoms of fever, right hypochondria pain and tenderness , only to be discovered later and the patient having a second operation to remove the second gallbladder. Although a rare condition, this paper illustrates the importance of maintaining a high index of suspicion of duplex gallbladders. On-table cholangiography though not used in this case due to unavailability is advocated as an efficient and definitive way of elucidating the extrahepatic biliary anatomy especially in anomalous cases like this one.

Conclusion

Successful management of a rare congenital malformation of the gallbladder complicated by the pathology of cholelitiasis and its complications has been presented. A high index of suspicion and awareness of congenital biliary tree anomalies should be maintained if these conditions are to be managed successfully. A comprehensive internet literature search using Pub Med Central failed to find any case like the one described here. We conclude that this is the first case of its kind described in world literature. The multiplicity of concurrent problems made it difficult to coin a terse terminology for the described condition. Accurate radiological imaging techniques play a major role in the management of congenital anomalies and pathology of the biliary tree especially as regards planning surgical strategy. Cholecytitis complicating intrahepatic gallbladder is difficult to treat surgically. Open as opposed to laparoscopic cholecystectomy is the surgical technique of choice as employed in the case presented. The management of a gangrenous ruptured intrahepatic gallbladder has never been described before. The minimalistic approach presented here seems appropriated for fear of causing damage to the

liver parenchyma with the attendant risk of heamorrhage in a heamodynamically unstable patient.

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