IJNRD.ORG ISSN : 2456-4184
INTERNATIONAL JOURNAL OF NOVEL RESEARCH AND DEVELOPMENT (IJNRD) | IJNRD.ORG
An International Open Access, Peer-reviewed, Refereed Journal

GALLBLADDER AGENESIS IN AN ADULT FEMALE PATIENT DIAGNOSED BY MAGNETIC RESONANCE CHOLANGIOGRAPHY: REPORT OF A CASE AND REVIEW OF THE LITERATURE

Case Report of Gall Bladder Agenesis in Adult Indian Female

[1]Dr Neeraj Goyal, [2]Dr Shraiya Tiwari, [3]Dr Varnika Gupta

[1]Principal Consultant Surgeon

[1-3] Max Superspeciality Hospital, Patparganj, New Delhi, India

Abstract: Gallbladder agenesis (GA) is a congenital entity with an incidence of around 1 per 6500 live births globally. The majority of the patients, estimated between 50 to 70 percent, remain asymptomatic while those who are symptomatic report symptoms mimicking biliary colic. The ultrasonography can be misleading or inconclusive in the initial workup for suspected gallbladder pathology. Furthermore, modern diagnostic techniques such as the hepatobiliary iminodiacetic acid (HIDA) scan and endoscopic retrograde cholangio-pancreatography (ERCP) may show no gallbladder visibility, leading to an incorrect diagnosis of cystic duct obstruction rather than GA. As a result, some GA patients are only diagnosed during surgery. Surgery can be dangerous in these people because needless dissection in search of a non-existent gallbladder can harm the biliary network, hepatic veins, or small bowel. As a result, doctors should keep GA on their differential diagnosis list and consider imaging modalities like magnetic resonance cholangiopancreatography (MRCP) if other tests are equivocal. We report a 56-year-old female presenting with upper abdominal pain, bloating, and dyspepsia later reported as cholelithiasis. She underwent MRCP to confirm the diagnosis of GA. No filling defect was detected or calculus was seen.

Index Terms **- Gallbladder Agenesis, Bile System, Magnetic Resonance with Cholangiopancreatography, No Gall bladder, Hepatobiliary iminodiacetic acid scan, Symptoms mimicking biliary colic.**

Introduction Gallbladder agenesis (GA) is an uncommon biliary system congenital abnormality. Bergman characterized it for the first time in humans in 1702, and it has subsequently been recorded multiple times in case reports [1].GA's cause is uncertain; it is generally a spontaneous occurrence with no obvious cause. However, some families have had the illness in many members, implying that there are familial hereditary types of GA.

Because gallbladder agenesis is caused by a defect in embryonic development, most occurrences of gallbladder agenesis are linked to other congenital abnormalities, such as those of the bile system [2]. It occurs in around one-sixth of all cases of biliary atresia; the absence of the gallbladder and cystic duct on their own is uncommon. The average rate of gallbladder agenesis at birth is roughly 0.02 percent (A6), with varying penetrance and no sex-linked characteristics [3].

In 23% of cases, patients develop symptoms, and GA is almost usually misdiagnosed as cholecystitis with cystic duct obstruction or sclero-atrophic gallbladder, resulting in unnecessary surgery. The treatment of GA is conservative with antispasmodic drugs.

Because of the vague character of the symptoms, establishing a reliable preoperative diagnosis of GA in symptomatic individuals is difficult. We present a case of congenital GA in an adult patient with cystic duct absence and a biliary tract anomaly discovered by Magnetic Resonance Cholangiopancreatography (MRCP). The genesis and physiopathology of this anomaly, as well as the diagnostic methods used, are also discussed [4].

In this article, we discuss a case report of GA that was accurately diagnosed preoperatively in India using radiological imaging, specifically MRCP. This is one of the rare examples reported in the literature that we are aware of.

CASE REPORT

A 56-year-old woman came to our attention for upper abdominal pain, bloating, and dyspepsia for nearly last 15 days. Her blood pressure and pulse rate were regular and her body temperature was 36.7◦C, and other vitals were also normal.

Results of all hematological and biochemical investigations were within normal limits. Ultrasonography (US) examination did not visualize the gallbladder clearly (Figure 1). MRCP was subsequently performed with a 1.5 Tesla magnet (Philips Gyroscan Intera; Best, Medical Systems, Netherlands), equipped with a Master dynamic gradient system (30 mTm maximum power and 150 mTm/msec slew rate) using a phased array body coil.

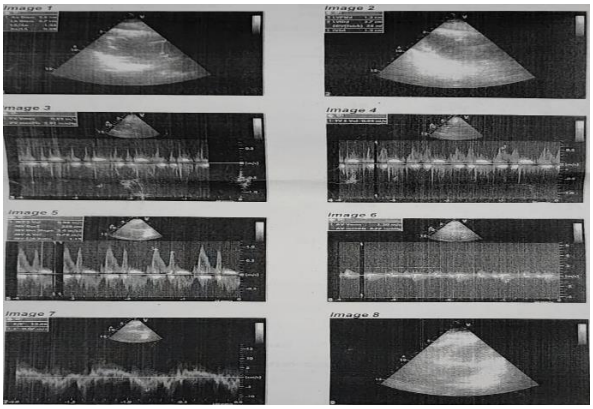

Figure 1

The patient underwent the MRCP study after fasting for 8 hours. The examination protocol consisted of an axial T1-weighted 2D FLASH and axial T2-weighted TSE sequences and axial SPIR to localize the acquisition volume for the MR Cholangiography sequences. We performed BALANCE sequences to obtain an accurate anatomical resolution and gallbladder fossa visualization. The MRCP study consisted of a 3D-MIP breath-hold acquisition of a single slice in the coronal plane, positioned so as to obtain a complete visualization of the intra- and extrahepatic biliary tract, with a single-slab RARE sequence.

The MRCP showed the absence of both the gallbladder and the cystic duct. Bile duct was prominent measuring 7mm with no intrahepatic biliary radical dilation or distal intraluminal obstructing lesion. Pancreas was normal in course and calibre. No filling defect was detected or calculus was seen.

The intrahepatic bile ducts appeared normal with no images of stenosis. Moreover, MRCP demonstrated an anatomical variation of choledochopancreatic duct junction. In fact, the bile duct was visualized up until the second part of the duodenum, in correspondence to the Major papilla. The patient underwent medical treatment with complete control of symptoms

DISCUSSION

Anatomic malformations of the biliary tract are common, but gallbladder and cystic duct agenesis is uncommon; it is frequently identified by chance and is usually asymptomatic. GA can be found in both children and adults, with an average age of 46 at the time of diagnosis. It's frequently discovered by chance after abdominal surgery or at an autopsy. The prevalence rate varies between 0.007 and 0.13 percent. In surgical cholecystectomy series, the incidence of this deformity is slightly lower (0.007–0.027 percent) than in autopsy reports. GA detected during surgery had a three-to-one female predominance, but cases discovered in autopsies have an equal male-to-female ratio. In the fourth week of pregnancy, the gallbladder develops from the caudal section of the hepatic diverticulum.

There are two theories regarding nondevelopment of the gallbladder. According to one theory, the foregut's hepatic diverticular bud does not grow properly into the gallbladder and cystic duct [4]. The other theory is that after solid-phase development, the cystic duct and gallbladder fail to recanalize. When the cystic bud does not develop, isolated GA occurs. Because cystic bud growth impairs development between the sinus venus cordis and paired omphaloenteric and umbilical veins, GA is frequently associated with cardiovascular and gastrointestinal problems. In 70–82 percent of instances, GA occurs on its own (31.6 percent asymptomatic cases and 55.6 percent symptomatic cases). In the remaining 12.8–30% of instances, it is associated with additional malformations, which are divided into two subgroups: those with bile duct atresia or choledochal cyst (9%) and those with normal bile ducts but distant numerous foetal defects (12.8–21%) [4].

In new-borns, GA is associated with one or more defects, some of which are life-threatening. GA has been linked to a variety of gastrointestinal, skeletal, cardiovascular and genito-urinary malformations, including ventricular septal defect, imperforate anus, duodenal atresia, gut malrotation, pancreas divisum, right hepatic lobe hypoplasia, hepatic flexure duplication cysts, renal agenesis, undescended testes and syndactyly. Patients become symptomatic in roughly 23% of cases, despite the fact that GA has no distinct symptomatology [5]. 90 percent of cases have right upper quadrant abdominal pain, 66 percent have nausea and vomiting, 37 percent have fatty food intolerance, 30 percent have dyspepsia, and 35 percent have jaundice. These symptoms, which are similar to other common biliary tract conditions often lead to misdiagnosis of this condition.

Most of the adult patients with GA are asymptomatic. The symptoms may be secondary to concomitant biliary pathologies such as primary duct stones and biliary dyskinesia (patients may have a congenital abnormality of function in the form of a significant higher sphincter of Oddi resting pressure and an increase in the proportion of retrograde propagation of phasic muscular contraction with regurgitation of pancreatic or duodenal contents), or it may be related to nonbiliary causes such as esophagitis and duodenitis [6].

The different clinical presentation of GA reflects the variety of embryological anomalies and were classified by Bennion et al into three categories:

1) Association with other congenital malformations often leads to perinatal death (12.8-30%),
2) Asymptomatic cases (31.6%),
3) Symptomatic patients (55.6%).

Recently, Tang et al proposed a new classification including symptomatic subtype 1a GA accompanied by lethal deformities such as biliary atresia, imperforate anus, ventricular septal defect, duodenal atresia (majority of these die after birth), subtype 1b which is accompanied by nonlethal deformities such as intestinal malrotation, right liver agenesis, cryptorchidism and choledochal cyst [7].

Reviewing the literature, it was found that, with the exception of two cases of GA, symptomatic patients are still unnecessarily operated on because the preoperative investigations carried out failed to demonstrate the exact diagnosis. GA represents a difficulty for the surgeon during laparoscopic surgery, the biliary or portal structures can easily be wounded during dissection as one searches for a gallbladder that does not exist. If the diagnosis of GA is made during the operation, the surgeon must prove GA by thoroughly examining the most common sites for ectopic gallbladder, which are intrahepatic, retrohepatic, on the left side, or within the leaves of the lesser omentum or within the falciform ligament, retroduodenal, retropancreatic, and retroperitoneal. The absence of normal anatomical structures and the inability to pull on the gallbladder to dissect the triangle of Callot represent a risk of iatrogenic injury, and it is the most common cause of conversion from a laparoscopic procedure to a traditional open laparotomy [8].

In our patient, MRCP has been used in addition to other diagnostic methods. MRCP is a noninvasive and well-demonstrated imaging method in the evaluation of the biliary tract. As it does not require contrast administration to visualize the bile, it is not compromised by biliary stasis. It can also demonstrate an excluded and/or ectopic gallbladder. In our case, MRCP allowed to make the correct preoperatively diagnosis with a noninvasive examination, avoiding unnecessary surgical exploration, and minimizing the risk of complications. Moreover, it provided accurate anatomical details about the bile tree conformation excluding the condition of ectopic gallbladder too. In conclusion, GA should be kept in mind whenever the gallbladder is improperly visualized in routine imaging methods in patients with biliary-type pain.

Although the MRCP approach may not yet be able to completely replace ultrasound as the gold standard for acute gallbladder imaging, it has proven to be an excellent supplement to inconclusive ultrasonographic tests. The precise preoperative diagnosis of GA is critical in order to avoid unnecessary and perhaps dangerous surgical exploration.

CONCLUSION

In conclusion, Gall Bladder agenesis presents a significant diagnostic challenge. Clinicians should have a strong index of suspicion if gall bladder is not visualised on ultrasound. MRCP is considered as a test of choice in case of suspicion. It can also demonstrate other anomalies of the biliary tract.

Management of Gallbladder agenesis is conservative and an accurate preoperative diagnosis will avoid unnecessary surgical intervention.

I. ACKNOWLEDGMENT

The authors would like to acknowledge the technicians of the laboratory of our hospital department for their help in offering me the resources in running the program and staff involved in this case report. Authors would also like to extend their gratitude to the medical writing team of Quod Genesis for their support.

CONFLICT OF INTEREST

Any of the authors have no conflict of interest.

REFERENCES

1. Latimer EO, Mendez Jr FL, Hage WJ. Congenital absence of gallbladder: report of three cases. Annals of surgery. 1947 Aug;126(2):229.
2. KOBACKER JL. Congenital absence of the gall bladder—a possible hereditary defect. Annals of Internal Medicine. 1950 Oct 1;33(4):1008-12.
3. Rabinovitch J, Rabinovitch P, Rosenblatt P, Pines B. Congenital anomalies of the gallbladder. Annals of Surgery. 1958 Aug;148(2):161.
4. Fiaschetti V, Calabrese G, Viarani S, Bazzocchi G, Simonetti G. Gallbladder agenesis and cystic duct absence in an adult patient diagnosed by magnetic resonance cholangiography: report of a case and review of the literature. Case Reports in Medicine. 2009 Jan 1;2009.
5. Bedi N, Bond-Smith G, Kumar S, Hutchins R. Gallbladder agenesis with choledochal cyst—a rare association: a case report and review of possible genetic or embryological links. Case Reports. 2013 Jan 9;2013:bcr2012006786.
6. Belli G, D'agostino A, Iannelli A, Rotondano G, Ceccarelli P. Isolated agenesis of the gallbladder. An intraoperative problem. Minerva Chirurgica. 1997 Sep 1;52(9):1119-21.
7. Tang LM, Wang XF, Ren PT, Xu GG, Wang CS. The diagnosis of gallbladder agenesis: two cases report. International Journal of Clinical and Experimental Medicine. 2015;8(2):3010.
8. Peloponissios N, Gillet M, Cavin R, Halkic N. Agenesis of the gallbladder: a dangerously misdiagnosed malformation. World journal of gastroenterology: WJG. 2005 Oct 21;11(39):6228