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# "DOUBLE GALL BLADDER WITH CHOLESTEROLOSIS, MUCOCELE AND ADENOMYOMATOUS HYPERPLASIA": A RARE ENTITY

Pathology

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## ABSTRACT

Double gallbladder is a rare congenital anomaly with an incidence ratio of 1:4000. Congenital anomalies of the gallbladder and anatomical variations of their positions are associated with an increased risk of complications. Pre- operative imaging will be helpful for diagnosis. Laparoscopic cholecystectomy of both the gallbladder with intra-operative cholangiography seems to be the appropriate treatment. Hereby we report a rare case of double gallbladder in a young girl, presented with abdominal pain and underwent cholecystectomy. Histopathology revealed very interesting and several findings.

## KEYWORDS

Double Gallbladder, Cholesterolosis, Mucocele, Adenomyomatous hyperplasia.

## INTRODUCTION:

Double gallbladder (GB) is a rare congenital anomaly with an incidence ratio of 1:4000.[1] Congenital anomalies of the GB and anatomical variations of their positions are associated with an increased risk of complications.[2] Double GB is classified according to Boyden's classification and the two main types: (i) Vesica fellea divisa or bilobed GB, and (ii) Vesica fellea duplex or true duplications with two different cystic ducts. The true duplications are further classified into two types: Y shaped type (two cystic ducts unite before entering the common bile duct (CBD). Usually the two GBs are adherent and occupy the same fossa. (b) H shaped type (two separate GB and cystic ducts entering separately into the CBD.[2] Pre- operative imaging will be helpful for diagnosis. Laparoscopic cholecystectomy of both the GB with intra-operative cholangiography seems to be the appropriate treatment.[3] Hereby we report a rare case of double GB in a young girl, presented with abdominal pain and underwent cholecystectomy. Histopathology revealed several interesting findings.

## Case details:

A 17 year young girl presented to surgery out-patient department with chief complains of recurrent abdominal pain in the right upper quadrant and intermittent vomiting for the past three months. She presented with colicky type of pain which aggravated on taking meals and getting relieved on fasting. She also had intermittent fever and anorexia, but no jaundice. She had several episodes of vomiting but normal bowel and bladder habits. General physical examination revealed no other significant findings. All her vitals were normal and stable. On inspection, abdomen was flat, and no dilated veins. On palpation of abdomen, severe tenderness noticed over right hypochondriac region. Percussion revealed no free fluid. On auscultation, normal bowel sounds were heard. Murphy's sign was positive and suggestive of gall bladder pathology. All routine investigations were within normal limits including the liver function tests. With provisional clinical diagnosis of chronic cholecystitis, patient was subjected for abdominal and pelvic ultrasonography. USG abdomen revealed presence of two cystic structures in the gallbladder fossa (Fig no 1).

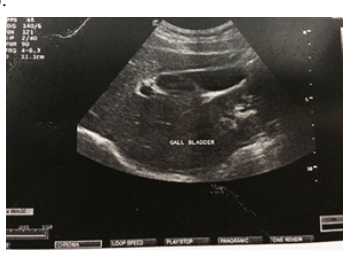



**Fig No 1: USG abdomen revealed presence of two cystic structures in the gallbladder fossa.**

One larger gall bladder measured 10.5x3 cm, and smaller oval shaped gall bladder seen near neck of larger one and it measured 1.9x1.2cm. Wall thickness of both the gallbladders was comparably equal and no stones were seen. Other parts of the extra hepatic biliary tract were normal and no significant findings seen. Based on these investigation findings, pre-operative diagnosis of double gallbladder with chronic cholecystitis was made and she was posted for surgery. Laparoscopic cholecystectomy was done with the excision of both the gallbladder. Patient recovered well in post-operative period and discharged after 7 days. Six months follow-up period was uneventful.

The gross examination of resected specimen revealed presence of two gallbladders with the smaller gallbladder attached to the neck and medial aspect of the larger gallbladder (Fig no 2).

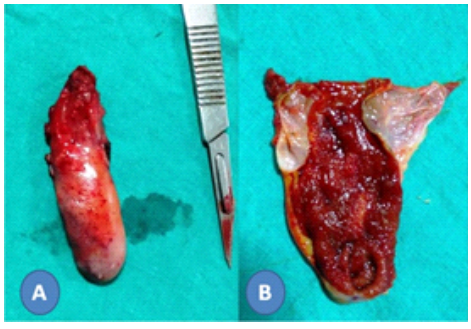



**Fig No 2: Gross specimen revealed presence of two GB with the smaller GB attached to the neck and medial aspect of the larger GB (A). Cut section of larger GB showed reddish velvety mucosa with yellow specks and smaller GB showing mucocele (B).**

Cut section of larger gall bladder showed reddish velvety mucosa with yellow specks scattered diffusely over the mucosa. No gall stones were found. Cut section of smaller gallbladder revealed mucin filled lumen due blockage of the opening by thick mucous plug leading to formation of mucocele. Both gallbladders had a common dividing wall.

Microscopic examination from larger gall bladder revealed lipid laden foamy macrophages filled in the subepithelial tissue and entire lamina propria (Fig no 3) suggesting diagnosis of Cholesterolosis. There was associated villous mucosal hyperplasia. There were Rokitansky Aschoff sinuses where mucosa was dipping deep into muscle layer in extensive manner suggesting diagnosis of adenomyomatous hyperplasia. Whereas sections from the smaller gall bladder revealed features of mucocele of gall bladder with presence of thick mucinous secretions in the lumen. Some of the detached mucosal portions were seen floating within these mucinous secretions. There was minimal

dysplasia in the lining epithelium and focal stratifications of the lining epithelial cells. Subepithelium showed focal dense lymphocyte and plasma cell infiltrations. The thickened wall of gall bladder showed fibrosis and extensive Rokitansky Aschoff sinuses. We offered diagnosis of double gallbladder with larger gallbladder showing Cholesterolosis and adenomyomatous hyperplasia and smaller gallbladder with Mucocele, mild chronic cholecystitis and adenomyomatous hyperplasia.

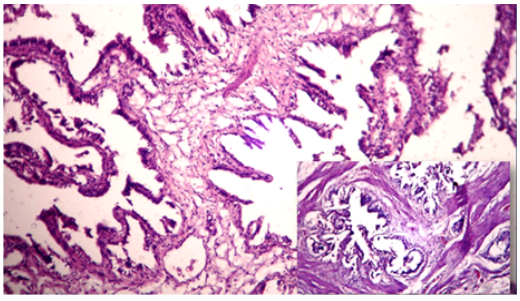


**Fig No 3: Microscopy of larger GB: Lipid laden foamy macrophages filled in mucosa suggesting Cholesterolosis with villous mucosal hyperplasia. Inset showing extensive Rokitansky Aschoff sinuses. (H & E stain, 10x)**

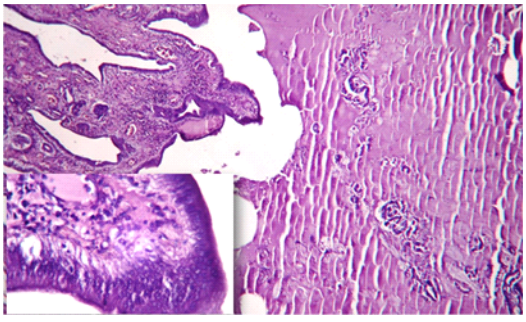
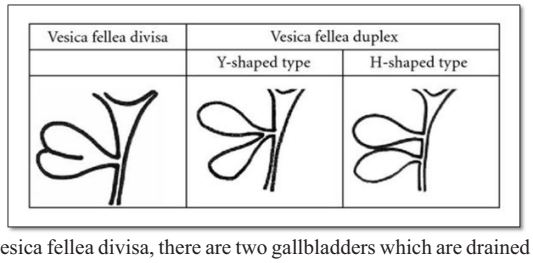


**Fig No 4: Microscopy of smaller GB: Mucocele with presence of thick mucinous secretions in the lumen. (H & E stain, 10x). Inset showing mild dysplasia in the lining epithelium and stratification with dense lymphocyte and plasma cell infiltration. (H & E stain, 40x)**

## DISCUSSION:

Double gallbladder is a rare congenital anomaly with an incidence of 1:4000.[1] However, the exact incidence of this rare anomaly cannot be accurately assessed, since the only cases which can be identified are those that have become symptomatic or incidental findings during radiological investigation or surgery or autopsy. There are around 62 cases of double GB in the review of literature.[6] It is important to diagnose this anomaly pre- operatively to avoid damage to ductal and arterial system during surgery.[3]




In vesica fellea divisa, there are two gallbladders which are drained by a single cystic duct. In vesica fellea duplex, there are two gallbladders which are drained separately by their own cystic duct. It is again sub classified into Y shaped type (two cystic ducts joins to form a common cystic duct before opening into the common hepatic duct) and H shaped type or ductular type (two cystic ducts drain separately into the common hepatic duct). So according to this classification, our case of double gallbladder comes under vesica fellea divisa due to the presence of single cystic duct.

During the fifth or early sixth week of intrauterine life, the distal part of the cystic duct might have divided resulting in the formation of double gallbladder. Of these two, one gallbladder might have undergone normal growth and the other was underdeveloped.[3]

The true duplication is more common and occurs due to bifurcation of GB primodium during the 5th and early 6th week of embryonic life. There are nearly 41 group enriched genes which encode for the development of the gallbladder. Some of them are FGF19, CHST4, MOGAT1 and MUC5B. A common variant of the sterol transporter encoded by the ABCG8 gene is associated with an increased risk for the development of cholesterol stones.[4]

Double GB does not presents with any specific signs or symptoms and incidence of the disease is similar to the normal variant. Normally one GB functions actively till adulthood and later may show predisposition to inflammation and the other GB remains as a mucocele, as seen it happened in our case.

Cholelithiasis is most common complication in double GB, usually involving only one GB, though both may be involved. There is no increase in incidence of disease in the double GB, so prophylactic cholecystectomy is not recommended. In symptomatic patient cholecystectomy is recommended with excision of both the GB, even if disease is present in only one GB. Complete preoperative evaluation of the anatomy is a must to avoid the potential damage to the duct system.[6]

Differential diagnosis includes gallbladder diverticula, gallbladder fold, Phrygian cap, choledocal cyst, pericholecystic fluid, focal adenomyomatosis, and intraperitoneal fibrous bands. There are no specific symptoms associated to double gallbladder. The patients can present with the signs and symptoms of acute or chronic cholecystitis, cholelithiasis, empyema, torsion, cholecystocolic fistula and rarely adenocarcinoma.[5]

Ultrasonography is the initial modality that can diagnose this entity. Computed tomography and MRCP being noninvasive are the preferred modalities for characterization of this anomaly.[7] If biliary anatomy is delineated preoperatively, serious biliary and vascular injuries can be prevented during surgery. Intra-operative cholecystography and cholangiography is essential to delineate the ductal anatomy and to help identify the additional anomalous structures. Cholecystectomy can be done by either open or laparoscopic approach depending on the expertise.[8,9] Laparoscopic cholecystectomy is the gold standard treatment and can be done safely in double GB.[10]

## CONCLUSION:

Double gallbladder is a rare congenital anomaly. Pre- operative diagnosis of the anatomy of the double gallbladder is important to avoid damage to biliary ductal and arterial system during surgery. In symptomatic patients, cholecystectomy is recommended with the excision of both the gallbladder. Unless symptomatic, the prophylactic cholecystectomy is not recommended.

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