Case Report

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Adenocarcinoma of gall bladder in a patient with situs inversus totalis: a very rare clinical case

Abstract

Introduction
Situs inversus totalis represents a rare congenital anomaly in which the thoracic and abdominal viscera are transposed to the opposite side through the sagittal plane. It often occurs concomitantly with other disorders that present a challenge for making the diagnosis and management of abdominal pathology. The relationship between situs inversus totalis and cancer remains unclear. This study reports a rare case of adenocarcinoma of the gall bladder in a patient with situs inversus totalis.

Case report
We report a first clinical case of a 50-year-old man with situs inversus totalis and adenocarcinoma of gall bladder, which was successfully treated with surgery and doing well with no evidence of local recurrence or metastasis during six months of follow-up.

Conclusion
The occurrence of gall bladder adenocarcinoma in a patient with situs inversus totalis accounts as a very rare occurrence, as no case is reported till date. In this context, when the tumour is resectable, surgical management should be considered and must be preceded by a careful preoperative staging and full adaptation to the mirror image anatomy.

Introduction
Situs inversus totalis (SIT) is a rare congenital condition with autosomal recessive inheritance and it occurs in the range of 1:8000–1:25000 of the normal population. The male to female ratio is 1:1 with no racial predilection. It was first reported in humans by Fabricius in 1600. Most of the individuals with SIT are asymptomatic and are detected accidentally at the time of clinical examination and radiological investigations for some other condition. It is not considered to predispose to cancers; however, rare cases of malignancies of lung, stomach, liver, common bile duct, colon, rectum, kidney and gastric lymphoma have been reported in the literature.

Gall bladder cancer (GBC) is a rare and usually aggressive malignancy occurring predominantly in the sixth and seventh decade of life, with a strong predilection for females (F>M = 2–3:1). It is one of the most common malignancies in North India, particularly in females. It is not usually found until it has become advanced and becomes symptomatic. Risk factors for GBC are gallstones (size >3 cm), porcelain gall bladder, adenomatous gall bladder polyps (size >10 mm), Salmonella typhi infection, choledochal cysts, primary sclerosing cholangitis, anomalous pancreato-biliary junction, obesity and family history. Histologically, adenocarcinoma is the most commonly isolated variety (>90% cases) followed by squamous cell cancer. Management of this malignancy requires thorough preoperative assessment; knowledge of anatomical variations and their complications, a strategic management plan and vigilant perioperative care to bring out best clinical outcomes. We herein report a rare case of moderately differentiated gall bladder adenocarcinoma in a 50-year-old man with SIT who presented at our institution.

Case report
A 50-year-old male, with known SIT presented at the surgical outpatient department of our institution in January 2013, with a history of intermittent dull aching pain in left upper abdomen for last six months followed by appearance of a hard lump in the left upper abdomen for the last month. No history suggestive of obstructive jaundice, altered bowel and bladder habit was present. He was a chronic smoker for the last 30 years and there was no history suggestive of diabetes mellitus, tuberculosis, any chronic illness or any previous surgery. Family history was non-contributory regarding occurrence of malignancies in the family. On general examination, he was afebrile with mild pallor and no jaundice. On examination of the cardiovascular system, apex beat was present more clearly in the right fifth intercostals space lateral to the right border of the sternum. Per abdominal examination revealed mild tenderness and a hard globular lump size of 3 × 4 cm coming out of the left subcostal margin in the midclavicular line, which was moving freely with respiration. Clinically there were no signs of free fluid, bowel sounds were normal and per rectal examination revealed no growth and any abnormality. Chest X-ray posteroanterior view showed apex and gastric fundal gas shadow towards the right side and the electrocardiography showed transposition of first and second lead and inversion of the first lead. These findings were confirmatory signs for situs inversus in our patient. Ultrasonography of...
the upper abdomen showed situs inversus abdominis with isoechoic, well defined mass lesion of a size of $3.6 \times 2.0$ cm with irregular margins in gall bladder lumen along with echogenic calcification. Contrastenhanced computed tomography image of the upper abdomen showed transposition of all major organs with liver, gallbladder and head of the pancreas on the left side and spleen, stomach and tail of pancreas on the right side. Gall bladder revealed thickened walls (3–10 mm), more marked in the fundal region with moderate enhancement and pericholecystic fat planes appeared normal (Figure 1). There was no evidence of infiltration of adjacent liver parenchymal by this mass as well as lymph node involvement in hepatoduodenal ligament. Laboratory investigations, including complete blood cell count, liver function tests, kidney function tests and biochemical tests, were within normal limits. A provisional diagnosis with the highest probability of the carcinoma gall bladder and SIT was established. As, clinically, tumour was confined to the gall bladder, but to decide the operability and confirm the absence of metastatic disease, first staging laparoscopy and intraoperative frozen section biopsy from the gall bladder mass was done. On confirmation of diagnosis of adenocarcinoma gall bladder, open radical cholecystectomy with 3 cm non-anatomical wedge resection of the gall bladder bed of liver and Roux-en-Y hepaticojejunostomy was done, via left-sided Kocher’s incision. A western style portal nodal dissection which included a complete portal lymph node dissection including suprapyloric lymph nodes and skeletonisation of the portal structures along with celiac lymph nodes dissection was done. Definitive histopathological examination of the resected gall bladder showed moderately differentiated adenocarcinoma, with neoplastic cells forming papillary and acinar structure without lymphovascular or perineural invasion. Hepatic and common bile duct cut margin were not involved by the tumour. Paracholedochal and celiac lymph nodes did not show metastatic involvement. Now the patient is on his regular follow-up and doing well. Postoperative imaging studies performed six months after surgery showed no evidence of recurrence at local sites or distant metastasis.

**Discussion**

SIT is a rare congenital condition in which the precise aetiology is unclear; however, it is believed to be due to an autosomal recessive gene located in chromosome 14 and deletions affecting chromosomes 7 or 8. In BTCs are invasive adenocarcinomas that account for 4% and 3% of all gastrointestinal cancers, respectively. BTCs are invasive adenocarcinomas that arise from the epithelial lining of the gallbladder and intrahepatic and extrahepatic bile ducts. GBC represents the most common and aggressive cancer among all the BTCs and carries an extremely poor prognosis. The incidence of GBC steadily increases with age; women are affected two or three times more often than men, and more common in

**Figure 1**: Contrast-enhanced computed tomography image of the abdomen, showing transposition of all major organs with liver, gall bladder and head of the pancreas on the left side and spleen, stomach and tail of the pancreas on the right side. Gall bladder showing thickened walls in the fundal region (arrow) with moderate enhancement.

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