Case Report

Pyloric perforation in situs inversus totalis and Ladd’s band with dextrocardia: a collection of rare anomalies and associated operative challenge

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INTRODUCTION

Situs inversus is a rare congenital anomaly where the intra-abdominal viscus is reversed or mirrored from their normal position.1 When associated with dextrocardia it is termed as situs inversus totalis. Situs inversus is a rare autosomal recessive condition associated with complete transposition of the abdominal and thoracic organs. In cases of 'situs inversus totalis’ or ‘mirror image dextrocardia’ the heart is on the right side of the midline while the liver and gall bladder are on left side along with a 'mirror image’ of normal bowel, caused by a clockwise rotation of the viscera in early embryonic life.2 Surgical diagnosis and procedures in patients are tricky because of mirror image anatomy of intra-abdominal organs.

CASE REPORT

A 22-year-old male, presented to our emergency department with severe abdominal pain. He presented with history of acute onset abdominal pain in the epigastrium 7 days back which had increased in severity for 1 day, pain aggravated on movement and reduced on leaning forwards. He also complained of nausea and vomiting. A physical examination revealed a pulse rate of 94 beats/minute, blood pressure of 100/70 mmHg, and he was afebrile. Examination of his abdomen revealed guarding and rigidity. The laboratory results showed a serum haemoglobin (Hb) of 17 g% and a white blood cell (WBC) of 19,600 cmm with neutrophilia but other biochemical test results were essentially normal. Results of an X-ray of the chest showed dextrocardia, X-ray erect posture showed fundic gas shadow on the right side with free gas under both dome of diaphragm, left>right. A clinical diagnosis of hollow viscus perforation was made.

Subsequently an ultrasound sonography test (USG) scan was performed and suspicion of situs inversus was confirmed with liver with gall bladder was noticed in the

ABSTRACT

Situs inversus totalis with dextrocardia is a rare entity where the liver and gall bladde are on the left side and the spleen on the right. It is an autosomal recessive mode of inheritance and the patients are usually asymptomatic with a normal life span. Ladd’s band on the other hand is a fibrous band of peritoneum lying over the duodenum to the caecum, which usually presents as intestinal malrotation and is most commonly diagnosed and treated with Ladd’s procedure in infancy itself. Asymptomatic adult cases presenting later in life are rarely seen. Here we present a case of a 22-year-old male who presented with symptoms of perforation peritonitis and was diagnosed on clinical and radiological examination to have situs inversus. Intra-operative findings later revealed presence of a Ladd’s band too.

Keywords: Pyloric perforation, Situs inversus surgery, Ladd’s bands, Dextrocardia
left side and spleen on the right side without any pathology and with normal echo texture.

After resuscitation with intravenous fluids, and administration of antibiotics, proton pump inhibitors, and nasogastric aspiration, patient was posted for an exploratory laparotomy. On exploration there was an acute perforation of about 5 mm diameter in pylorus part of the stomach. There was complete situs inversus ‘mirror image’, with the liver and gall bladder on the left side and spleen on the right side. A Ladd’s band was noticed extending from the greater curvature of the stomach and wrapped around the duodenum, to the transverse colon and on further exploration inter bowel adhesion were present and caecum with the appendix were pulled upwards. The large intestine up to the proximal 1/3 of transverse colon was distended and twisted along the longitudinal axis of the mesentery.

Primary closure of the pyloric perforation with overlay of omental (Graham’s) patch was performed with absorbable sutures, an edge biopsy was sent of the perforation. Ladd’s band was divided and inter bowel adhesions were released caecopexy was done and a thorough peritoneal lavage was performed; an incidental appendectomy was also performed to avoid further diagnostic problems and the abdomen was closed in layers.

Post-operative recovery—the patient recovered well with oral feeds being started on day 3 and the patient being discharged on post-operative day 8. Biopsy returned negative for malignancy and was reported as an acute inflammatory pathology.

DISCUSSION

Situs inversus abdominalis is an uncommon anomaly with an incidence varying from one in 4,000 to one in 20,000 live births. Situs inversus usually remains undiagnosed, as exemplified by the present case, unless it is diagnosed incidentally while investigating another associated ailment. A diagnostic dilemma arises whenever pathology occurs in the unusual located abdominal viscera situs inversus with dextrocardia is also known as situs inversus totalis. There are also reported cases of situs ambiguous or heterotaxy, where liver may be in midline, absent spleen or multiple spleen and malrotated bowel leading to diagnostic confusion. Various modalities such as electrocardiograms, radiographic studies, computed tomography (CT) scans with oral and intravenous contrast, ultrasound, and barium studies can be used to diagnose situs invertus. Adult gut malrotation is very rare and its incidence has been reported to be between 0.0001 and 0.19%. To choose a proper surgical incision for abdominal exploration, pre-operative recognition of the condition is important. In our case the diagnosis was made pre-operatively and an exploratory laparotomy was performed with an upper midline incision.

In this case, we diagnosed the condition by a chest radiograph and abdominal ultrasonography.

There have been isolated reports of situs inversus associated with peptic ulcer, ulcer perforation, amoebic liver abscess, acute cholecystitis, cholelithiasis, acute appendicitis, and intestinal obstruction. To the best of our knowledge, this is first case of patient with situs inversus totalis presenting with pyloric perforation with Ladd’s band.

CONCLUSION

Combinations of rare anomalies present a surgical challenge in the presence of acute or chronic pathologies and strict adherence to the individual patient anatomy gives the best postoperative result.

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