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Case Report

Incomplete Situs Inversus Presenting as a Case of Neonatal Intestinal Obstruction

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ABSTRACT

Situs inversus totalis is a rare congenital abnormality characterized by a mirror-image transposition of both the abdominal and the thoracic organs. Laterality is achieved established early in development, and any failure in that process might lead to a wide variety of positional derangements which may be partial or complete. Situs solitus describes the normal anatomy, situs inversus is the complete reversal, and situs ambiguous is used for any other abnormality of left-right development and shift. Complete situs inversus is a very rare anomaly is characterized with the total inversion of all abdominal and thoracic ORGANS. During the normal embryonic development, laterality (left-right-sidedness) is controlled by a group of signal molecules and genes. Any disturbance in the establishment of normal anatomical left-right asymmetry during this period results in left-right axis disorder which may express as complete situs inversus, incomplete situs inversus or situs ambiguous. The amin of this report was to present a case of situs inversus ambiguous which presented as neonatal intestinal obstruction. The index case would be reported here as a presented from admission to mortality.

Keywords: Incomplete, Situs Inversus, Presenting, A Case, Neonatal, Intestinal, Obstruction.

INTRODUCTION

Situs inversus is a rare condition (1 in 8,500 people) causing mirror image positioning of thoracic and abdominal organs. Situs inversus abdominus also known as situs inversus with levocardia or left-located heart is a condition with right-to-left reversal limited to the abdomen.[1] In situs inversus totalis, other structural malformations are uncommon in most individuals, but they are slightly more frequent than in people with situs solitus.[2] Many people with situs inversus are unaware of their anomalies until when evaluated medically. Situs inversus totalis complicating neonatal intestinal obstruction is very rare and presents with dilemmas in management. The index case is a case of situs inversus solitus associated with jejunal atresia which manifested as jejunal obstruction.

It is a broad term that includes variety of congenital anomalies which could involve almost every organ in the body. The association of this condition with duodenal atresia is very rare, but has been reported in about 20 patients.[3,4,5] More recently, the association of

heterotaxy with jejunal atresia was reported once by Rasool et al.[6] Abdur-Rahman et al.[7] in separate studies.
A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia. It is usually associated with transposition of the great vessels and a right-sided aortic arch, atrial and ventricular septal defects, tetralogy of Fallot.[8] There are evidences in the literature that showed the association of situs inversus with other congenital anomalies such as Kartagener syndrome, duodenal atresia, biliary atresia and gastroschisis.[9],[10,11]

Situs inversus is mostly diagnosed by chance during investigative modalities or exploratory laparotomies for other abdominal conditions. This index case is being reported because of the unusual presentation of this case as jejunal atresia which was associated with situs inversus ambiguous.[12]

CASE PRESENTATION

A male neonate who was seen 2 hours after birth on account of bilious vomiting. He was a product of unplanned pregnancy. His mother had antenatal care at the obstetric department of the hospital. There was no history of use of unprescribed drug during pregnancy. Prenatal Ultrasound scan done at gestation age of 6 months showed maternal polyhydraminous and dilated foetal bowel loops. The mother however developed premature rupture of membrane at 33 weeks. Steroid was administered to aid lung maturation and she subsequently had emergency caesarian delivery due to suspected Chorioamnionitis at gestational age of 34 weeks. The baby weighed 2.5kg at birth and APGAR scores were 8 and 9 at 1st minute and 5th minute respectively.

However, about an hour after birth, he developed repeated of bilious vomiting and upper abdominal distension was also observed. He was thus transferred to paediatric surgical service for expert management.
He is the first child of unmarried couple. The mother is 23 years old $G_5P_0^{+4}$ and she has had four previous voluntary termination of pregnancy.

Examination on admission revealed an active, pink, anicteric, acyanosed, well hydrated neonate. His respiratory rate was 50/min with broncho-vesicular breath sounds. His heart rate was 140/min, heart sound 1 and 11 were heard and there was no added sound. His apex beat was located at the 5th left intercostals space, mid clavicular line. The upper abdominal region was full but soft. Bowel sounds were present and hyper-active.
Rectal examination revealed patent but empty rectum. Initial clinical diagnosis was Jejuno-ileal Atresia.

The patient immediately had a wide bore nasogastric tube inserted and intravenous fluid and antibiotics were commenced. A plain abdominal radiograph and upper gastro-intestinal tract series

Figure 1 showing asymmetric swelling of the abdomen with a localized swelling at the right hypochondrium

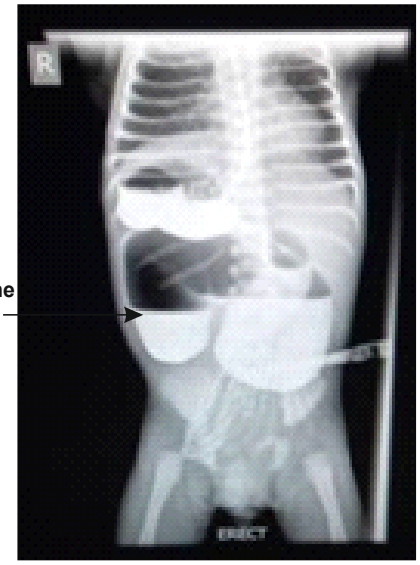

Figure 2 showing gastric shadow on the right side of the midline

(figures 1 and 2) were obtained and both suggested jejunal atresia with abdominal situs inversus and levocardia. The patient was thus worked up for laparotomy. His haematocrit was 42% and white cell count was 4.56 x 10^9/L. Serum urea and electrolytes were within normal limits.

He subsequently had laparotomy done on the 5th days of life under general anaesthesia. Intra- operative findings included Type 1 proximal jejunal Atresia, liver/gall bladder located on the left hypochondrium, spleen on the right and multiple congenital bands. He had division of congenital bands done as well as excision of atretic portion of jejunum with end-to-end jejuno-jejunal anastomosis.
The post-operative period was uneventful until 6th day post-operation when he developed tachycardia (Heart R

= 168/min) and fever. There was also

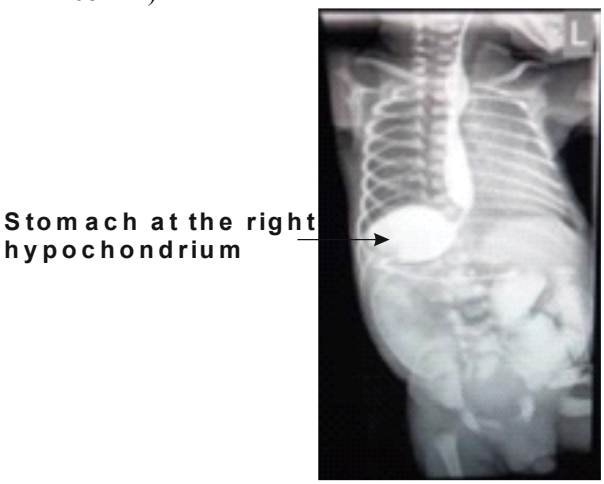

Figure 3: Upper gastrointestinal contrast studies findings of the patient

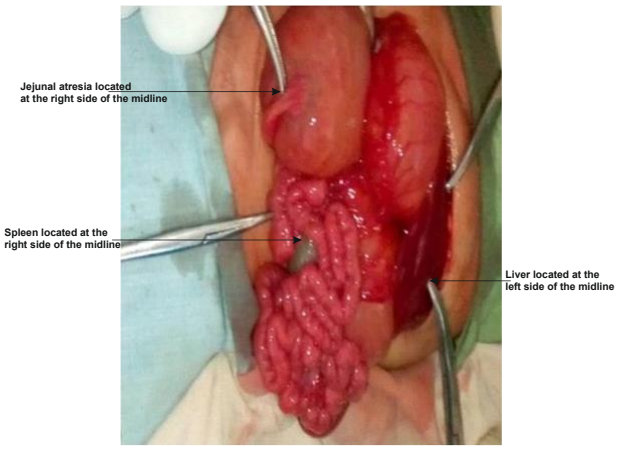

Figure 4 showing intra-operative findings of the patient

progressive abdominal distension and significant reduction of daily nasogastric effluent. He was thus considered to have developed an anastomosis leak.

Patient was taken in for repeat laparotomy. The major finding intra-operatively was partial dehiscence of previous anastomosis. The wound edges of dehisced anastomosis site were freshened and primary single layer repeat anastomosis was done. The peritoneal cavity was lavaged with warm saline. He also had sessions of blood transfusion to correct anaemia post operatively.

He first passed greenish stool on 3rd post-operative day. However, he was noticed to drain greenish effluent from the abdominal incision wound 5th day after re-exploration. He also developed persistent fever, tachycardia and progressive altered consciousness. His antibiotic regimen was changed to ampicillin with clavulanic acid based on culture sensitivity pattern. He, however continued to deteriorate and subsequently stopped breathing on 8th day after re-exploration. A post mortem examination was conducted, findings included the following: the heart normally positioned in the left hemithorax, liver and gall bladder in the left hypochondrium and spleen in the right hypochondrium. **(**Figure 3**)**

DISCUSSION

Situs inversus is a congenial anomaly with inverted position or transposition of internal organs from their normal anatomical position; and also referred to as situs inversus viscerum or situs transversus, or opposites.[13] The normal anatomical position of internal organs is termed as situs solitus. Situs inversus was first described in humans by Fabricius,[14] while Vehemeyer[15] gave the radiological diagnosis.

The index case was a case of situs inversus ambiguous which presented as neonatal intestinal obstruction. Pre-operative features were apex beat located at the fifth left intercostal space mid- clavicular line, bilious vomiting, abdominal distension and constipation. Pre-operative assessment included upper gastro-intestinal tract series and plain abdominal radiographs (erect and supine) which suggested the presence of intestinal atresia. At exploratory laparotomy, the spleen was in the right hypochondrium while the liver and the gall bladder were in the left hypochondrium. The presence of type 1 jejunal atresia was confirmed intra-operatively. These findings are similar to the findings of similar authors.[16]

In situs inversus totalis, there is complete right to left reversal of all of the viscera including dextrocardia; the morphologic right atrium is on the left and the left atrium is on the right. The normal pulmonary anatomy is reversed such that the left lung has three lobes and the right lung has two. The liver and gallbladder are located on the left, and spleen and stomach are on the right side.[17,18] Other congenital anomalies associated with situs inversus anomalies are splenic malformations, jejunal atresia and dextrocadia. Situs inversus is usually asymptomatic and if not picked during physical examination of the precordium; may also be incidentally diagnosed during investigations for other conditions or laparotomies for other abdominal surgical conditions.[19,20]

Associated of situs inversus with intestinal atresia reveals its presence early especially during neonatal period. Heterotaxy syndrome or situs ambiguous found in about 4 in one million live births is defined as the abnormal positioning of organs and vessels as opposed to the proper arrangement in situs solitus. There are 3 sub-types of heterotaxy based on the presence and location of the spleen; they are described as follows: (i) asplenia, (ii) polysplenia and (iii) normal spleen located in the right upper quadrant of the abdomen and more commonly

associated with congenital heart disease in 50-90% of cases, intestinal atresia and malrotation in 70-100% of cases. The etiologic agents implicated in situs inversus are autosomal recessive gene, cocaine abuse, retinoic acid exposure and maternal diabetes. Though, there was no sex predisposition in the prevalence of situs inversus as revealed in the literature searches;[21,22] some authors[23,24] have reported the occurrence of situs ambiguous in males more than females as witnessed in our report.

Situs inversus presenting in the neonatal period with gastrointestinal complaints is usually an abdominal emergency just like the index case who presented as a case of intestinal obstruction and was investigated through upper gastrointestinal series, plain abdominal x-ray and exploratory laparatomy. He had excision of jejunal atresia and maintenance of intestinal continuity through end-to-end jejuno-jejunal anastomosis. However, the anastomosis failed after two repeated attempts and he finally succumbed to septicaemia. Postmortem confirmed situs ambiguous with the spleen in the right hypochondrium.

CONCLUSION

The index case presented with neonatal intestinal obstruction secondary to jejunal atresia which was associated with situs inversus ambiguous.

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