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A Rare Case Presentation of Meckel’s Diverticulum with Situs Inversus Totalis

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ABSTRACT

Situs inversus, a rare congenital condition, is a complete mirror image of the thoracic and abdominal viscera, in which the positions of major visceral organs are reversed. Here, we report a case of a 6 year old boy who presented with abdominal pain in both the iliac fossae, fever and vomiting. Imaging studies showed situs inversus totalis and a provisional diagnosis of acute appendicitis was made and a possibility of meckel’s diverticulitis was suspected. A final diagnosis of meckel’s diverticulitis was made during emergency laparotomy. This case report is being presented because of its rare occurrence.

Keywords: Situs inversus totalis, Meckel’s diverticulitis, Congenital positional anomaly, Heterotaxia

Introduction

Situs inversus is a rare congenital positional anomaly with an incidence of 0.01% in the USA [1]. Meckel’s diverticulum is found in 2% of the population with normal visceral configuration [2]. Situs inversus with meckel’s diverticulitis is a rarer entity with only 2 cases reported earlier [3,4], to the best of our knowledge. Documenting situs inversus in an individual is important in order to correctly interpret any future symptoms and avoid any inadvertent clinical or surgical mishaps. This case report is being presented because of its rare occurrence.

Case report

A 6 year old boy presented with abdominal pain of 4 days duration. The patient also had fever and vomiting for 3 days. On clinical examination, tenderness was elicited over both the right and left iliac fossae with maximum tenderness over the left iliac fossa. Rebound tenderness was present. Bowel sounds were sluggish. On radiological examination, heart was found to be in the right side. It was confirmed by echocardiography. Roentgenographic examination of the chest and abdomen and CT scan of the abdomen confirmed situs inversus totalis (Figure 1).

A provisional diagnosis of situs inversus totalis with acute appendicitis was made. Due to tenderness over both the right and left iliac fossae, a possibility of meckel’s diverticulitis was suspected. Since the patient presented with peritonitis, as evidenced by rebound tenderness and sluggish bowel sounds, the patient was scheduled for emergency laparotomy. Usually in children, laparotomy is done by supra/infracumbilical transverse incision. Since the patient presented with situs...
inversus totalis, surgery was proceeded with midline longitudinal incision.

Under general anaesthesia, the abdomen was opened in layers. Meckel’s diverticulum was found to be inflamed (Figure 2).

Purulent secretion was found in the peritoneal cavity adjacent to meckel’s diverticulum. Sigmoid colon was found in the right iliac fossa. Caecum and appendix was found in the left iliac fossa. Appendix was found to be normal. Liver and gall bladder was found in the left hypochondrium. Resection of the ileal segment containing the meckel’s diverticulum and end to end anastomosis along with prophylactic appendicectomy was done. On histopathological examination of the diverticulum, mucosa showed no evidence of heterotopic epithelium (Figure 3).

**Discussion**

Mathew Baillie first described situs inversus totalis in the early 20th century [1]. Situs inversus is a rare congenital condition which is inherited in an autosomal recessive pattern [1,5], although it can be X linked [6]. Situs inversus is a complete mirror image location of the thoracic and abdominal viscer, in which the positions of major visceral organs are reversed. The normal arrangement of visceroauthal situs is situs solitus [8]. The molecular basis for the structural asymmetry of the body has been linked to a complex interaction of fibroblast growth factor 8, Nodal gene, Lefty 2 gene, PITX2 gene, Lefty 1 gene, Sonic HedgeHog (SHH) gene, Brachyury (T) gene and expression of Snail transcription factor [7]. Yet, it remains unclear.

The oldest and the most persistent theory of visceral inversion is that twins are often mirror image of each other that a single individual with situs inversus represents a survivor whose normal twin has died in utero. Woellwarth’s experiments suggested that some cases may arise from early embryonic accidents such as injury to the governing half while other cases appear to have a genetic basis. If the left side is injured the right side develops the characteristics of a mirror image of the left side while left becomes mirror image of the right. Heart loops to the left side instead of the right side to produce dextrocardia [7]. Situs inversus can be associated with right sided heart (dextrocardia) which is situs inversus totalis or can be associated with left sided heart (levocardia) which is situs inversus incompletes [1,8]. In situs inversus totalis, heart is located on the right side of the thorax and its apex pointing to the right side. Stomach and spleen are present on the right side. Liver and gall bladder are present on the left side of the abdomen. Normal pulmonary anatomy is also reversed i.e., left lung is trilobed and right lung is bilobed. Blood vessels, nerves, lymphatics, intestines are also transposed. There is a 5-10% prevalence of congenital heart disease in situs inversus totalis, most commonly transposition of great vessels. Incidence of congenital heart defects is more (95%) with situs inversus incompletes [1]. 25% of individuals with situs inversus have primary ciliary dyskinesia. They present as Kartagener syndrome [1] with the classic triad of situs inversus, chronic sinusitis and bronchiectasis. When the visceroauthal situs cannot be determined, the condition is called situs ambiguous or heterotaxia [8]. It may be either right isomerism (asplenia syndrome) or left isomerism (polysplenia syndrome). Heterotaxia is usually associated with severe congenital heart defects [1] (ASD, VSD).

Meckel’s diverticulum is a remnant of omphalomesenteric or vitelline duct [7]. It is situated on the antimesenteric boder of the small intestine [2]; 60cm from the ileo-caecal
valve and is 3-5 cm long. It has all the four coats of the intestinal wall and has its own blood supply [2]. Therefore, they are vulnerable for infection and obstruction in the same way as appendix. When a normal appendix is found at surgery for suspected appendicitis, meckel’s diverticulum should be sought by inspection of an appropriate length of terminal ileum.

In 20% of cases, mucosa contains heterotopic epithelium. It is usually difficult to demonstrate by contrast radiography. In the presence of ectopic heterotopic epithelium, the most sensitive study is Meckel Radionucleotide Scan [8]. The most accurate investigation is small bowel enema [2]. Situs inversus totalis usually does not cause any significant morbidity to an individual harbouring it. However, its timely diagnosis is crucial for the correct interpretation of future symptoms and results of diagnostic procedures. An antedated diagnosis of this disorder will form a baseline reference for future surgical procedures and as such will be invaluable in preventing an unintentional operative mishap.

References


