



Situs İnversus Totalis İle Komplike Olmuş Çocuk Cerrahisi Olguları

Pediatric Surgical Cases Complicated With Situs Inversus Totalis

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Öz

Situs inversus totalis (SIT), organların ayna görüntüsü olacak şekilde vücudun karşı tarafına yerleşmesiyle karakterize olan sıradışı bir anomalidir. Klinik pratikte pediatrik cerrahlar akut apandisit, intususepsiyon, duodenal atrezi, özofagus atrezisi ve bazen hidrokolpos ile sıklıkla karşılaşmaktadır. Sonuç olarak; anatomik ve mekansal durum bilindiği sürece cerrah, SIT ile komplike olan cerrahi durumlarda güncel tedavi prosedürlerini kolayca uygulanabilir. Bazı sendromlarla ilişkili olabileceği bilinmelidir.

ABSTRACT

Situs inversus totalis (SIT) is an unusual anomaly that characterized by complete mirroring of organ locations in the opposite side of the body. Acute appendicitis, intussusception, duodenal atresia, esophageal atresia, and occasionally hydrocolpos are frequently encountered in pediatric surgeons in clinical practice. As a result; the surgeon can easily apply current treatment procedures in surgical conditions complicated with SIT as long as the surgeon is aware of the anatomical and spatial state. Complicated diseases with SIT should be kept in mind as they may be associated with some syndromes

Anahtar Kelimeler: situs inversus totalis, çocuk, cerrahi

Keywords: situs inversus totalis, pediatric, surgery

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Başvuru Tarihi/Received: 09-06-2018

Kabul Tarihi/Accepted: 24-04-2019



Introduction:

Situs inversus totalis (SIT) is an unusual anomaly that characterized by complete mirroring of organ locations in the opposite side of the body. Acute appendicitis, intussusception, duodenal atresia, esophageal atresia, and occasionally hydrocolpos are frequently encountered in pediatric surgeons in clinical practice. In this study, 6 pediatric SIT cases that need surgery, follow up and treatment were evaluated with the review of the literature.

Material-Method: Between 2013 and 2018, a retrospective screening of the 830 patients who underwent follow-up and treatment at Pediatric Surgery Clinics was performed. 6 children who were operated due to SIT were identified and presented with literature review.

Case 1: A 14-year-old girl presented with a right lower quadrant pain starting 2 days ago. in examination tenderness was felt on her both iliac fossa more prominently on the right. Wbc: 14000 / mm³. the patient was learned to be SIT and in the abdominal tomography her appendix was found in left lower quadrant (figure 1). Laparoscopic appendectomy was performed. The patient was discharged without any problem.



Figure 1: CT shows appendix on the left side(White arrow)

Case 2: A 13-year-old girl was admitted to our clinic with a right lower quadrant pain in the abdomen, in the abdominal CT polysplenism without a main spleen , a tail deformity in the pancreas and left lower quadrant placed appendix was detected. Laparoscopic appendectomy was performed.

Case3: A 2-day, 1800-gram male patient who was evaluated for vomiting after a feeding period and on babygram had a double bubble view on the right (figure 2). SIT and duodenal atresia were put as diagnose. During operation, it was observed that the spleen was placed on the right side (figure3). Duodenoduodenostomy was performed without any complication.



Figure2: On Xray, double Bouble is on the right



Figure3: Spleen placed on right side of stomach

Case 4: A 4-year-old female patient who presented with intermittent abdominal pain and vomiting was evaluated, in USG target sign was detected at the umbilical level. No features were found on the X-ray. The patient, known to be a SIT from her history, her intussusception was treated non-surgically with ultrasound guided hydrostatic reduction.

Case 5: A 4-day-old girl was admitted to the neonatal intensive care unit due to abdominal tenderness, and distension. Hymen was not detected on the external genital examination and the vagina was terminated blindly. The presacral cystic appearance in MR (figure 4) was evaluated as hydrocolpos. Syndromic facial appearance was evaluated as Bardet-Biedl syndrome due to polydactyly, renal, cardiac anomalies and SIT. Transabdominal percutaneous drainage of the cyst was done and vaginal dilatation is continued with Hegar dilators.

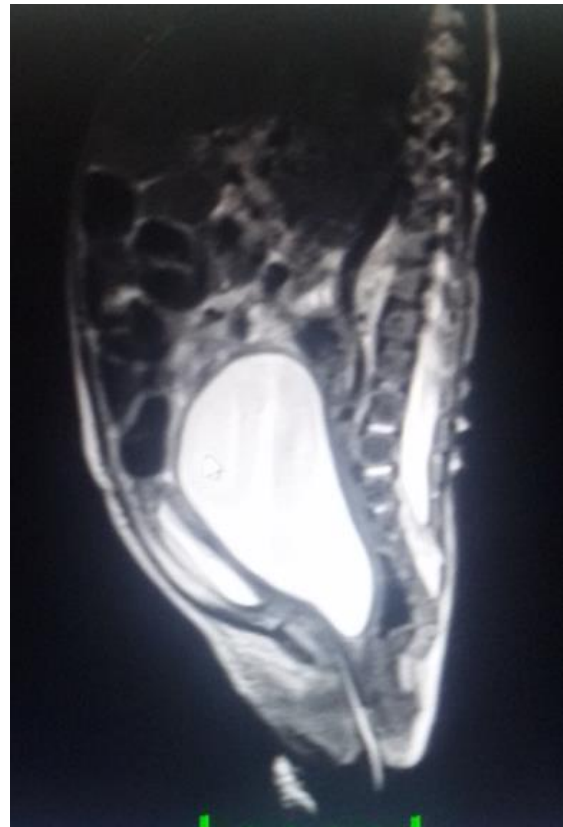


Figure 4: MR Shows Uterine Cystic Lesion

Case 6: A 2-day-old male with vomiting and nutritional intolerance was found to have esophageal atresia. In her esophagography due to lack of gas in abdomen and esophageal pouch image, it diagnosed as isolated esophageal atresia. The SIT was considered due to the right side placed heart in the echography and the left side localized liver in the ultrasound. The patient had right esophagostomy due to a long gap and gastrostomy was performed on the right quadrant because stomach was paced on right side (figure 6). According to our information; our case is the first defined SIT case in English literature which has pure esophageal atresia in the same time.



Figure 6: Right esophagostomy and right gastrostomy catheters show right-sided stomach

Discussion:

Situs inversus totalis (SIT) occurs with genetic predisposition as a mutation in the long arm of the autosomal recessive chromosome 14 (2). The incidence is 1 / 10,000th in the general population (3). SIT cases are generally asymptomatic healthy individuals and they are detected by examination in the radiographs (4). SIT occurs by 270 degrees clockwise rotation of the embryonic midgut (5).

SIT; renal dysplasia, pancreatic fibrosis, intrahepatic biliary dysgenesis, Cartagener syndrome, cardiovascular (dextrocardia ASD, VSD, Fallot, pulmonary stenosis and large arterial transposition), respiratory (absence of one lung, bronchiectasis, absence or deformity of the paranasal sinuses). Simultaneous association with the digestive system (atresia or stenosis of the duodenum, persistent of Meckel's diverticulum, absence of appendix, megacolon, atresia of anus, splenic and genitourinary anomalies have been reported (6). The diagnosis is put usually with imagings such as x-ray, ultrasound and CT images. CT is the most appropriate imaging method for determining the location of intestinal organs (1,7).

The incidence of acute appendicitis in SIT has been reported between 0.016 % and

0.024 % (1). Situs inversus totalis does not increase the incidence of acute appendicitis. Akbulut reported that 62% of the SIT patients had pain in the left lower quadrant and 14% in the right lower quadrant and %7 bilaterally (7). Our patients had pain on both sides more clearly in the right lower quadrant. Appendices were detected with CT on the patients which are known to be SIT. Laparoscopic appendectomy was performed. Laparoscopy is gold standard in the treatment of acute appendicitis cases (1).

Congenital duodenal obstruction is a congenital anomaly seen in 10000 to 15000 live births. Approximately 20 SITs has been reported with a duodenal atresia in the literature (8). The treatment of congenital duodenal atresia with or without SIT is duodeno-duodenostomy.

Intussusception is the most common cause of acute abdomen after acute appendicitis in children (9). Nearly 100% are diagnosed with ultrasound (9,10). Ultrasound guided hydrostatic reduction has 85% of success rate in the treatment (10). The association of SIT and intussusception is rarely reported in adults and children (4,11). As long as there is no intestinal perforation and necrosis in intussusception cases, nonsurgical treatment is the first choice (10). Our case which is known to be SIT, was treated with USGHR upon detection of invagination with USG. No recurrence was detected so surgical approach was not considered.

Hydrocolpos or pyocolpos is a very rare congenital disorder, occurring in 1 / 30,000 live birth (12). The association of Bardet-Biedl syndrome with hydrocolpos and SIT in coincidental association is known (13). In our case, cyst aspiration have done with transabdominal peritoneal drainage and dilatations have been continuing with Hegar dilators.

Esophagus anomalies with SIT are very rare. SIT with EA / TEF patients are frequently



clinically diagnosed due to dextrocardia. If preoperative evaluation have not done, it can make the esophagus anastomosis very difficult (14). In our case, gastrostomy was opened due to pure esophageal atresia and esophagostomy was performed due to long gap detection and colonic interposition surgery was planned.

As a result; the surgeon can readily apply existing treatment procedures in surgical conditions complicated with SIT as long as the surgeon is aware of the anatomical and spatial state. Complicated diseases with SIT should be kept in mind as they may be associated with some other syndromes. Also in patients with SIT, putting a marker on their bodies for possible future medical interventions will facilitate the work of health workers.

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