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Embryogenesis and types of subcostal hernia—A rare entity☆,☆☆,★

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Abstract

Background/Purpose: Four infants with congenital subcostal hernia are reported, as it is a rare entity with only two cases previously reported. Further, there are no reports concerning the complex multisystem subtype. Embryogenesis of the associated anomalies and subcostal hernia and their management are discussed.

Materials/Methods: Clinical features, history, investigations, associated anomalies, and management data of four patients with subcostal hernia were collected and analyzed.

Results: The following associated anomalies were detected: renal agenesis (2), musculoskeletal abnormality (3), congenital heart disease (2), müllerian–renal–cervicothoracic somite abnormalities and vertebral–anorectal–cardiac–tracheoesophageal–renal–radial-limb anomalies (1). The subcostal hernias were treated by laparoscopic assisted (3) or laparoscopic herniorrhaphy (1).

Conclusions: Subcostal hernia is a rare entity with varied clinical presentations and presents either as an isolated defect or as a complex multisystem defect. The exact etiology is still unknown. Phenotypic manifestation of the complex defect is probably due to developmental gene defect affecting the coordinated growth of mesoderm around 4th to 10th weeks of fetal life.

Abbreviations: ARM, anorectal malformation; RA, renal agenesis; CETV, congenital talipus equinovarus; CHD, congenital heart disease; ASD, atrial septal defect; VSD, ventricular septal defect; PDA, patent ductus arteriousum; ECHO, echocardiography; CECT, contrast enhanced computerized tomography; MRI, magnetic resonance imaging; MRKH, Mayer, Rokitansky, Kuster, Hauser syndrome; MURCS association, müllerian, renal, cervicothoracic somite abnormalities; VACTERL associations, vertebral, anorectal, cardiac, tracheoesophageal, renal, radial limb; BMP, Bone morphogenetic proteins; FGFs, Fibroblast growth factors; WNT gene, hybrid of Integration1 gene and wingless gene.

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Astley Cooper (1804) defined hernia as a protrusion of any viscus from its proper cavity [1]. Inguinal hernia is the commonest type of neonatal hernia [2] and its reported incidence varies from 1% to 5%. Subcostal hernias are usually post-traumatic entities and rarely congenital. In our cohort, four neonates presented with a protuberant mass in the left subcostal region, with a small abdominal wall defect and herniation of its contents. To the best of our knowledge, this is the largest series of the subcostal hernias reported. In addition to reviewing the literature of congenital subcostal hernia, we also discuss the embryology, associated anomalies, and provide a new classification.

1. Materials and methods

Four patients with congenital subcostal hernia were admitted to our hospital between January 2009 and November 2011. The facility is a teaching medical college hospital and tertiary care center. Three cases were managed by the first author and the corresponding author while the fourth case was managed by the third and corresponding authors. The fourth and fifth authors were involved in editing the manuscript and providing inputs for the embryology. The clinical features, associated anomalies, echocardiography (ECHO), radiological findings and surgical data were collected and analyzed. All but one, were term neonates and all had an uneventful pregnancy. The family history was noncontributory. All four infants were from a close geographical area within a radius of 120 km and presented as a cluster of four cases within a short span of 1 year. The exact etiology of occurrence of four cases in the same geographic area could not be determined. Detection of the four cases may be merely co-incidental or may be influenced by some environmental or genetic factors.

2. Clinical findings and case reports

All the neonates had a left subcostal hernia with a muscular defect varying from 3 to 10 cm. Antenatal sonograms (third trimester) did not pick up the hernia but left renal agenesis (RA) was detected in one fetus.

2.1. Case 1

A 2-day-old girl presented with abdominal distention, vomiting, absent anus and abnormal genitalia. Physical examination showed moderate abdominal distention, left subcostal hernia, imperforate anus, recto-vestibular fistula with vaginal agenesis and congenital talipus equinovarus (CTEV). Other systemic examination showed atrial septal defect (ASD) with tricuspid regurgitation. Initially she was investigated with a plain infant radiogram, abdominal ultrasound and ECHO. She underwent staged repair of the anorectal malformation (ARM). Diagnostic laparoscopy was carried out for completion of evaluation. The investigations revealed the occurrence of multisystem associated anomalies including agenesis of the müllerian ducts and left ovary, and congenital heart disease (ASD with tricuspid regurgitation). She also had butterfly 11th vertebra, in addition to partial sacral agenesis and left RA. The patient was a normal female with 46XX chromosomal pattern on karyotype. We performed laparoscopic repair of the subcostal abdominal wall defect. Postoperatively she had a mild residual diffuse bulge, which was attributed to early hernia recurrence. Presently she is 1.5 years old and vaginal reconstruction has been planned when she attains the age of onset of sexual intercourse.

2.2. Case 2

A 1-day-old boy presented with respiratory distress, deformed chest, and left reducible subcostal hernia covered by hypoplastic, reddish-appearing skin (Fig. 1). Antenatal and postnatal abdominal ultrasound studies confirmed left RA. Plain radiographs of the chest and abdomen showed crowding of the left upper ribs, agenesis of 7th and 8th ribs, approximations of 11th and 12th ribs (thoracic wall defect) and a subcostal abdominal wall defect with herniation of bowel loops. The ECHO appeared to be normal. Diagnostic laparoscopy revealed a hernial orifice with a smooth margin. This was repaired by an open approach in view of the dysplastic skin which also required excision and repair (Fig. 2). However, postoperatively he succumbed to neonatal sepsis secondary to pneumonia.

2.3. Case 3

A 6-month-old boy, who was born preterm, presented with an isolated subcostal hernia and underwent a successful

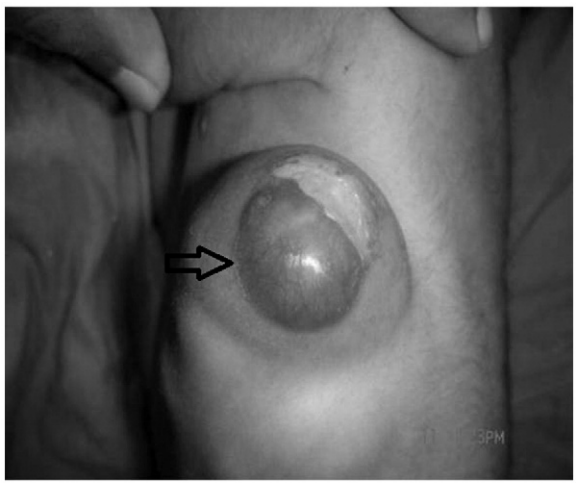

Fig. 1 Left subcostal hernia with dermal hypoplasia (arrow).