Congenital subcostal hernia in a patient with Lumbo-Costovertebral Syndrome, case report and review of the literature

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A B S T R A C T

Congenital subcostal hernias are a very rare type of flank hernia. They are typically asymptomatic but are more difficult to fix as the child grows, warranting early repair. They are typically found in the setting of multiple congenital anomalies. We report a congenital subcostal hernia associated with Lumbo-Costovertebral Syndrome, which is already known to be associated with congenital lumbar hernias. We will report the background and outcome of primary repair in such a hernia.

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1. Case report

A 19 month old male presented to clinic after discharge from the NICU with a right flank bulge. The patient was born full term to a mother of advanced maternal age and type II DM. At birth he was noted to have the right flank bulge (Fig. 1) along with T9/T10 vertebral abnormalities and underdeveloped/splayed out right-sided ribs 9, 10 & 11 (Figs. 2 and 3). The bulge itself was consistent with a hernia, containing a palpable defect with significant increase in size with crying. While admitted to the NICU for failure to thrive he was also found to have hydrocephalus, an ASD, and vesicoureteral reflux causing right-sided hydronephrosis. The patient received a gastrostomy and was discharged home after tolerating feeds.

When seen in the NICU it was felt that the patient would benefit from gaining weight before undergoing repair. After 18 months the patient had gained 6 kg. In addition, the parents reported the hernia was beginning to hinder his mobility, as he was beginning to pull up to attempt to ambulate.

The patient underwent primary repair of the hernia defect via an open approach (Figs. 4 and 5). The hernia, being located just lateral to the right costal margin, was exposed by directly opening the hernia sac. The sac was noted to contain both normal appearing liver and colon. After defining the edges of the defect the final size was measured as 4 cm x 4 cm. After minimal undermining of the skin was performed, the fascial edges were re-approximated using interrupted 2-0 vicryl. Skin was closed using a running subcuticular closure with only minimal excess skin. The patient was admitted overnight for observation and did well.

On follow-up with the patient at 3 months the child’s parents reported that he was walking without issues and his previous hernia site showed no signs of recurrence or wound complication. The patient’s family relocated and was unable to be contacted for any further follow-up.

2. Discussion

Flank hernias are rare in children but when they occur, they tend to occur in the same way as adults, after trauma or surgery [1]. Small subsets of these pediatric flank hernias are congenital in origin [2]. Most of these congenital hernias are termed lumbar...
hernias. They are defined by being in the lumbar triangle. A review of the literature in 1996 by Al-Salem et al. [3], found that only 42 cases had been reported in the literature. The first case report of a congenital lumbar hernia was by Charles Dowd in 1906 [4]. He reported the delayed primary repair of this hernia at 3 years of age. He noted that, with delay, the hernia defect tended to grow with the child, making repair more difficult. Although his repair was successful, today early repair is advocated for these hernias.

Lumbar hernias are known to be associated with Lumbo-Costovertebral Syndrome (LCVS) [2,3,5–7], a disorder involving vertebral, rib, and abdominal wall defects. First reported by Touloukian [7] in 1972, he theorized that the syndrome was due to a single somatic defect occurring at 3–5 weeks embryogenesis which resulted in malformations of the vertebral bodies, ribs, and trunk musculature. They are typically unilateral defects but isolated cases of bilateral hernias have been reported [8].

Even more rare is the congenital subcostal hernia. This hernia is found more anteriorly, as its name implies, in the subcostal region. Although it falls under a different term it is likely that the etiology of both subcostal and lumbar hernias are the same. Both types of congenital hernias tend to occur in association with vertebral [1,3,5–7,9] and rib abnormalities [2,3,5–7,9,10]. Maternal diabetes [3,11] and renal agenesis [3,9,11] have been reported with both hernias. Abnormalities that have been reported with subcostal hernia and not lumbar hernia are the use of fertility treatments [10,11] and heart defects [9,10], such as ASD and VSD.

On review of the literature only 6 congenital subcostal hernias have ever been reported [9–11] with a 7th possibly being mislabeled as a lumbar hernia [6]. Although the etiology of this process is unknown it is felt that the isolated defects are due to an early vascular accident while multisystem cases are due to a developmental gene defect affecting the growth of mesoderm around the 4th–10th week of life [9].

Some case reports advocate an early open repair of these defects to prevent issues [9,11]. Reports have been made discussing the use of laparoscopic techniques [9] for repair as well as the use of mesh [2,5,9]. Although the defect we encountered...
was fairly sizeable, the fascia was brought together without tension and the patient has done well without evidence of recurrence.

3. Conclusion

This case report is the first to report LCVS in a patient with subcostal hernia. This association has already been established with lumbar hernias. Although they have different anatomical locations, they can both be referred to as flank hernias. The diagnosis, treatment, and even the etiology of lumbar and subcostal hernias appear to be similar.

Conflict of interest statement

Authors have no conflicts of interest to disclose.

Consent

Written informed consent has been obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the written consent was available for review by the Editor-in-chief of this journal on request.

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