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Case Report

Paediatrics Section

Congential Lumbar Hernia with Malrotation of Left Kidney and Hydronephrosis in an Infant: A Rare Presentation

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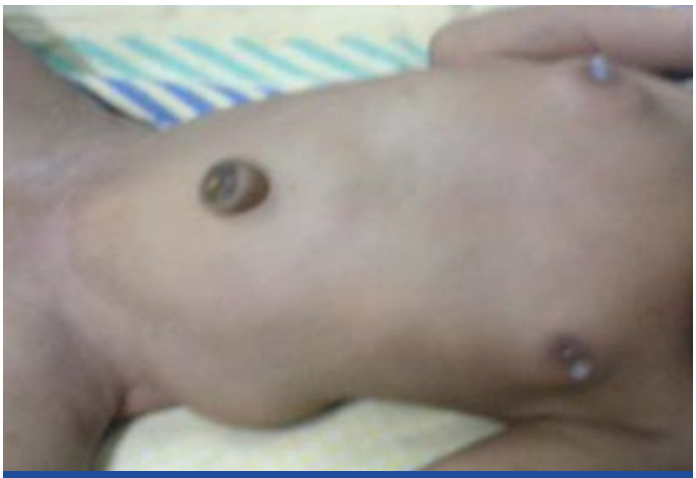
ABSTRACT

Congenital Lumbar hernia was previously known as Lumbocostovertebral syndrome. This syndrome consists of the presence of hemivertebrae, rib defects, abdominal wall anomalies and hypoplasia of the abdominal musculature. This case report describes a 40-hour-old infant with malrotation of left kidney and hydronephrosis in association with congenital lumbar hernia. Various congenital renal anomalies like renal pyelectasis, bilateral renal agenesis and pelviureteric junction obstruction, malascended kidneys, hypospadiasis have been reported in association with this disease. Malrotation has only been reported in adults so far. However ours is the first ever case in literature to report malrotation of kidney in association with this syndrome in neonatal age group. Treatment of this condition is done usually by mesh hernioplasty. Presence of this condition must prompt the surgeon to carry out all possible investigations to rule out various other congenital orthopedic, neurological, and urological anomalies.

Keywords: Lung hypoplasia, Malrotation, Renal anomaly, Scoliosis

CASE REPORT

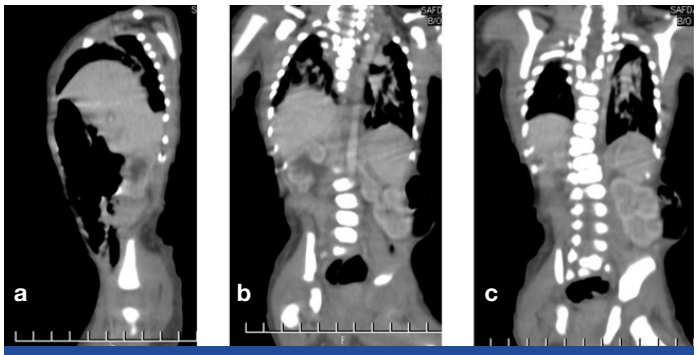
A 40-hour-old baby presented with respiratory distress since birth. The child also had a swelling arising from left lateral abdominal wall since birth [Table/Fig-1]. There were no symptoms related to digestive tract or urinary tract. On physical examination, inspection revealed a left posterolateral body wall swelling, palpation demonstrated a defect size approximately 5 cm in diameter; reducible and non tender, with a positive cry impulse. Spine palpation revealed scoliosis with concavity on the right. X-ray of chest and abdomen revealed scoliosis and 11th thoracic hemivertebrae and right lung hypoplasia and shifting of heart shadow laterally. Cardiac auscultation revealed a point of maximal impulse 1cm outside the mid-clavicular line. Liver dullness was present from 3rd intercostal space downwards. Hematological investigations revealed raised total leukocyte count (25.2 k/l) with predominance of neutrophils. However urine, blood and cerebrospinal fluid cultures were negative. Blood gases showed partially compensated respiratory alkalosis. The sonogram of the abdomen showed left sided lumbar hernia with herniated bowel loops, left sided grade II hydronephrosis with dilated renal pelvis and left sided malrotation of kidney. There were no anomalies noticed on echocardiography. Contrast study of the chest and abdomen were performed which demonstrated right sided eventration of the diaphragm with right lung hypoplasia, malpositioned kidney and grade II hydronephrosis [Table/Fig-2]. Hemivertebrae was noticed at 11th thoracic vertebrae, along with scoliosis, and musculoaponeurotic defect containing herniated bowel loops [Table/Fig-3]. This was further associated with attenuation of the posterolateral body wall musculature comprising of erector spinae, latissimus dorsi and iliocostalis. When the child was operated; the defect was identified in the superior or Grynfellt-Lesshaft's triangle as well as in the inferior triangle. The hypoplastic musculature with the sac was identified during the procedure and dissected all around upto the neck. The bowel loops were reduced and circumferential purse string sutures along with semi absorbable prosthetic mesh was placed in order to contain the defect. The flaps of the overlapping musculature were used for reinforcement of the weak musculature. Postoperative period and recovery was uneventful. The infant was discharged on the 5th postoperative day. Till date, the patient has been following up in the clinics without any recurrence.



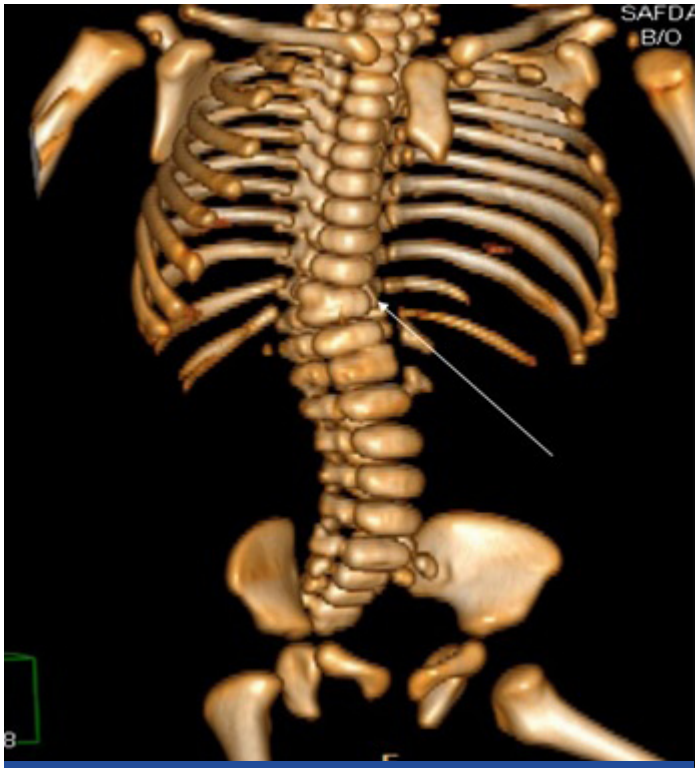
[Table/Fig-1]: Congenital lumbar hernia with mastitis neonatorum

DISCUSSION

Congenital lumbar hernia is a rare syndrome with only less than 50 cases reported worldwide; the first case was reported by Garangeot [1]. It is commonly seen as a part of lumbocostovertebral syndrome; and presents as the association of a congenital lumbar hernia with rib and vertebral anomalies and usually presents at birth but has



[Table/Fig-2]: (a) Contrast study of the chest and abdomen shows right sided eventration of diaphragm. (b) Contrast study revealing right sided Grade II hydronephrosis and malrotation of kidney. (c) The congenital lumber hernia with scoliosis of the spine



[Table/Fig-3]: Hemivertebra at T11 (arrow)

also been reported in older children [2]. The common site of hernia is the inferior triangle and herniation from the superior triangle is less commonly observed. The treatment of lumbar hernia is surgical and should be performed in the first year of life [3]. Several interesting associations of the lumbocostovertebral syndrome have been reported. It has been seen in association with VACTERL anomaly [4]. Various anomalies involving kidneys include renal pelvic pyelectasis, hypospadiasis [4], renal hydronephrosis, Unilateral renal agenesis [5], bilateral renal agenesis [6], crossed renal ectopia and malrotation [7]. Malrotation of kidneys has been reported only in adult patients, our case is one of the first presentation of this kind in an infant. Authors have reported presence of kidneys in the herniated sac causing pelviureteric junction obstruction [8] and have also reported duplication of bladder, urethra and external genitalia as well as malascended left kidney [9]. Our case presented with malrotation of the kidney but the baby had a normal renal function and partial pelviureteric obstruction. The incidence of incarceration in spontaneous lumbar hernia is less likely; as was also observed in our case, and which is in concordance with the findings by other authors [10]. This is due to the fact that the herniated sac has a broad neck; as was also seen in our case. Also the lumbar hernia ring is of a larger size.

Hernioplasty remains the treatment of choice; which can be open or mesh hernioplasy. Cavallro et al., report the successful use of Laparoscopic techniques for the repair of uncomplicated lumbar hernias [11].

Our case is unique as we report malrotation of the kidney and hydronephrosis in association with congenital lumbar hernia. Renal anomalies like renal pelvic pyelectasis , hypospadiasis have been reported previously [8]. However, ours is the first ever case in literature to report malrotation of kidney in association with this syndrome.

CONCLUSION

Congenital lumbar hernia is one of the rare types of hernias reported in neonates; and occasionally in older children. Early recognition of renal anomalies associated with this condition can prevent deterioration in renal function, bowel obstruction, infections involving the region around the kidneys. Thus cases presenting with lumbar hernia should undergo extensive urological workup. This case has been presented to put stress on the fact that malrotation of kidneys in congenital lumbar hernia can occur in pediatric population as well. Thus an ultrasound abdomen should be carried out to rule out malrotation of kidney in a case of suspected congenital lumbar hernia.

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