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Spondylocostal Dysplasia

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Spondylocostal dysplasias (SCD), are a group of disorders in which abnormalities of vertebral segmentation are associated with distortion, fusion or absence of some ribs. Clinically, the trunk is shortened with thoracic asymmetry and spinal deformity.

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Case Report

A 22-year-old second born girl of first cousins, aged 21 and 29 years, was referred with gross chest deformity and respiratory distress. Th elder sibling was a 3-year-old normal female child. The second pregnancy of the mother had ended in spontaneous abortion after four months. No periconceptional factors could be implicated either for the abortion or for birth of this malformed child.

This baby was normally delivered at term with a birth weight of 2 kg, length 43 cm (below 10th centile). She had a deformed chest with a short neck and bulging abdomen. Head circumference, facial features and limbs were normal. *X*-ray chest showed multiple hemivertebrae and block vertebrae. The ribs were reduced in number to only 5 pairs (*Fig.*). Ultrasonography of the abdomen did not reveal any abnormalities. Karyotype of the child and that of parents were normal.

This condition was diagnosed as spondylocostal dysplasia (SCD) with costovertebral segmentation defects of autosomal recessive inheritance in view of parental consanguinity. This patient died at 3 months of respiratory failure.

The parents were advised to report during the subsequent pregnancy for prenatal diagnosis by ultrasonography and were told of the probability of 1 in 4 chance of recurrence. The mother reported at 14 weeks of gestation during the next pregnancy. Ultrasonography revealed a fetus with multiple hemivertebrae, reduced and irregular ribs and a thin abdominal wall, similar to the first child. The pregnancy

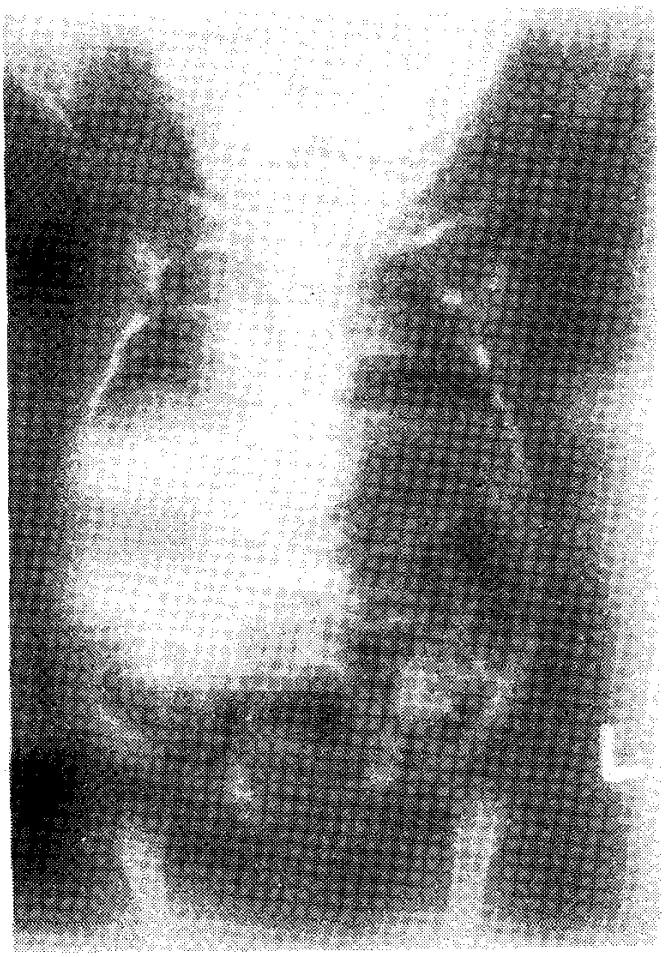

Fig. Infantogram showing reduced number of ribs and multiple hemivertebra and block vertebrae.

was terminated at 18 weeks. Postmortem skiagram of the fetus showed a reduction in number of ribs and hemivertebrae.

Discussion

Isolated costovertebral segmentation anomalies follow a mendelian pattern of inheritance. The severe variety which leads to death in childhood follows an autosomal recessive pattern of inheritance(1,2). A benign autosomal dominant form of SCD is recognized where even asymptomatic adults have been noted(3). Severe defect may be produced by hemozygosity for the allelles(4).

An infant with SCD was born to a woman who had taken lysergic acid diethylamide during early pregnancy(5). A phenotypically normal mother with a translocation of 14-15 chromosomes had given birth to a girl with the same chromosomal anomaly and SCD(4). The causal relationship has not been explained.

Costovertebrae segmentation anomalies can occur in syndromes like VATER and VACTREL. Syndromes have also been reported in association with limb anomalies, mental retardation and renal anomalies(6). SCD has to be differentiated from Spondylo thoracic dysplasia (STD), also lethal in infancy with a characteristic crablike radiographic appearance of the chest, named after Jarcho and Levin who described it(7).

Death often occurs in SCD in infancy or early childhood due to respiratory failure. Spinal cord compression is also a potentially dangerous complication. The condition is amenable to prenatal diagnosis by ultrasonography after 12 weeks of gestation.

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Benign Cystic Teratoma of the Mediastinum

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Cysts and tumors of the mediastinum may originate from any of the structures contained therein or as a result of a developmental abnormality and are rare in childhood(1). Willis(2) defined a teratoma as a true tumor or neoplasm, composed of multiple tissues of kinds foreign to the part in which it arises. The components are derived from all three embryonic layers and may contain mature or immature skin, hair, teeth, bone, cartilage, CNS, respiratory or alimentary tissue.

In our case, the teratoma contained pancreatic tissue, which is an unusual finding(3). Also, in the last decade, although few cases of teratomas have been reported in Indian literature(4-7), to our knowledge, there has been no case report of a mediastinal teratoma containing pancreatic tissue in a child.

Case Report

A 3-year-old boy, weighting 8 kg, was admitted with complaints of dry brassy cough and progressively increasing breathlessness since last one year, with exacerbation of symptoms for the last 15 days. Clinical examination revealed a conscious, well-oriented child with heart rate 140/min and respiratory rate of 64/min. There was no flushing of the face, dilated veins or edema of head and neck. On chest examination, we noticed a marked left sided precordial bulge with apex beat not well visualized. The trachea was shifted to the right side and pulsations were seen in the right parasternal region. Percussion revealed a dull note in the anterior left hemithorax, extending from the 2nd to 6th intercostal space, right upto the anterior axillary line. The left border of the heart could not be delineated, and on the right side the dull note extended upto 1 cm parasternally. Heart sounds were normal, but heard on the right side of the parasternum. Other systems were normal. The development milestones were normal.

Chest roentgenogram revealed a large mass in the left hemithorax with a mediastinal shift to the right (*Fig. 1*). Two-dimen-

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