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CASE REPORTS

Congenital Spinal Canal Stenosis (CSS): A Case Report of Two Similar Rare Anomalies

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Abstract Congenital spinal canal stenosis is a very rare vertebral disorder. The etiology is not clearly defined. Genetic, nutritional, and environmental factors have all been presumed to play a role in the evolution of this defect. The incidence of congenital spinal canal stenosis is also unknown. To our knowledge, there are no prenatally described cases in literature. We report two unusual cases of congenital spinal canal stenosis associated with rib and vertebral anomalies. Both presented with vertebral anomalies at the thoracic level, congenital talipes equinovarus (CTEV) and rib anomalies. In view of multiple anomalies, both the pregnancies were terminated and the diagnosis was confirmed by postnatal pathological examination.

Case 1

A 25-year-old primigravida was referred at 22 weeks for a second opinion for hemivertebrae. Sagittal view of the spine showed abnormal angulation at the thoracic level. 2D/3D imaging revealed multiple hemivertebrae with bilateral talipes, narrow thorax, hypoplastic lungs and abnormal ribs. The patient underwent termination of pregnancy in view of multiple anomalies. Postnatal pathological examination identified spinal canal stenosis at T5-T8 vertebrae, abnormal thoracic cage with missing and misshapen ribs, bony shelf between ribs 5 and 6, bilateral hypoplastic lungs and bilateral CTEV. Case 1—Figs. 1, 2, 3, 4, 5 and 6.

Case 2

A 30-year-old G3P1 L1 A1 was referred to us at 20 weeks for second opinion in view of an abnormal spine and bilateral talipes equinovarus. 2D/3D imaging findings were a single umbilical artery, completely disorganised lower thoracic and upper lumbar spine leading to lordosis and compression of spinal cord, abnormal ribs and bilateral CTEV. Cervical, upper thoracic and sacral spines appeared normal. In this case, although spinal cord compression was noted prenatally, this finding was attributed to the multiple segmentation defects in the vertebrae. Patient underwent termination of pregnancy considering the guarded prognosis. Postnatal pathological examination showed a single umbilical artery, focal spinal canal stenosis at T10 vertebra, mild thoracic asymmetry (13 ribs on right and 10 ribs on the left), abnormal and misshapen T9 and T10 vertebrae, hemivertebra at T11, abnormal and fused ribs 8–12, vertical left ilium and bilateral CTEV. Case 2—Figs. 7, 8, 9, 10, 11 and 12

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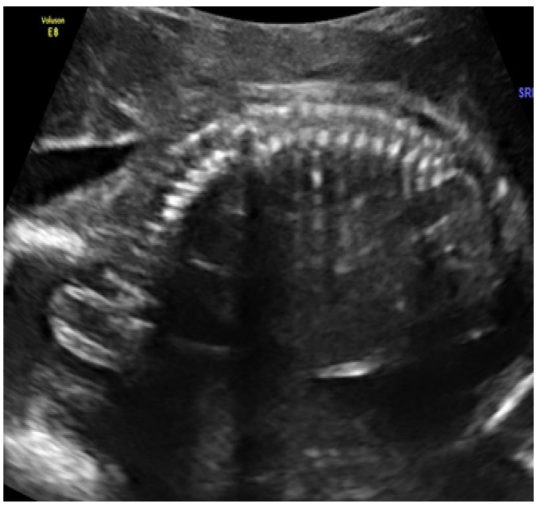

Fig. 1 2D ultrasound sagittal spine showing hemivertebrae in thoracic spine with an echogenic area in the spinal cord level

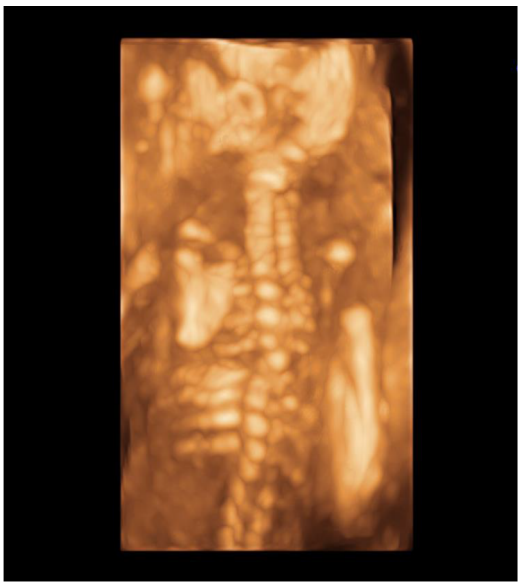

Fig. 2 3D ultrasound showing narrow thorax and abnormal ribs

Discussion

Vertebral anomalies can be classified into vertebral body and neural arch anomalies. Vertebral body anomaly include segmentation defects and hemivertebrae. Defects in the posterior neural arch result in anomalies of the spinal cord like open spina bifida and closed spinal dysraphisms.

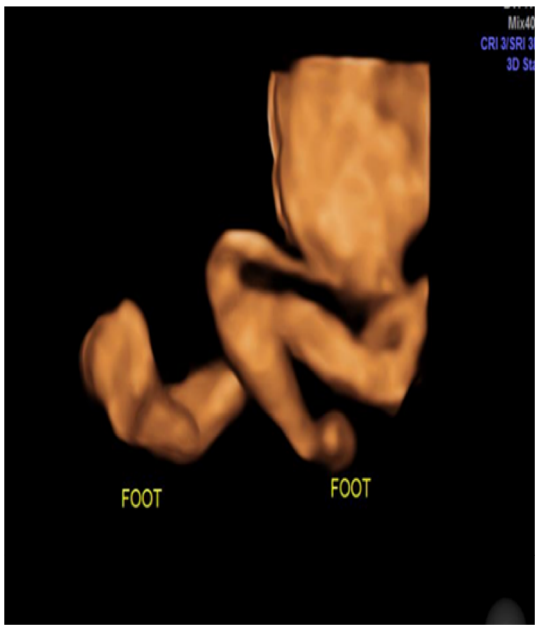

Fig. 3 3D ultrasound showing bilateral CTEV

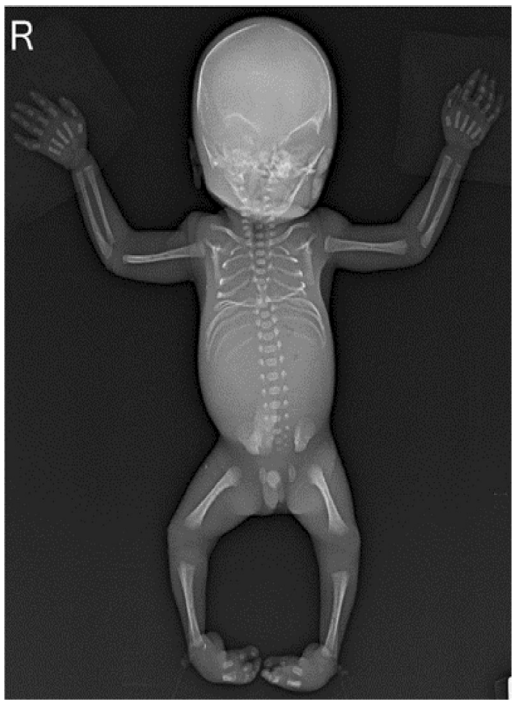

Fig. 4 Fetogram showing narrow thoracic spine, abnormal ribs and CTEV

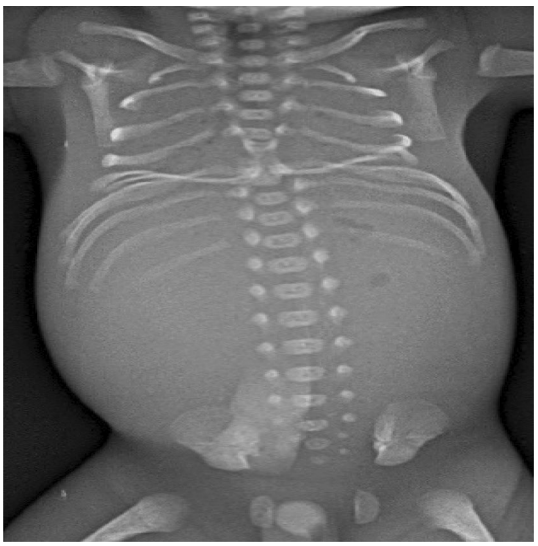

Fig. 5 Fetogram—Abnormal thorax with missing, short and misshapen ribs, vertebral stenosis at T5–T8 and a bony shelf between ribs 5 and 6

Spinal canal stenosis is a rarer entity which also results from posterior neural arch abnormality. The advent of 3D ultrasound has made evaluation of spine more comprehensive. It helps in the evaluation of the entire spine in all three different planes which is not always possible on 2D due to the curvature of the spine. Skeletal mode rendering helps evaluation of the spine, entire rib cage and the scapulae [1–3]. However, congenital fusion or spinal canal stenosis may be better appreciated with MRI [4].

Both our cases had multiple spinal abnormalities associated with rib anomalies and congenital talipes equinovarus. There were no cardiac, central nervous system, gastrointestinal tract or genitourinary tract anomalies to point towards a syndromic association. The similarity noted by ultrasound in these cases was the appearance of an echogenic area in the spinal canal (as a bony spur) which results from the narrowing of arches. The difference in the appearance of posterior arch in the coronal and sagittal plane is that of widening or splaying of the laminae in neural tube defects, whereas in congenital spinal canal stenosis the laminae are seen close together. In diastematomyelia, a type of closed spinal dysraphism, there is widening of the neural arches (laminae) with a bony spur/spicule, which splits the spinal cord into two.

In case 2 of our report**,** spinal cord compression was reported in the prenatal scan due to the close proximity of the bony elements of the laminae in the coronal plane. However at postnatal pathological examination, X ray of the spine (fetogram) complimented by clear dissection, demonstrated and confirmed spinal canal stenosis, the diagnosis of which was unknown at the time of antenatal scan. Though invasive testing was offered to ascertain chromosomal normalcy and DNA storage, both families opted against the procedure. However considering the rarity of the anomaly and the absence of any other associations like VACTERL, the low risk of recurrence in subsequent pregnancies was explained. The association between spinal and rib anomalies substantiates the common embryological origin from the sclerotome cells.

Implications on Clinical Practice

Open neural tube defects are the most common spinal malformations seen in fetal life followed by hemivertebrae. Spinal canal stenosis, a rarer entity, presents as a constriction of the posterior laminae in coronal scan which results in spinal cord compression. To our knowledge these

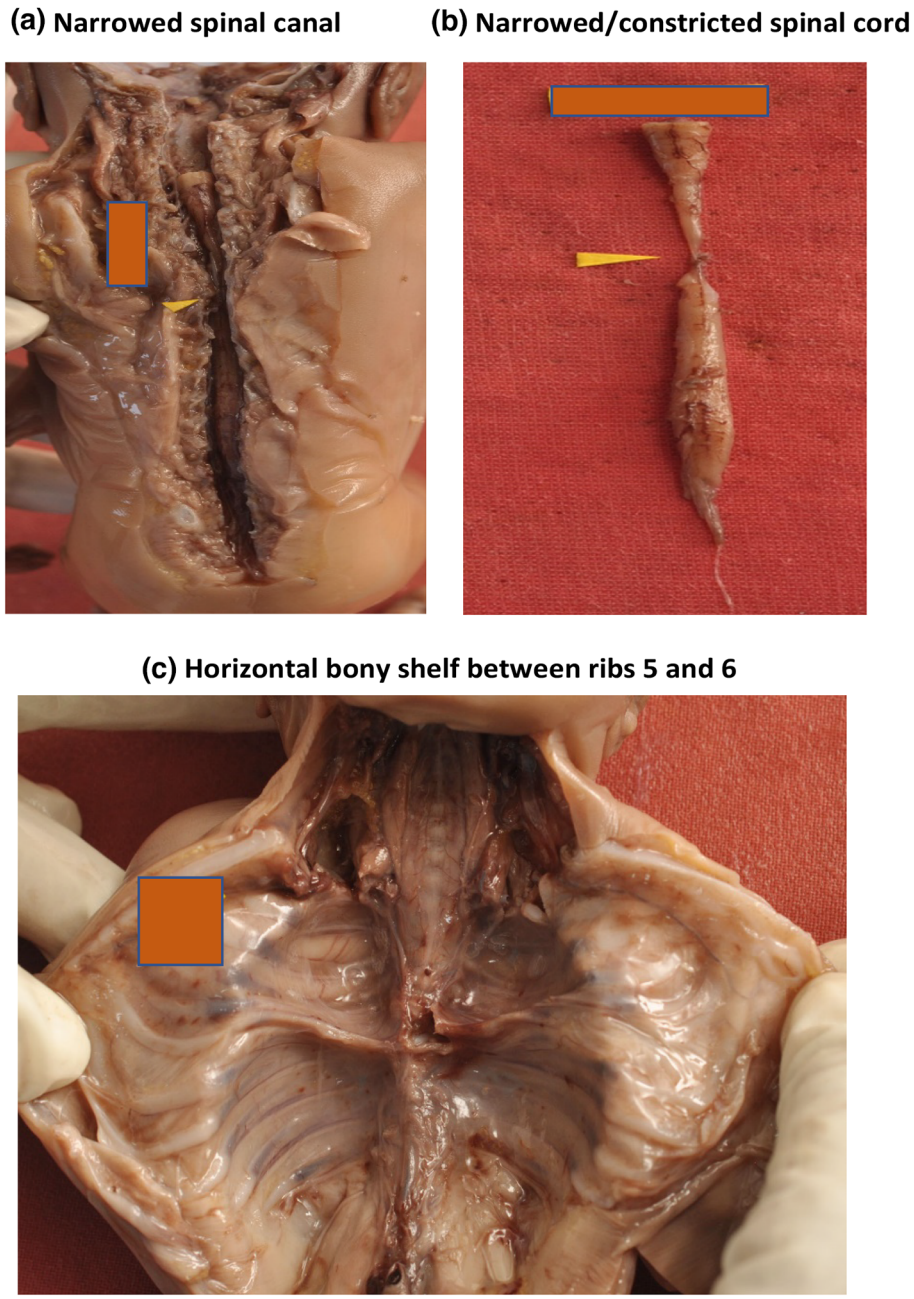

Fig. 6 Postnatal pathology images. **a** Narrowed spinal canal, **b** Narrowed/constricted spinal cord, **c** Horizontal bony shelf between ribs 5 and 6

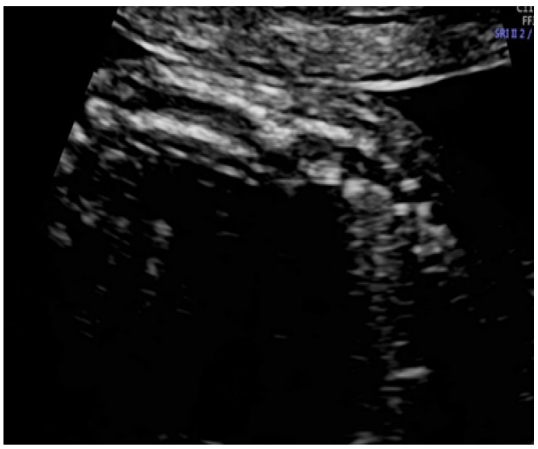

Fig. 7 2D sagittal view of the spine showing irregular vertebrae with an echogenic area in the spinal cord level

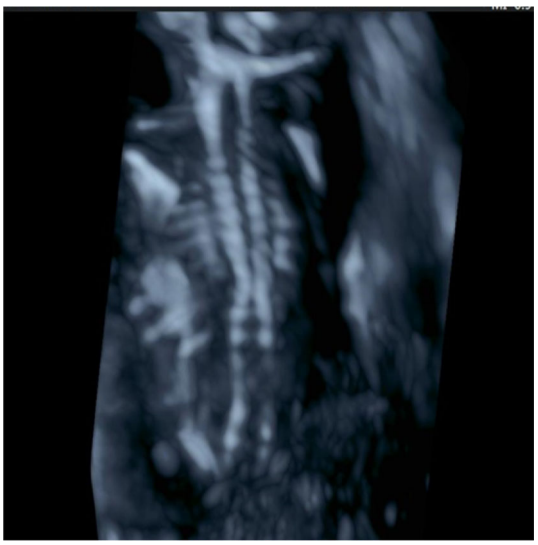

Fig. 8 3D ultrasound showing a narrowed spinal canal and neural arches

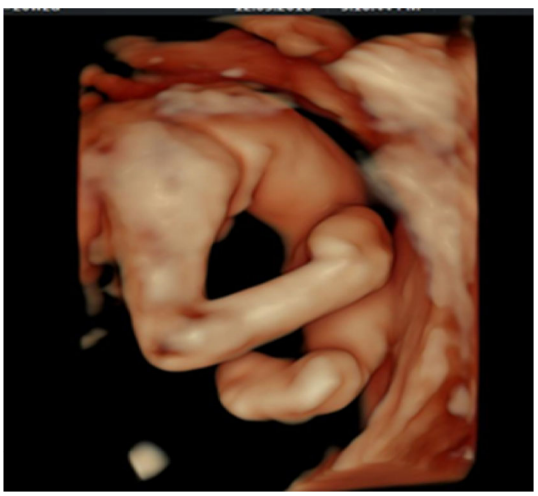

Fig. 9 3D ultrasound showing a bilateral CTEV

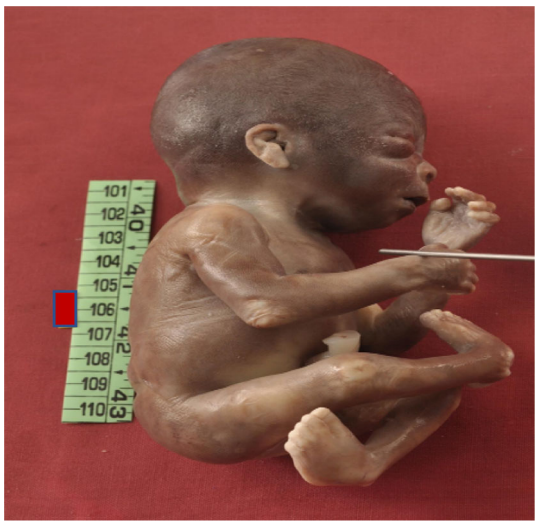

Fig. 10 Autopsy: Gross appearance with bilateral CTEV

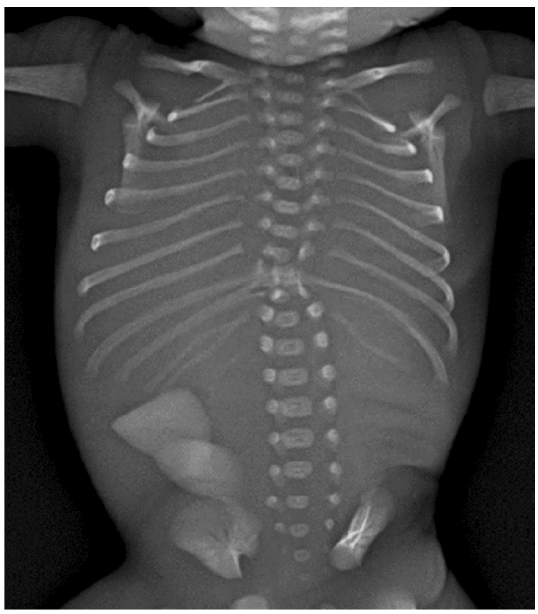

Fig. 11 Fetogram Focal spinal canal stenosis at T10, mild thoracic asymmetry (13 ribs on right/10 ribs on left), abnormal and misshapen T9–T10, hemivertebra at T11, abnormal and fused ribs 8–12 and vertical left ilium

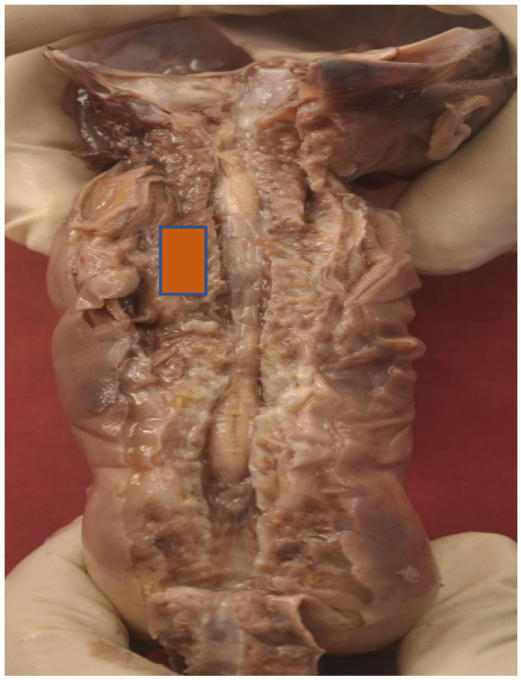

Fig. 12 Autopsy Spinal canal stenosis at T10

are the first prenatally reported cases which were confirmed by postnatal pathological examination.

Declarations

Conflict of interest The authors declare that they have no conflict of interest regarding the publication of this case report.

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