An Unusual Case of Rib Anomalies in Number and Morphology with Association of Vertebral and Sternum Anomalies: Computed Tomography Diagnosis

Dharmraj Meena

Department of Radiodiagnosis and Modern Imaging, Government Medical College, Kota, Rajasthan, India

Correspondence: Dr. Dharmraj Meena, Department of Radiodiagnosis, Government Medical College and Associated Group of Hospitals, Kota - 324 001, Rajasthan, India. E-mail: dr.dharmrajmeena9875@hotmail.com

ABSTRACT

A significantly less number of ribs with multiple developmental anomalies in ribs, spine, and sternum is very rare anomaly. We report a case of a 50-year-old man with 15 number of ribs, absent first, second, fourth, seventh and eleventh ribs on the left and absent first, third, eight and tenth ribs on the right side, along with a bifid anterior end of the left sixth rib, fused posterior ends of right fifth and sixth ribs, and bilateral hypoplastic 12th ribs. These anomalies were also associated with thoracolumbar block and hemivertebrae as well as a small sternum. All findings were identified incidentally on computed tomography. No similar type of rib anomalies was found in previous literature. We discuss the details of this case, and suggest a perspective of morphology and developmental biology.

Key words: Jarcho Levin syndrome; rib anomalies; vertebral anomalies

Introduction

A case of rib anomalies in number and morphology with association of vertebral and sternum anomalies is found unique. Normally, the number of ribs are 24 (12 on each side) was noted by the Flemish anatomist Vesalius in his key work of anatomy- De humani corporis fabrica- in 1543.[1] Variations in the number of ribs occur. About one in 200–500 people has an additional cervical rib, and there is a female predominance.[2]

Congenital vertebral anomalies include alterations of the shape and number. Among the congenital vertebral anomalies, hemivertebrae, transitional vertebrae, and block vertebrae are included.[3,4]

Case Report

A 50-year-old man presented in the medical out-patient department with complaint of right hip pain for 6 months, for which a chest computed tomography (CT) examination of the chest and upper abdomen was done following a wrong suspicion of a lower chest pain. Incidentally, we noted absence of the first, second, fourth, seventh, 11th ribs on the left side and the first, third, eight and tenth ribs on the right side, along with a bifid anterior end of the left sixth rib, fused posterior ends of right fifth and sixth ribs, and bilateral hypoplastic 12th ribs; thus, the total number of normal and abnormal ribs was 15 [Figures 1-3]. In vertebral anomalies, first thoracic vertebra has the characteristics of two types of vertebrae, normal right transverse process, and cervical foramina in enlarged left transverse process (cervicothoracic transitional). The second thoracic vertebra was normal. The left sided third thoracic vertebra was hemivertebra and fused with right sided fourth thoracic hemivertebra. The left sided fifth thoracic was hemivertebra and fused with right sided sixth thoracic hemivertebra. The right-sided seventh thoracic hemivertebra and left sided eighth thoracic hemivertebra were fused. The right-sided ninth thoracic hemivertebra and left sided 10th thoracic hemivertebra were fused. The 12th thoracic vertebra was block vertebra with right sided 11th thoracic vertebrae and showed enlarged transverse processes bilaterally. The first and third lumbar vertebrae were fused with left sided second lumbar hemivertebra. Fourth lumbar vertebra was partially hemivertebra [Figures 4-6]. Sternum was small in size with normal manubrium, three fused segments body and tapered xiphoid [Figure 1]. Cervical

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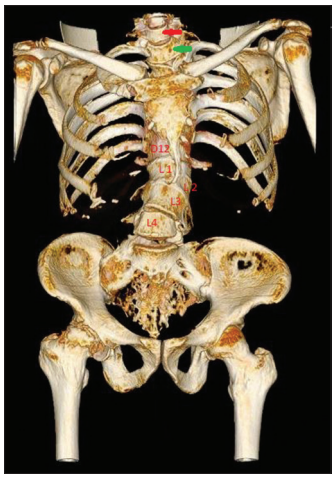

Figure 1: Computed tomography volume rendering technique anterior view image of patient showing less numbers and anomalous ribs, small sternum, transitional T1 (red arrow), normal T2 (green arrow), and four numbers lumbar vertebrae with L2 hemivertebra (red annotations)

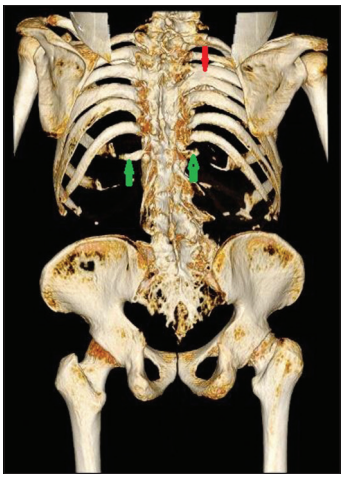

Figure 2: Computed tomography volume rendering technique posterior view image of patient showing fused posterior ends of right fifth and sixth ribs (red arrow) with hypo plastic bilateral twelfths ribs (green arrows)

Figure 3: Computed tomography volume rendering technique lateral image of patient showing bifid anterior end of left sixth rib (red arrow) and lumbar hemivertebra (green arrow)

Figure 4: X-ray dorsolumbar anteriorposterior view showing multiple hemivertebrae and blocked vertebrae

Figure 5: X-ray dorsolumbar lateral view showing multiple blocked vertebrae

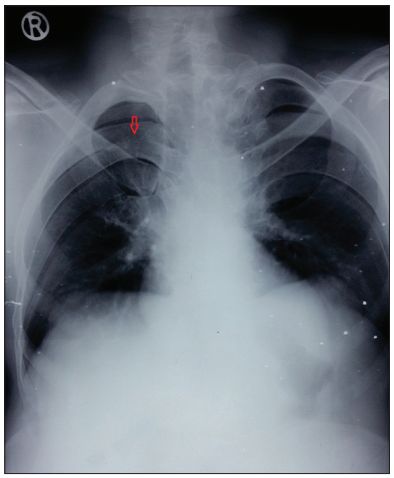

Figure 6: X-ray chest (posterioranterior) view showing less numbers and anomalous ribs and posterior ends fused ribs (red arrow)

spine, both scapulae, and clavicles appear normal. The patient had no dyspnea or chest infection. His intrathoracic

organs were not affected despite the multiple vertebral and ribs anomalies. Extremities were normal. Abdominopelvic ultrasound scanning was normal.

Further CT scan examination of both hip joints showed clumping and distortion of trabeculae in the right femoral head. The patient had a history of trauma. These findings were favor in right humeral head avascular necrosis.

Discussion

A typical human rib cage consists of 24 ribs, the sternum, costal cartilages, and the twelve thoracic vertebrae. The number of ribs was noted by the Flemish anatomist Vesalius in his key work of anatomy De humani corporis fabrica in 1543, setting off a wave of controversy, as it was traditionally assumed from the Biblical story of Adam and Eve that men's ribs would number one fewer than women's.[1] Variations in the number of ribs occur. About one in 200 people have an additional cervical rib and there is a female predominance.[2] All ribs are attached in the back to the thoracic vertebrae. The upper seven true ribs (costae verae, vertebrosternal ribs, I-VII), are attached in the front to the sternum by means of costal cartilage. Due to their elasticity they allow movement when inhaling and exhaling. The eighth, ninth, and 10th ribs are called false ribs (costae spuriae, vertebrochondral ribs, VIII-X), and join with the costal cartilages of the ribs above.[5] The 11th and 12th ribs are known as floating ribs (costae fluitantes, vertebral ribs, XI-XII), as they do not have any anterior connection to the sternum.

Abnormalities of the rib cage include pectus excavatum ("sunken chest") and pectus carinatum ("pigeon chest"). Bifid or bifurcated ribs, in which the sternal end of the rib is cleaved in two, is a congenital abnormality occurring in about 1.2% of the population. The rib remnant of the seventh cervical vertebra on one or both sides is occasionally replaced by a free extra rib called a cervical rib.

Twelve paired ribs develop from cartilaginous costal processes of the developing thoracic vertebrae. Rib development begins at 9 weeks; secondary ossification centers appear at 15 years.[6,7] The first seven "true" ribs connect to the sternum via the costal cartilages by day 45. The lower five "false" ribs do not articulate with the sternum. Developmental rib abnormalities may be isolated or occur in association with other congenital anomalies.

Cervical ribs arise from the seventh cervical vertebra. They resemble hypoplastic first thoracic ribs, from which they are reliably differentiated by means of the adjacent transverse process, which is angulated inferiorly in the cervical spine and has a more cranial course in the thoracic spine. The reported prevalence of cervical ribs varies from 0.2% to 8%.[8] Cervical ribs occur unilaterally or bilaterally.

Congenital vertebral anomalies include alterations of the shape and number. Among the congenital vertebral anomalies, hemivertebrae are wedge shaped vertebrae. The most common location is the midthoracic vertebrae, especially the eighth (T8).[3] The probable cause of hemivertebrae is a lack of blood supply causing part of the vertebrae to not form. Block vertebrae occur when there is improper segmentation of the vertebrae, leading to parts of or the entire vertebrae being fused. The adjacent vertebrae fuse through their intervertebral discs and also through other intervertebral joints and this can lead to an abnormal angle in the spine. Certain syndromes like Klippel–Feil syndrome are associated with block vertebrae. They are most commonly an incidental finding or are associated with the Klippel–Feil anomaly. The Klippel–Feil syndrome characterized by the congenital fusion of any two of the seven cervical vertebrae due to the presence of a congenital defect in the formation or segmentation of cervical vertebrae. Short ribs do not extend as far anteriorly as the sternum. Supernumerary ribs may rarely be seen incidentally as a normal variant. Increased numbers of ribs are seen in trisomy 21 syndrome and with the vertebral anomalies, anal atresia, tracheoesophageal fistula and/or esophageal atresia and renal anomalies (VATER) association.[9,10] In the VATER association, vertebral anomalies are common and they tend to occur together ribs anomalies in numbers and morphology more frequently. It is more common to see 11 pairs in the absence of associated anomalies; this situation occurs in 5–8% of normal individuals. Eleven pairs of ribs occur in one-third of patients with trisomy 21 syndrome,[10] as well as in association with cleidocranial dysplasia and campomelic dysplasia. Multiple vertebral and rib anomalies are present in Jarcho Levin syndrome, a congenital disorder characterized by the presence of rib and vertebral defects at birth. This syndrome is usually diagnosed in newborns with short neck, trunk, and stature. They present multiple vertebral anomalies at different levels of the spine, including "butterfly vertebrae," hemivertebrae, and fused hypoplastic vertebrae. The small size of the thorax in newborns frequently leads to respiratory compromise and death in infancy.[11] We report a case with vertebral and rib anomalies without respiratory problems in a 50-year-old patient.

Short ribs constitute an integral part of several syndromes, e.g. short rib polydactyly syndrome. In pediatric age group, definitive diagnosis of the type of any skeletal dysplasia requires a complete skeletal survey. On thoracoabdominal CT, we found congenital anomalies in ribs, thoracolumbar vertebrae, and sternum. Normally, skeletal survey should be performed early in the neonatal period to exclude a lethal dysplasia and short narrowed chest with critical respiratory distress; in our case, this was done in a patient that was 50-year-old. The patient had no dyspnea or chest infection. His intrathoracic organs were not affected despite the multiple vertebral and rib anomalies.

Summary

Most of these cases present at birth and early pediatric age with respiratory problems due to short and narrow thoracic cage. These anomalies lead to early mortality hence cannot be evaluated in older age groups. The presented case of rib, vertebrae, and sternum anomalies without associated respiratory problems later in this age group is so far not documented in literature, which makes it a very rare case. This suggests that vertebral and thoracic cage anomalies can present without respiratory signs and symptoms; so any asymptomatic external morphological variation should undergo further radiological workup and future morbidity can be prevented.

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