Segmental Costovertebral Anomaly Presenting with Horn-like Appearance in the Chest

Cagli B., Unlu E., Tuncel S. A., Temizoz O., Genchellac H.
Department of Radiology, Trakya University School of Medicine, Edirne, Turkey

Received June 10, 2010; Accepted August 6, 2010.

Key words: Costovertebral anomaly – Rib – Horn-like appearance – Chest X-ray – CT

Abstract: Several types of abnormalities of the ribs have been reported. Many of these anomalies usually result in the correct diagnosis thanks to the characteristic radiological appearances. But some of the anomalies are rare and it is important to recognize and get accustomed to differing appearances of these osseous abnormalities in order to differentiate the pathological processes and to diagnose the syndromes which may accompany rib anomalies. The localized costovertebral anomaly with horn-like appearance, which has not yet been described in literature, is presented in this paper.

Mailing Address: Assoc. Prof. Ercument Unlu, Department of Radiology, Trakya University School of Medicine, 22030 Edirne, Turkey; Phone: +902 842 357 553; Fax: +902 842 352 730; e-mail: drercument@yahoo.com

Introduction

Manifestations of many congenital and acquired conditions can be seen in the ribs of child or adult patients in the radiological practice (Glass et al., 2002). These abnormalities are usually considered to be either isolated abnormalities or a part of a generalized skeletal disorder. Some types of rib anomalies are usually well-recognized; however some anomalies are rather rare. Currently, more than 100 syndromes involving rib abnormalities have been identified (Jeung et al., 1999; Hannam et al., 2000). Therefore it is important that the radiologists and clinicians knows the radiological appearances of variation and local rib anomalies to be able to differentiate these anomalies from pathological conditions (Donnelly et al., 1999; Kurihara et al., 1999).

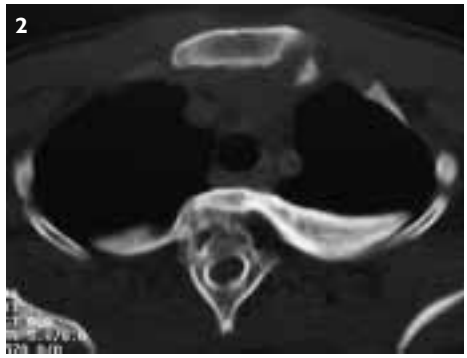
In this paper, not yet described localized thoracic costovertebral anomaly is presented, documented with chest X-ray, computed tomography (CT) and with its three dimensional (3D) reconstructed images.

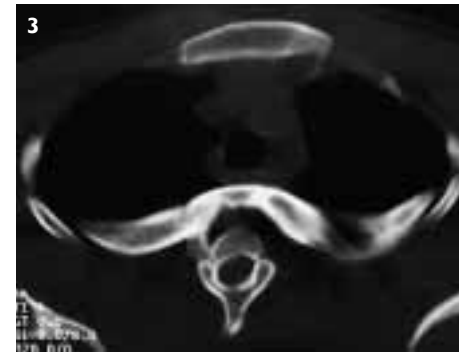
Case report

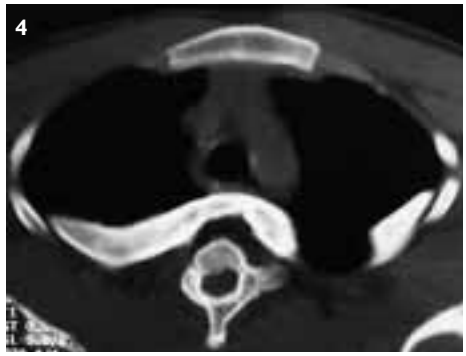
A 21-year old-boy applied to our hospital with non-specific complaints and following the physical examination, chest X-ray was taken. He had no history of any type of significant trauma, severe infection or surgical operation. Posterior closure defect abnormalities at the upper dorsal region vertebrae accompanied with abnormal course and architecture of the ribs at the same level are revealed at frontal chest X-ray of the case. Bilaterally close proximity of the posterior parts of the 4^{th} and the 5^{th} ribs in the chest X-ray. The density of the clavicle and the ribs were normal. There were 12 ribs at each of the hemithoraces. Hyperlucent appearance was observed in the middle zone due to the abnormal course of

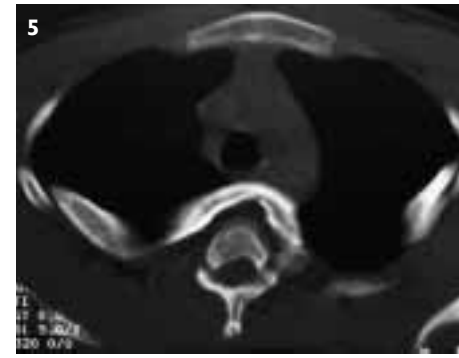
Figure 1 – In the upper dorsal vertebrae, posterior closure defects; disorganization in the ribs; and in the bilateral midthoracal zones, hyperlucent areas free of rib superpositions are seen on the frontal chest radiography.

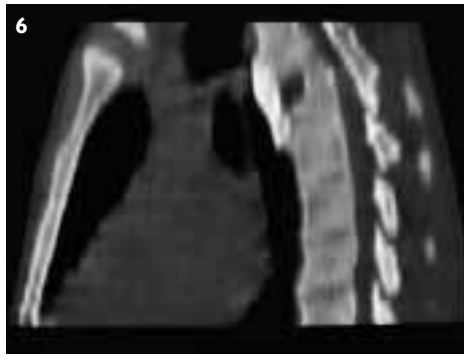
costae (Figure 1). It was observed that both 4th ribs were fused in the prevertebral area and through their course the proximal and distal segments was localized more anteriorly compared to the corresponding parts of the other ribs in the CT scan of the patient. It was also seen that the anomalous and bilateral 4th rib were wider than the other ribs. The structure of cortex and medulla of the anomalous rib observed was ordinary. At this level hemivertebra was the accompanying abnormality (Figures 2–6). Sternum was morphologically normal. No vascular structural or other mediastinal abnormality detected. Pulmonary segmentation











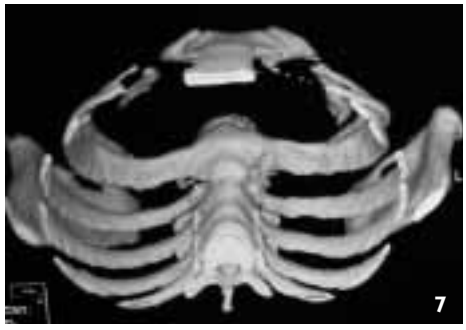
Figures 2–6 – At the prevertebral area at the level of 4th ribs, rib fusion and hemivertebra anomaly are seen in the consecutive axial CT slices and sagittal two-dimensional reconstructed images through the upper dorsal level.

and parenchymal aeration were normal. Sagittal and coronal dimensions of both hemithoraces were normal. Bilateral rib fusion at the costovertebral junction was observed on the reconstructed chest 3D CT scans. Three-dimensional reconstruction images of CT examination displayed horn-like appearance of the abnormal ribs (Figures 7 and 8). The abdominal sonography of the patient was normal. No visually detectable external morphological pathology was seen in this case.

Discussion

After the 3rd week of embryogenesis, the somites formed of paraxial mesoderm differentiate into three distinct areas: the dermatome, myotome, and sclerotome. While medial part of the sclerotome is specifically deemed to form the vertebral mesenchymal anlages, ventrolateral part develops into the ribs in the thorax in the 9th weeks of embryogenesis. These mesenchymal parts finally fuse and ossify to form the bones of the vertebrae and ribs. Persistence and overgrowth of the notochord may result axial skeletal disorganization. At this mesenchymal formation phase, stagnation (teratogenous or related to other causes) or a deformation in the vertebral column and ribs might occur as congenital anomalies (Huang et al., 2000; Nacke et al., 2000; Qi et al., 2004; Aoyama et al., 2005).

Including cervical rib, developmental fusion of two or more ribs, articulation or bridge formation between two ribs, bifid rib, abnormal number of ribs, abnormal size or shortening of ribs, abnormal bone density and abnormal rib shape are some of the examples of various types of congenital anomalies and deformities of the ribs. Some of these anomalies that are isolated generally have little or no clinical significance, some of them may be a part of a generalized skeletal or a metabolic disorder. The existence of over 100 or more kinds of syndromes that are related with rib anomalies is known (Jeung et al., 1999; Kurihara et al., 1999; Hannam et al., 2000; Glass et al., 2002). In this case, no associated anomaly was present, so it was thought that this abnormality was not related to any syndrome.



Figures 7 and 8 – In the reconstructed three-dimensional CT images, the anomalous ribs are seen to localize more anteriorly on their posteroanterior course compared to the other normal ribs and they form horn-like shape.

Pathological processes of the bony thorax may include congenital and developmental anomalies, inflammatory and infectious diseases, and tumours. In most patients, the bony thorax anomalies are revealed on the posteroanterior X-ray of the chest which is usual routine initial imaging study. Exact diagnosis is frequently possible because the lesion has a typical radiological appearance. On the other hand, radiologists and clinicians sometimes have some difficulty in detecting, localizing and defining these anomalies on the chest X-ray images. As in this case, advanced cross-sectional imaging methods such as CT and its reconstructed images provides detailed information about abnormalities. Besides, CT or magnetic resonance imaging is preferred for detection and diagnosis of inflammatory or neoplastic masses on the chest (Jeung et al., 1999; Kurihara et al., 1999; Glass et al., 2002).

Morphologic anomalies and anatomic variants such as developmental fusion of two or more ribs and articulation or bridge formation between two ribs occur in 0.15–0.31% of the population and occur either on the left or right side (Kurihara et al., 1999). On the other hand, hemivertebra and segmentation defects were realized on the upper dorsal parts of vertebrae, besides, posterior fusion was also found at this level of the 4th bilateral ribs in the prevertebral localization in this case. As far as it is known, this kind of an anomaly has never been identified. In cases similar to this case, where there is a localized rib fusion, no clinical significance has been found or thought to have a minor importance (Kurihara et al., 1999; Glass et al., 2002). Although there are minor similarities (multiple vertebral segmentation anomalies, sickle shape appearance of the vertebral body, posterior rib fusion etc.) between the Jarcho-Levin syndrome and this case, more widespread involvement is seen in vertebrae and ribs in the spondylocostal or spondylothoracic dysplasia. These cases recognized with related findings and can be diagnosed due to the respiratory problems and typical appearances in the new-born period or sonography in the intrauterine period. Crab-like configuration of the bony thorax is typical for the patients of such cases. Also, associated features in spondylocostal dysostosis include congenital heart disease, urogenital and anal anomalies, limb abnormalities, torticollis, and diaphragmatic, umbilical and inguinal hernias (Roberts et al., 1988; Cornier et al., 2004). In this case, associated anomalies were not seen, and the bony thorax anomaly was localized. It can be speculated by the help of these findings that there were changes and deformations in one or more sclerotome of somites during embryogenesis. However, this case could not be analyzed genetically due to lack of equipments.

As a result; radiological and 3D CT appearance of this localized costovertebral anomaly, which was identified in this case, has not been described in literature before. Even though the anomaly identified in this case has no major clinical significance, it is still important to diagnose and differentiate the real rib disorders from these kinds of anomalies and variations.

References

Aoyama, H., Mizutani-Koseki, S., Koseki, H. (2005) Three developmental compartments involved in rib formation. *Int. J. Dev. Biol.* **49**, 325–333.

Cornier, A. S., Ramirez, N., Arroyo, S., Acevedo, J., García, L., Carlo, S., Korf, B. (2004) Phenotype characterization and natural history of spondylothoracic dysplasia syndrome: a series of 27 new cases. *Am. J. Med. Genet. A* **128**, 120–126.

Donnelly, L. F., Frush, D. P., Foss, J. N., O'Hara, S. M., Bisset, G. S. III. (1999) Anterior chest wall: frequency of anatomic variations in children. *Radiology* **212**, 837–840.

Glass, R. B., Norton, K. I., Mitre, S. A., Kang, E. (2002) Pediatric ribs: a spectrum of abnormalities. *Radiographics* **22**, 87–104.

Hannam, S., Greenough, A., Karani, J. B. (2000) Rib abnormalities arising before and after birth. *Eur. J. Pediatr.* **159**, 264–267.

Huang, R., Zhi, Q., Schmidt, C., Wilting, J., Brand-Saberi, B., Christ, B. (2000) Sclerotomal origin of the ribs. *Development* **127**, 527–532.

Jeung, M. Y., Gangi, A., Gasser, B., Vasilescu, C., Massard, G., Wihlm, J. M., Roy, C. (1999) Imaging of chest wall disorders. *Radiographics* **19**, 617–637.

Kurihara, Y., Yakushiji, Y. K., Matsumoto, J., Ishikawa, T., Hirata, K. (1999) The ribs: anatomic and radiologic considerations. *Radiographics* **19**, 105–119.

Nacke, S., Schafer, R., Habre de Angelis, M., Mundlos, S. (2000) Mouse mutant "rib-vertebrae" (rv): a defect in somite polarity. *Dev. Dyn.* **219**, 192–200.

Qi, B. Q., Beasley, S. W., Arsic, D. (2004) Abnormalities of the vertebral column and ribs associated with anorectal malformations. *Pediatr. Surg. Int.* **20**, 529–533.

Roberts, A. P., Conner, A. N., Tolmie, J. L., Connor, J. M. (1988) Spondylothoracic and spondylocostal dysostosis. Hereditary forms of spinal deformity. *J. Bone Joint Surg. Br.* **70**, 123–126.