Bifid Intrathoracic Rib: a Case Report and Classification of Intrathoracic Ribs

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Abstract

Bifid intrathoracic rib is a very rare anomaly of the ribs that is characterized by an osseous prominence of a rib into the thoracic cavity. We report a 21-year-old woman with bifid intrathoracic rib arising from the anterior-lateral portion of a depressed 4th rib, based on findings from chest radiography and computed tomography (CT). This is only the second reported case of this type of intrathoracic rib worldwide. We discuss differential diagnoses for this case and suggest a classification of intrathoracic rib from the perspective of morphology and developmental biology.

Key words: bifid intrathoracic rib, intrathoracic rib, bifid rib, Hox, Pax

(DOI: 10.2169/internalmedicine.45.1502)

Introduction

One percent of the population shows some variation in rib structure, but intrathoracic rib is a rare congenital anomaly. The condition was first described by Lutz as an unusual 4th rib, based on radiological observations (1). Most cases of intrathoracic rib involve articulation with a vertebral body and lateral downward extension, or arise from a posterior portion of a rib. Here, we report a unique case of bifid intrathoracic rib arising from an anterior-lateral portion of a deformed 4th rib, and classify the cases of intrathoracic rib that have been reported in the literature (1-23).

Case Report

A 21-year-old healthy woman underwent a routine chest radiograph during a health check. The radiograph showed a thin line at the inferior-lateral edge of the left 4th rib, and also indicated hyperlucency of the lung (Fig. 1). Physical and laboratory examinations were unremarkable, and there was no past history of fracture, thoracic disease or other serious diseases. Unenhanced helical CT scans of the mediastinal window demonstrated that the left 4th rib was thicker than the right 4th rib and was depressed into the thoracic cavity, and that a linear osseous structure protruded into the thoracic cavity from the anterior-lateral portion of the 4th rib (Fig. 2A). There were no abnormal findings in the lung window (Fig. 2B).

Discussion

Differential diagnoses for the line at the inferior-lateral edge of the 4th rib observed in the chest radiograph (Fig. 1) include an accessory lobe, calcification of pulmonary sequestration and an abnormal 4th rib. The first two possibilities were ruled out because no anomalies were found in the vessels, bronchus and interlobar fissure and no pulmonary sequestration was evident in the chest CT of the lung window (Fig. 2B). Hyperlucency of the lung was also noted in the chest radiograph (Fig. 1), and this may have been due to the conditions used in the X-ray examination or to an effect of the dose on the soft tissue, because no abnormalities were present in the CT of the lung window (Fig. 2B). From these findings, the line at the inferior-lateral edge of the 4th rib was interpreted as an abnormality of the left 4th rib, which was depressed into the thoracic cavity, and this case was diagnosed as bifid intrathoracic rib arising from the 4th rib. Forty-one cases of intrathoracic rib (1, 3-23) have appeared in the literature, but these have not been previously

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Received for publication July 22, 2005; Accepted for publication March 2, 2006

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classified. Here, we suggest classification of this condition into four categories, as presented in Fig. 3A. Type I-a intrathoracic rib is a typical supernumerary intrathoracic rib, arising from an anterior-lateral portion of a vertebral body and extending laterally downwards, passing anterior to the origin of other ribs (2). Type I-b intrathoracic rib is also a supernumerary intrathoracic rib, arising from the posterior portion of a rib close to a vertebral body (proximal rib) and extending laterally downwards, passing anterior to the origin of other ribs (2). Cases of type I-a and I-b intrathoracic rib frequently exhibit attachment to the diaphragm, since the intrathoracic ribs run downwards. Type II intrathoracic rib is a so-called bifid intrathoracic rib; it is a rare variant of bifid rib, with the bifurcation in a distal portion of the normal rib (distal rib) (19). Type III intrathoracic rib is caused by local rib depression, in which one or more ribs is depressed into the thoracic cavity (19).

We would like to propose this morphological classification of the intrathoracic rib from the viewpoint of developmental biology. Considering various knockout mice, other animal experimental models and human disease related to the anomaly of the ribs (24-30), supernumerary intrathoracic rib and bifid intrathoracic rib would most likely be attributable mainly to some alterations in gene expression. The type I intrathoracic rib is associated with an abnormality of vertebra or proximal ribs, and expression of the Hox gene family is reported to be important in the development of vertebra, while the Pax1 gene is responsible for proximal rib development (24-26). The type II condition reflects a bifid rib originating from a distal rib, and it has been reported that bifid rib is found frequently in Gorlin syndrome, which is characterized by a mutation in the PTCH gene (27). Development of the distal rib is controlled by genes that are not responsible for proximal rib development, and the proximal and distal rib also differ in the tissue interactions required for their development (26). The depressed intrathoracic rib would most likely be attributable to some alterations in gene expression and/or to effects of mechanics in the thoracic cavity. The type III intrathoracic rib may be caused by a pressure effect against the chest wall at the time of development of the embryo in the uterus (19), but there is also a possibility of altered gene expression or tissue interactions associated with the orientation of rib development (31, 32). Detailed analysis of the signal transduction system associated with rib development will be necessary to resolve the possible causes of intrathoracic rib.

Figure 1. Chest radiograph showing a thin linear shadow extending along the 4th rib (arrows).

Figure 2. A: Unenhanced helical CT scan of the mediastinal window showing the depressed thicker left 4th rib (arrow) and the bifid intrathoracic rib from the anterior-lateral portion of the 4th rib (arrow). B: Unenhanced helical CT scan of the lung window.
Of the 41 cases of intrathoracic rib in the literature (1, 3-23), we classified 20 as type I-a, 14 as type I-b, 1 as a type Ia+type Ib that has two intrathoracic ribs, 1 as a type I variant that has no articulation with a vertebra or rib, 3 as type II, 1 as type III and 1 as type II+III with characteristics of a type II bifid intrathoracic rib and a type III locally depressed intrathoracic rib (Fig. 3B). To our knowledge, the case described here is the second case of type II+III (1, 3-23). Such observations are of importance, because identification and classification of the characteristic radiological features of intrathoracic rib may help to prevent the performance of unnecessary procedures, including exploratory surgery, which have been used in three previous cases of patients with intrathoracic rib (5).

References