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By

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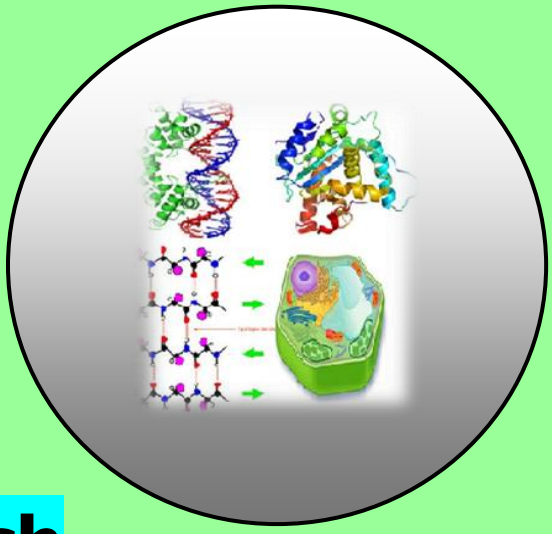
ISSN 2319-3077 Online/Electronic
ISSN 0970-4973 Print

Journal Impact Factor: 4.275
Global Impact factor of Journal: 0.876
Scientific Journals Impact Factor: 3.285
InfoBase Impact Factor: 3.66
Index Copernicus International Value
IC Value of Journal 47.86 Poland, Europe

J. Biol. Chem. Research
Volume 33 (1) 2016 Pages No. 510-513



Journal of Biological and Chemical Research

An International Peer Reviewed / Referred Journal of Life Sciences and Chemistry

Indexed, Abstracted and Cited in various International and National Scientific Databases

Published by Society for Advancement of Sciences®

J. Biol. Chem. Research. Vol. 33, No. 1: 510-513, 2016
(An International Peer Reviewed / Refereed Journal of Life Sciences and Chemistry)
Ms 33/2/33/2016

ISSN 0970-4973 (Print)
ISSN 2319-3077 (Online/Electronic)

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CASE REPORT

Received: 14/05/2016 **Revised: 22/05/2016** **Accepted: 23/04/2016**

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ABSTRACT

Congenital anomalies involving ribs are rare anatomical entity and are incidental findings on routine chest radiographs. Structural defects of first rib is far less common than the other ribs, but if present may lead to compression of neurovascular bundle resulting in Thoracic Outlet Syndrome. We report a case of synostosis of first and second thoracic ribs of the right side. The specimen showed the fusion of posterior ends while the shafts and the anterior ends were free. Although such an anomaly is infrequently seen but if a patient presents with symptoms suggestive of Thoracic Outlet Syndrome, the possibility of such a fusion anomaly should be kept in mind and properly looked for.
Keywords: Rib, Synostosis, Forked and Thoracic Outlet Syndrome.

INTRODUCTION

Ribs are twelve pairs of bony arches which form the osseous framework of thorax. Rib defects are rare anatomical entity and are usually detected during routine radiographical procedures. They are often associated with many syndromes. Few of the syndromes include Klippel- Feil Syndrome, Jarco Levin Syndrome, Poland Syndrome, Basal Cell Naevus Syndrome (Gorlin Syndrome), Neurofibromatosis and Spondylocostaldysostosis (winter and Baraister, 1991). Congenital rib defects can be studied under two categories namely Numerical and Structural. Numerical defects include extra ribs for example cervical, lumbar, sacral ribs or deficient pair of ribs as 11 pair of ribs in Down syndrome. Structural defects include various fusion anomalies and pseudoarthrosis of first rib (Rani et al.2009). Congenital malformations of first rib are rare. Incidence of the fused first and second rib is reported to be 0.3% in a study based on chestradiographs (Elter et al.1944).In the present

study, we report a case of synostosis of first and second ribs. Knowledge of such a condition can be useful for anatomists, radiologists and cardiovascular thoracic surgeons. This condition may cause severe neurovascular compromise and should be corrected surgically without delay (Baumgartner et al. 1989).

CASE REPORT

Present specimen of the anomalous rib was found incidentally from the osteology lab of King George's Medical University, Uttar Pradesh, Lucknow, India. Meticulous study of the rib showed that it was a synostosis of first and second ribs of the right side. Presence of single circular articular facet on the head and scalene tubercle on the inner border of the upper segment confirmed it to be the first rib while presence of two articular facets on the head and a costal groove on the internal surface of the lower segment confirmed it to be the second rib. The ribs were fused at two points. First point of fusion was 1cm and 1.2 cm from the head of the first and the second rib respectively. Second point of fusion was 2.3 cm and 2.5 cm from the head of the first and the second rib respectively. The fusion resulted in the formation of a deep groove. It was 1.2cm long. The longest transverse diameter of the groove was 2.5 cm while the shortest transverse diameter was 0.7 cm, thus obliterating the first intercostal space. There were two separate tubercles, shaft and posterior ends (Fig. 1 & 2).

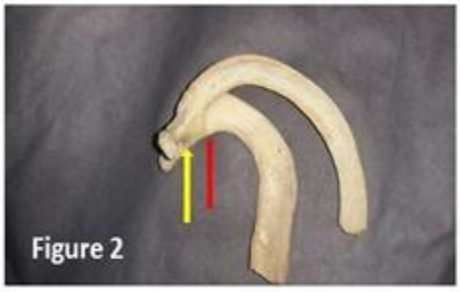

Figure 1. Photograph showing synostosis of first and second ribs (superior aspect).
Figure 2. Photograph showingfirst (yellow arrow) and second points of fusion (red arrow) of thesynostosis of ribs (inferior aspect).

DISCUSSION

Ribs are developed from the sclerotome portion of the paraxial mesoderm which in turn forms the costal processes of vertebrae. The ribs are derived from the costal processes of the thoracic vertebrae. Mal-segmentation of the axial skeleton before 20th day of intrauterine life leads to various structural anomalies of ribs and vertebrae (Helegman et al. 1987). The potential cause of such morphological defects can be attributed to the mal expression of myogenic determination factors like MyoD, Myogenin. Myf 5, MRF 4 which are detected in the medial half of the somites several hours before the formation of myotome (Rani et al.2009).

Fusion defects of ribs can be classified into three major categories i.e. bicipital rib, bridged rib and forked rib. Bicipital ribs have fused anterior ends and shafts but free posterior ends.

Ribs with fused shafts but separate anterior and posterior ends are called bridged ribs. Third types of ribs have fused posterior ends but separate anterior ends and shafts.

The first case was reported in 1740 by M. Hunauld who stated that "The skeleton of an adult in which the first rib on each side is well formed posteriorly and articulated with first dorsal vertebra, joins and fuses with second rib, which by union becomes larger than usual" (Turner 1883).Schaeffer (1947) reported a case of bicipital rib. According to him, this anomaly involves the first thoracic rib exclusively. Synostosis of rib can result from the fusion of two ribs, eithercervical rib and firstthoracic rib or fusion of first and second thoracic ribs which is more commonly encountered. Du Plessis (1975) studied the congenital anomalies of first thoracic rib in great detail. He stated thatin fusion anomalies, the first rib is poorly developed resulting in joining with the second rib by synostosisorpseudoarthrosis or ending freely in a muscle. Anomalies involving the rudimentary first rib are associated with post fixed brachial plexus with contribution from second thoracic nerve(Gupta etal.2009).Rani et al. (2009) reported two cases of synostosis of first and second ribs which were fused at the anterior end and the shafts respectively. Sampada et al.(2014) also reported a case of bicipital rib. The present case differs from the above cases in being fused at the posterior ends while being free at anterior ends and shafts.

In the studied specimen, the anomaly was reported as the synostosis of the first and the second thoracic ribs. As the ribs were fused at the posterior end while the shafts and the anterior ends were free this specimen can be reported as the forked rib.

Due to non-availability of the complete skeleton of the reported specimen, few assumptions regarding the consequences of this malformation could be made. This type of anomaly can make the first intercostal space narrow which may lead to compression of the neurovascular bundle, anomalous articulation of the thoracic vertebrae and formation of abnormal thoracic wall. From these observations it can be said that the reported anomaly can cause signs and symptoms of thoracic outlet syndrome.

Hashimoto et al. (1994) and Iida et al. (1997) reported cases of thoracic outlet syndrome resulting from synostosis of first and second ribs. In both the instances, patients made a good recovery after surgical resection of the involved ribs.

CONCLUSION

Although synostosis of first and second ribs is a rare anatomical entity and an uncommon cause of thoracic outlet syndrome, but the knowledge of such a condition is important for radiologists and cardiovascular-thoracic surgeons. If a patient presents with signs and symptoms of neurovascular compression, the possibility of fusion defect of first rib should be kept in mind and adequately treated.

ACKNOWLEDGEMENTS

We would like to express our gratitude to the staff of Department of Anatomy, King George's Medical University, Lucknow, UP, India for providing infrastructure facilities and help.

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