Original Research Article

Congenital anomalies of ribs and its clinical significance

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Abstract

Introduction: The thoracic skeleton includes sternum, 12 pairs of ribs and associated costal cartilages, 12 thoracic vertebrae and the intervertebral discs interposed between them. The ribs and costal cartilages form the largest part of the thoracic cage. Congenital anomalies of the ribs usually reported are cervical, lumbar or bifid rib and synostosis of ribs. These abnormalities can cause nervous and vascular symptoms and misinterpretation during counting of ribs in physical examination. **Materials and methods**: The present study was carried out on 50 dry adult human ribs belonging to both sides. The morphological variations of bones were studied in detail and noted. **Results:** In first specimen, rib of right side revealed fusion of the two ribs in midshaft region with free anterior and posterior ends and in the second specimen of aleft side rib bifurcation was seen in anterior end. **Conclusion:** This congenital abnormalities may cause compression of neurovascular bundles at the root of the neck causing thoracic outlet syndrome. This abnormalities may also be an indication for few systemic disorders.So knowledge of this congenital anomalies will be helpful to diagnose systemic diseases and also to avoid misinterpretation during counting of ribs.

Keywords: bridged, synostosis, thoracic outlet syndrome

Introduction

The thoracic skeleton forms the osteo-cartilagenous thoracic cage which protects the thoracic viscera and some abdominal organs.It is formed by twelve pairs of elastic archescalled ribs which articulate with vertebral column posteriorly and sternum anteriorly.The twelve pairs may be increased by cervical or lumbar ribs or reduced to eleven by absence of the twelfth pair. The superior seven pairs are connected by costal cartilages to the sternum,called as true ribs. The remaining five are called as false ribs, Cartilages of the eighth to tenth ribs join the superjacent costal cartilage to form costal margin. The eleventh and twelfth being free at their anterior ends are termed floating ribs. Upper ribs are less oblique than the lower,obliquity being maximal at the ninth and decreasing to the twelfth.Regarding breadth,they decrease downwards,in upper ten, the greatest breadth is anterior. The first two and last three present special features, the rest conforming to a common plan classified as atypical and typical respectively. A typical rib has a shaft with anterior and posterior ends. The anterior costal end has a small concave depression for its cartilage.The shaft has an external convexity and is grooved internally near its lower border,which is sharp in contrast to its rounded upper border. The posterior vertebral end has a head, neck and tubercleIt is curved,bent at the posterior angle 5-6 cm from the tubercle and also twisted in its long axis.The spaces between the ribs are called intercostal spaces.There are 11 pairs of intercostal spaces normally.They contain three muscles of respiration:the external intercostal,internal intercostal and the innermost intercostal muscles.The intercostal nerves and blood vessels run between the intermediate and deepest layers of muscles in the costal groove.

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Respiration consists of two phases-inspiration and expiration, which are accomplished by the alternate increase and decrease of the capacity of the thoracic cavity.In inspiration the anteroposterior diameter of the thoracic cavity would be increased if the downward-sloping ribs were raised at their sternal ends.The transverse diameter of the thoracic cavity will be increased when the ribs are raised because the ribs curve downward as well as forward around the chest wall. Both this can be accomplished by fixing the first rib and raising the other ribs to it by contracting the intercostal muscles.Expiration is largely a passive phenomenon[1]. The thinness and flexibility of the ribs allow remarkable flexibility, permitting it to absorb many external blows and compressions without fracture and to change the shape of the thoracic cage repetitively and sufficiently as required for respiration.Rib anomalies are traditionally classified into numerical and structural. Numerical anomalies are common such as supernumerary(cervical/lumbar) and deficient pair of 11^{th} rib.Structural abnormalities are quite rare and they can be further classified as bifid,short,fused ribs.The fusion anomalies can be classified into 3 types(1)Bicipital rib-fused anterior ends and shafts but free posterior ends(2)bridged rib –fused shafts but free anterior and posterior ends(3)Forked rib-fused posterior ends but separate shafts as well as anterior ends[2].Costal anomalies occur frequently at the thoracic outlet.The abnormal 1^{st} and 2^{nd} rib can present with thoracic outlet syndrome.Fusion anomalies of ribs can cause scoliosis and restriction of chest movements which may require surgical correction.Fusion of two ribs can cause nervous and vascular symptoms clinically similar to the presence of cervical rib. Pressure on a nerve causes motor and sensory effects in the structures they supply.Ribs are used as an essential landmark in surface marking during general physical examinations and clinical procedures.Theyalso provide information that aids in the interpretation of radiologic images. Hence it is essential that the radiologist and clinicians be familiar with incidence of rib variants, to

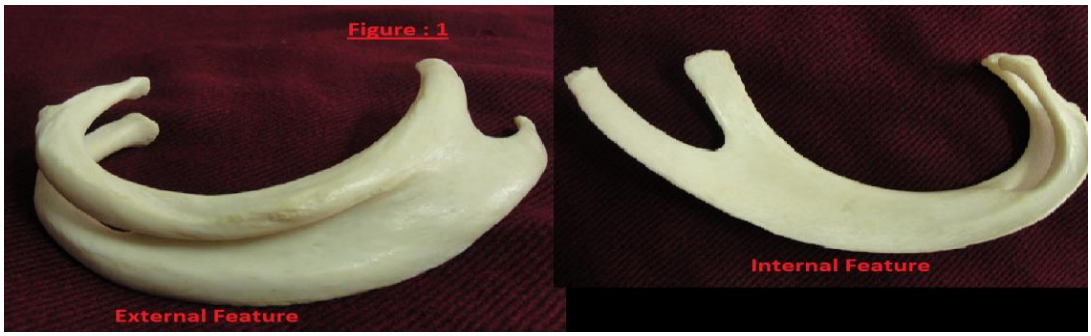
prevent misdiagnosis.This study is done to highlight the congenital anomalies of ribs and its clinical significance.

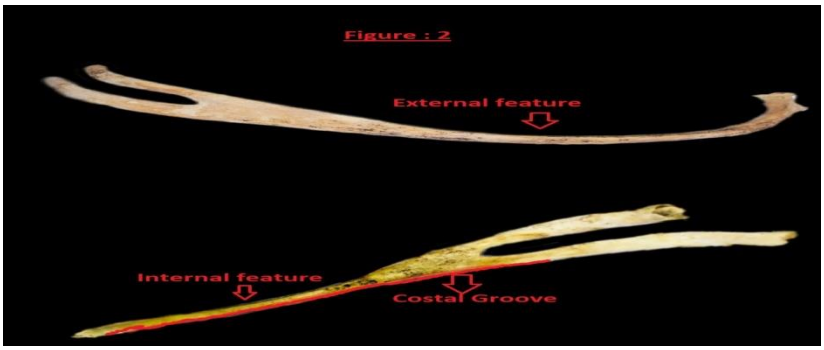
Materials and methods

The present study was carried out on 50 dry adult human ribs belonging to both sides of unknown age and sex in the Department of Anatomy, RajahMuthiah Medical College, Chidambaram. Dry adult ribs of intact features were included in the study. Themorphological features of the bones were studied in detail and the variations were noted.

Results

In the first specimen the morphological examination revealed fusion of the first two ribs of right side in midshaft region with free anterior and posterior ends. Thus the common shaft formed by their junction made it a bridged variety. This has resulted in the obliteration of 1st intercostal space in the conjoint shaft region. But a small intercostal space was present in the anterior and posterior ends. This common shaft had a maximum width of 42.6mm and length of 34.8mm(fig.1). In second specimen,a typical rib of left sidebifurcation was seen in anterior end.This rib had head with articular facets, tubercles,angle and costal groove which are features of typical rib(fig.2).





Discussion

Embryological significance

The paraxial mesoderm is divided by number of transverse clefts into a series of cubical blocks, the mesodermal somites. Each somite differentiates into a ventro-medial part, the sclerotome and a dorsolateral part, the dermo-myotome. During the fourth week of development the polymorphous cells in the sclerotomes of the somites change their position. They migrate ventro-medially around the notochord and form the rudiment of centrum of the blastemal vertebra.Thereafter the cells of the sclerotome extend dorsally between the segmental spinal nerves and dorsal root ganglia to enclose the neural tube. These bilateral extensions form the neural arches which eventually meet at the mid-dorsal line to form the rudimentary spines.Close to the centrum each neural arch gives rise to ventral extensions which extends laterally through the segmental myotomes.These extensions are present not only in the thoracic region but also in the cervical, lumbar and sacral regions.NThey are called as costal process in the thoracic region.They lie ventral to the mesenchymal basis of the transverse processes with which they are continuous. The ribs develop from these ventral extensions of the sclerotomic mesenchymal process of the primitive vertebral arches in the thoracic region.On 35th day ribs begin to lengthen by endochondral ossification of costal process.By 45th day,first seven ribs are connected ventrally to sternum through costal cartilages. Primary ossification centres appear near the angle of each rib in the sixth week and further ossification occurs in a distal direction. Secondary ossification centres develop in the tubercle and heads of the ribs during adolescence·At birth the ventral part of the costal arch remains cartilaginous, since ossification has to extend for a longer distance, and this persists as such throughout life to establish elastic recoil of the thoracic wall.After second year the vertebral ends of the upper ribs migrate headwards and transgress the territory of the upper vertebra. Thus a typical rib articulates with two vertebrae-numerically corresponding vertebra and the one which immediately lies above it[3].Malsegmentation of the axial skeleton before 20th day of embryonic lifeleads to multiplemorphological anomalies of vertebra and ribs. Malexpression of myogenic determination process like Myo D, Myogenin, MYF5, and MRF 4 could be potential cause of such anomalies. Rib abnormalities at the upper end of the thoracic cage are due to errors in segmentation of bony tissue during development and may be associated with variations in the disposition of nerves and vessels[4]. Recent experimental studies in mice have implicated splotch gene mutations for various structural abnormalities in the ribs[5]. Sclerotomal cells of one somite forms the caudal part of one rib and cranial part of the next caudal rib. The dermomyotome derived from one somite contributes to the intercostal muscle, including its origin and insertion. Ablation of the ectoderm which affects dermomyotome development results in severe malformations of the ribs,probably due to disturbed interactions between dermomyotome and sclerotome[6].A possible reason for the congenital anomalies of ribs may take place during the mesenchymal and chondrogenic stages of development around 4th to 6th week of development[7]. Fusion anomalies and bifid rib may be due to irregular segmentation of primitive vertebral arches. Fusion of sternal ends of ribs occurs in old age and is due to ossification spreading into and from costal cartilages.Fusion of vertebral ends of ribs occur

following lesions of intrathoracic contents, especially in long-standing collapse of lung[8].

Clinical significance

Usually rib abnormalities are observed during chest radiography studies or sometimes associated with systemic disesases or syndromes. The first case to be recorded was communicated in 1740 to Royal Academy of Sciences in Paris by M.Hunauldwho stated that the skeleton of an adult in which the first rib in each side is well formed posteriorly and articulated with the first dorsal vertebra, joins and fuses with second rib, which by this union becomes larger than usual[9]. Turner in 1883 described bicipital ribs in two cases.

Synostosis of first and second ribis also called as bicipital rib.Bicipital rib is formed due to the fusion of shafts of two distinct ribs into a common body and is seen exclusively in relation to the first rib, either due to fusion of a cervical rib with first rib or more commonly due to the fusion of the first rib with the second[9]. Rudimentary first rib forming synostosis with second rib was also reported[10,11].In congenital anomalies of first rib, there is post fixation of brachial plexus with a large contribution from T2. As the shoulder girdle sags traction affects the lower trunk causing neurological symptoms[12]. Fused ribs cause curvature of spine towards the area of involvement causing an impediment both for normal lung growth and respiration resulting in a condition which has been aptly called thoracic insufficiency[13]. Abnormal first and second ribs,fused first and second ribs presents with thoracic outlet syndrome. Baumgartner F et al in a radiological study reported two cases of fused first and second rib associated with thoracic outlet syndrome. Fused ribs are encountered in Gorlinsyndrome. Rib fusion will cause scoliosis and restriction of chest wall expansion, which may require surgical intervention.

In a bifid rib the sternal end of the rib will be bifurcated and so it is also called as bifurcated rib .It is a congenital abnormality, occurring in almost 0.15-3.4% of population and accounts for about 20% of congenital rib anomalies. This is generally common in males and occurs more frequently on the third and fourth ribs of right side.Bilateral bifid ribs can also be present[14]. It is usually asymptomatic and in children it may present as a painless lump in the chest wall. It will be an incidental finding during a routine physical examination or noticed in a chest radiography which is taken for other reasons[15]. But few people diagnosed with this condition had complaints of respiratory difficulty, neurological symptoms, chest pain, dyspnea. It is also a finding during routine cadaveric dissection. Kaneko et al reported bifid ribs in nine children out of which seven presented with chest wall mass and two were asymptomatic and were incidentally noted on chest radiographs[16]. Lung growth is limited to the anatomic boundaries of the thorax,So any spine or rib cage malformation that reduces the thoracic volume early in life may adversely affect the size of the lungs at skeletal maturity[17]. Patients with this anomaly become symptomatic when they become involved in sports and other exertional activities.Since ribs originate from mesoderm, malformations of other organs of mesodermal origin may be associated with rib anomalies. Common organs involved are heart and kidneys. It has been stated that there are atleast 22 known syndromes in which fused ribs are a constant component. klippelFeil, JarcoLevin, Poland and Gorlin syndrome are few among them .Abnormalities detected in the ribs can sometimes be the initial indication of systemic diseases. Rib changes can indicate generalized bone dysplasias, metabolic diseases and trauma. Extensive thoracic congenital scoliosis associated with fused ribs may affect thoracic function as a result of three dimensional thoracic deformity and growth and have an adverse effect on the function and growth of the lungsSupernumerary intra thoracic rib is a rare variant of a bifid rib. It may be associated with deformities of a vertebral body. Frequently remains non-symptomatic, but thoracic pain, dyspnea or even hemoptysis have been reported. Other rib anomalies include cervical ribs, supernumerary ribs, short ribs, defect in bone density, abnormal rib shapes.

Comparative anatomy

In Cetacea, more especially in the skeleton of some of the whalebone whales a peculiar bifurcated subdivision of the vertebral end of the first rib has been observed.DrJ.E.Gray of British Museum considered this as a character of much importance for the basis of classification not merely into distinct species but even into new genera.

Conclusion

Synostosis of first and second rib may cause compression of neurovascular bundles at the root of the neck causing thoracic outlet syndrome.Bifid rib may lead to misinterpretation of counting of ribs. .This abnormalities may also be an indication for few systemic disorders. Hence the knowledge about these variations of ribs will be helpful to reduce incidence of misinterpretation during radiological and physical examination,diagnose systemic diseases, and for surgeons operating on the anterior thoracic wall.

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