Bifid Rib with Additional Oval Intercostal Space: A Rare Case of Anterior Chest Wall Deformity

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Abstract

Morphological variations in the anterior osseous thoracic skeleton are a rare entity. Its incidence includes accessory bones atypical fusion or non bony fusion of ribs and accessory foramina. Knowledge of these rare skeletal variants is vitally important for surgeons and radiologist performing thoracic surgeries and counting ribs, because such anomaly could mislead them during the procedure. During the routine dissection of adult male cadaver for undergraduate medical students, we observed a variation of bifurcation of anterior end of right third rib and corresponding costal cartilage. These bifurcated ends were joined together to form oval additional intercostal space. Bifid ribs are usually asymptomatic, and usually detected during routine radiographical procedures. It is peremptory that detection of bifid rib can help early diagnosis of Gorlin's syndrome.

***Key words**: Bifid ribs, Intercostal space, Radiographs, Gorlin's syndrome.*

Introduction

Ribs contribute majority of the thoracic skeleton in the mammals, which are twelve pairs of elastic arches that articulate ventrally and dorsally by sternum and vertebral column respectively, forming normally eleven fair of intercostal spaces. These osseochondral spaces contain thin multiple layers of muscular fibres from outside inwards intercostalis externa, interna and intimi respectively, and intercostal nerves and vessels courses through the intercostal groove formed between intercostalis interna and intimi[1]. The ribs are derived from ventral development of the sclerotomic mesenchyme that designs the vertebral arches, initially ribs develop as part of the cartilage model for each vertebra, but in the thorax region, the rib portion separates from the vertebra by the seventh to eighth week. The ossification of the rib takes place in the cartilage model, except for the anterior portion, which remains as the costal cartilage. Growth of the cartilage models for the ribs, sternum and vertebrae allow the enlargement of the thoracic cavity by providing the protection to vital organs. The thoracic skeleton act as dynamic, as it moves at is its various joints, increasing or decreasing the various diameter of the cavity for an extremely important process of respiration. A bifid rib is a congenital neuroskeletal abnormality of the anterior chest wall, which occurs in about 1.2% of humans[2]. Incidence of bifid rib is more common in male than female [3]. Additional intercostal space results from incomplete fusion of cephalic and caudal segments of sclerotomic mesenchyme during 4th to 6th week of intra uterine life[4]. During the mesenchymal and chondrogenic state of development, faulty fusion or anomalous chondrification may give rise to unusual fusion deformities of the ribs[5].

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Case Report

During routine dissection for medical students in Department of Anatomy, BLDE Deemed to be University's Shri B M Patil Medical College and Research Centre, Vijayapura. Right sided anterior chest wall variation was observed in a 68 year old male embalmed cadaver. Additional oval intercostal deformity was examined in detail and morphological measurements were recorded. Bifid third rib was present enclosing an

additional intercostal space. Its bifurcation point was near its costochondral junction, the dimensions of the additional space were about 3.4 cm in transverse length and 0.7 cm in vertical length. The upper arched bony bar above the window at costochondral junction measured 1cm in length and the lower arched bony bar below the window at costochondral junction measured 1.3 cm in length. The intercostal space between second and third rib was 1cm at costochondral junction and intercostal space between third and fourth rib costochondral junction was 1.8 cm. The additional external intercostal space was covered by thin fibrous external inter costal membrane. Deep to the membrane the internal costal space was covered by internal inter costal muscle, fibres running down and laterally. Posterior to the internal inter costal muscle, fibres of Sternocostalis muscle were extending over this region. The intercostal vessels were entering additional intercostal space from right internal thoracic artery. The 3rd intercostal nerve running in costal groove was giving a twig to additional intercostal space.

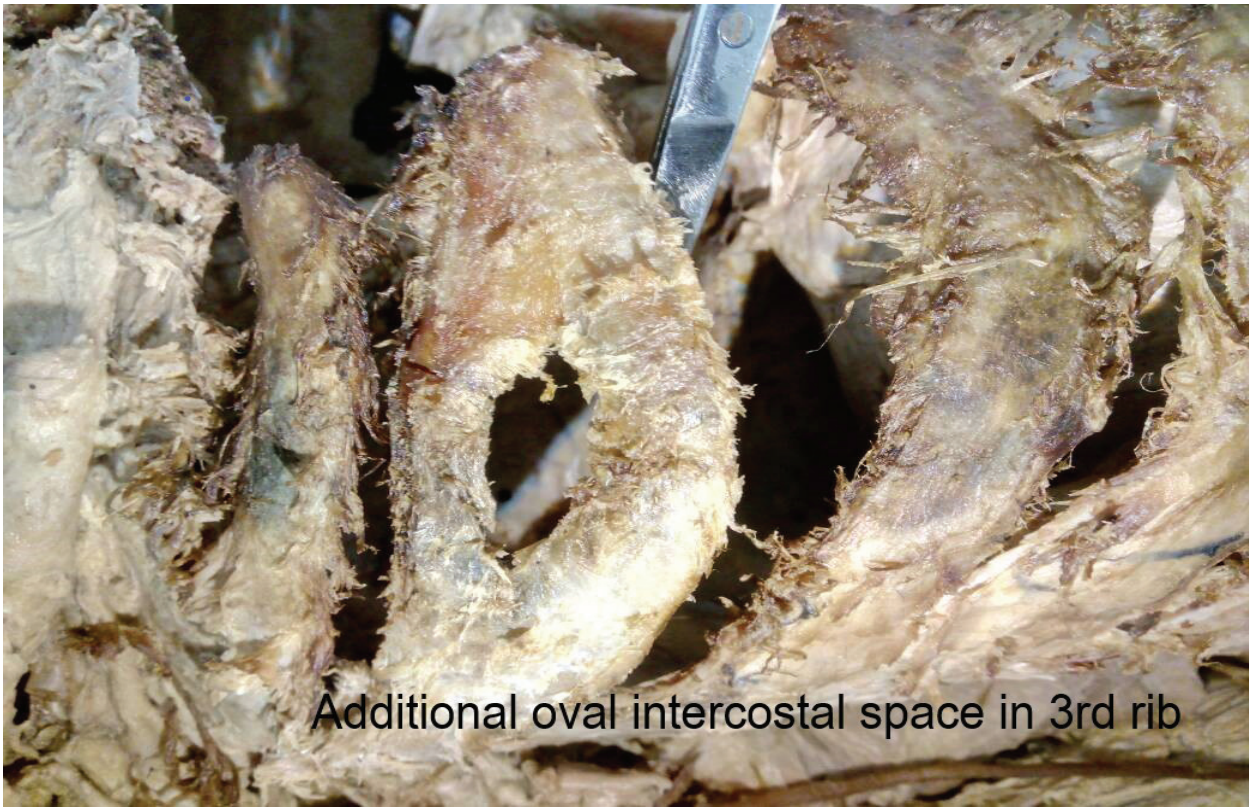

Figure 1 Dissection of Cadaveric thorax of right side showing oval intercostal space intervening between second and third intercostal spaces, encircled by bifid third rib and bifid third costal cartilage

Discussion

Skeletal anomalies in the bones of thorax are recognized as potential cause of thoracic out let syndrome (TOS). Bifurcation of ribs and presence of supplementary intercostal spaces is a very rare variation of the thoracic skeleton. This may be due to gene mutation that strongly alters the anatomy of bone development [6], and bifid rib immensely linked to the alteration of rib development during embryogenesis [7]. The ribs develop from sclerotomal cells of somite at approximately 7th to 8th weeks of gestation, and play a crucial role in subsequent body patterning by governing the formation of all adult segmented structures, the vertebrae and the intervertebral discs, the ribs, muscles, tendons, ligaments, dorsal root ganglia, peripheral spinal nerves, and blood vessels of the adult vertebrate trunk. Disturbance in the segmentation process in vertebrates can result in conditions characterized by fusion of the ribs and spinal deformities or truncations[8]. The segmental border of a somite is defined under the influence of Mesp2, a transcription factor that acts by suppressing the Notch signaling pathway [9], involved in the pace of somite formation. Each somite in the thoracic region

forms the caudal part of one rib and the cranial part of the next caudal rib. Finally, after their formation from unsegmented somatic mesoderm (PSM). Occurrence of congenital skeletal malformation in patients with mutations in Notch-associated genes which exhibit a short trunk due to multiple hemi vertebrae formation accompanied by rib fusions, bifurcations, and deletions. The dermomyotome derived from one somite contributes to the intercostal muscle [10]. Aberrations of ribs probably occur during the process of segmentation and re segmentation of the developing of costal processes from the somites [2]. Mesp 2 also plays a crucial role in defining these regions, by establishing the gene expression pattern within the two halves [9]. The formation of a rib follows a complex pattern that involves the caudal half and the rostral half of two adjacent somites, in children, the bifid and fused ribs are associated with pathologic malformations such as Gorlin-Goltz syndrome [3], which is a rare, autosomal dominant inherited condition. Rib anomalies are generally more common in females than in males, occur more frequently on the right side and usually they are asymptomatic. Studies reported that genetic background influence negative impact on rib development in children, such as trisomy 18, neurofibromatosis, achondroplasia, thalassemia major and mucopolysaccharidosis can increase the relative risk of ribs malformations. Progressive loss of bone matrix and demineralization occurs in case of overactive (hyper) parathyroid gland and vitamin D deficiency, also known as rickets happens when the there is hepato renal dysfunction. Prostaglandins medication, used to treat heart disease in children can also cause abnormal rib development; Additional intercostal space in our case gets nerve supply from 4th intercostal nerve. This indicates that the muscles of this additional space are originated from muscles of the fourth intercostal space. Development of the bifid rib is probably due to disrupted interactions between cephalic and caudal segments of sclerotome during embryogenesis, occurring around the 4th-6th week of fetal life[11].

Conclusion

Skeletal variations in the anterior chest wall may indicate underlying systemic disease. Mal-development of rib at upper end of thoracic cage is due to defective bone segmentation and also associated with variation in disposition of vessels and nerves. Identification of rib anomalies can yield important diagnostic clues in the work-up of patients with congenital bone dysplasia, acquired metabolic diseases, iatrogenic conditions, trauma, infection, and neoplasm's. Awareness of such variations is necessary for physician's radiologist and surgeons during clinical diagnosis and treatment.

Ethical Clearance- Not Taken (for Cadaveric study Ethical clearance is not applicable)

Source of Funding- Self

Conflict of Interest - Nil

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