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A Rare Presentation of Grossly Deformed Lungs in a Kyphoscoliotic Female Cadaver

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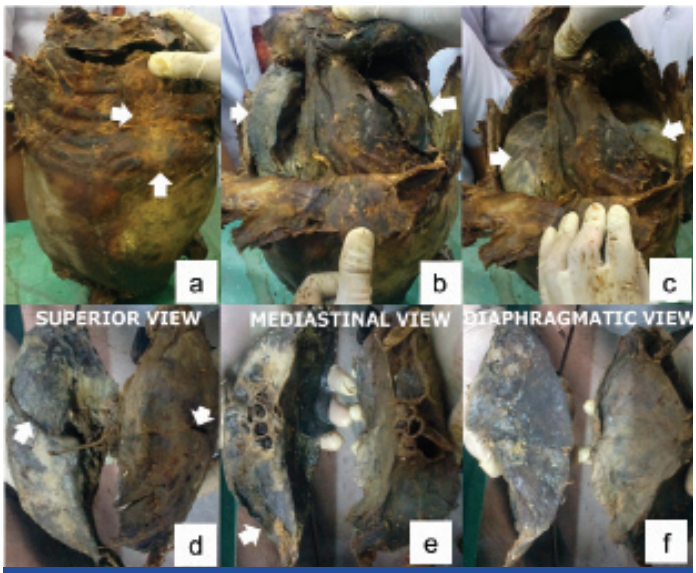
A corpse of around 50-year-old female destitute was received for dissection for undergraduate MBBS and BDS classes in a government medical college. The cadaver had kyphoscoliosis with anterior chest wall deformity and had died from cerebrovascular accident in the government hospital. There were no other medical records available. The length of the body was around 140 cm. This body was preserved in an embalming fluid containing 10% formalin solution and was dissected for undergraduate studies.

In [Table/Fig-1a] concavity on the sternum (horizontal arrowhead), in [Table/Fig-1b] left and right lungs (arrowheads), in [Table/Fig-1c] right and left diaphragms, in [Table/Fig-1d], oblique fissures (arrowheads), in [Table/Fig-1e] sharp anterior border of right lungs (arrowhead), in [Table/Fig-1f] the diaphragmatic surface of the lungs, are shown.

The torso was unusually small with visible deformities of the anterior chest wall [Table/Fig-1a], arrowheads. There was concavity [Table/Fig-1a], horizontal arrowhead) in the sternum and hence the deformity was identified as pectus excavatum. When the sternum and anterior part of the rib cage were opened the unusual appearance of the lungs first came into view. The lungs were quite small. We took out both the lungs and took photographs both before [Table/Fig-1b] and after removing the lungs [Table/Fig-1c]. On examination, the lungs' antero-posterior dimensions exceeded the vertical dimensions. The left lung's maximum antero-posterior length was 18 cm and that of right lung was 20 cm respectively. The maximum dimensions in vertical axis as measured in the mediastinal surface were 8 cm and 7 cm for left and right lungs respectively. [Table/Fig-2] compares the dimensions with that of the average lungs dimensions of adult females. The viscera have undergone considerable compression in vertical axis, leading to rounded appearance of the apex. The fissures demarcating the lobes were clearly visible [Table/Fig-1d], arrowheads and were complete. The right lung had two lobes instead of the usual three. Both the lungs had ill-defined apex, sharp anterior border, rounded posterior border and concave inferior or diaphragmatic surface [Table/Fig-1f]. The left lung had two lobes and cardiac notch. On the mediastinal surfaces [Table/Fig-1e] the impressions were difficult to identify. The hilar structures were distorted, but were preserved. We could not find any other gross variations in this cadaver.



[Table/Fig-1]: a) Concavity on the sternum (horizontal arrowhead); b) Left and right lungs (arrowheads); c) Right and left diaphragms; d) Arrow heads showing oblique fissures; e) Sharp anterior border of right lungs (arrowhead); f) The diaphragmatic surface of the lungs are shown.

Presence of normal fissure and normal anatomy in the lungs increases chances of uniform expansion, help as good landmarks and thereby help in proper planning of surgery [1]. There were two lobes instead of three lobes in the right lung. Any deviation in branching morphogenesis may be associated with vascular anomalies as Vascular Endothelial Growth Factor (VEGF) signalling from the developing lung epithelium leads to proper vascularization of lungs during organogenesis of lungs [2].

Embryology of lungs: The number of lobes is dependent upon formation of fissures during intrauterine development. In the fourth week of intrauterine life the endodermal lung diverticulum starts from the ventral part of primitive pharynx. It then separates from the foregut tube and divides into right and left primary bronchial buds. The right primary bronchial bud divides into three secondary bronchial buds and the left divides into two secondary bronchial buds. By further division the branching continues to form bronchopulmonary buds that fuse with each other except at certain places. These places form the fissures. Unusual fusion among bronchopulmonary buds may lead to decreased or increased number of fissures thereby decreasing or increasing the number of lobes [3].

Parameters	Left (Average) (5)	Left (In this specimen)	Right (Average)(5)	Right (In this specimen)
Height	20.3 cm	8 cm	19.3 cm	7 cm
Width	9.7cm	9 cm	11.2 cm	8 cm
Depth	16.2 cm	18 cm	15.9 cm	20 cm

[Table/Fig-2]: Comparison of linear dimensions of lungs in adult females.

Various molecular factors are responsible for regulation of individual events in lung development. The interaction between sonic hedgehog (shh) and the Gli transcriptional factor family leads to lung bud development and separation from the primitive gut tube [4].

Branching of primary bronchus determines the lobes of the lungs. Both shh and Gli transcription factors can be considered as the factors influencing lobe formation in lungs. This is further supported by murine model with Gli2 –/– that leads to defective primary branching in right lungs [2]. Details of molecular regulation of lung development is quite complicated, hence is beyond the scope of this article.

Studies on normal dimensions of lungs in Indian population are difficult to find. We could only find a Canadian study with linear dimensions of lungs calculated from CT images. In [Table/Fig-2] data for adult females is given [5]. Linear dimension data of CT images and that of cadaveric data is bound to differ, but the difference in this case is much higher.

We determine and demonstrate side of a lung that has been taken out of the body by relying on the anatomical features like, sharp anterior border, rounded posterior border and concave diaphragmatic surface. In spite of gross degree of compression the lungs maintained these features, hence traditional teaching for side determination of lungs held true even in such a deformed specimen. Counting lobes for determining side of lungs is considered fallacious and the same was demonstrated in the right lungs that had only two lobes instead of the usual three lobes.

The degree of compression found in the patient is quite rare. The patient was a destitute without a full medical report, so the cause as well as the effects of the malformed lungs, if any is only a subject of speculation.

Anterior chest wall deformities like pectus excavatum is the most common chest wall deformities associated with adults with congenital heart diseases. It is associated mostly with Mitral Valve Prolapse (MVP) [6]. Though the patient had pectus excavatum, but we were unable to corroborate MVP as it was neither in the clinical history nor we were able to identify it in the dissected heart.

Kyphoscoliosis being a restrictive lung disease is a risk factor for development of pulmonary hypertension and cor pulmonale. This leads to increased workload of right ventricle leading to right ventricular hypertrophy. Right heart failure with peripheral oedema more specifically ankle oedema is an important feature of cor pulmonale [7]. Death of the patient was due to cerebrovascular accident and there was no further medical history available, but considering the gross degree of kyphoscoliosis, it is quite probable that the patient was suffering from pulmonary hypertension.

Pectus excavatum with kyphoscoliosis and short stature were present in this cadaver that narrows the diagnosis towards Noonan syndrome. Noonan syndrome is mostly diagnosed clinically by looking up the following features. The patients have facial features like low-set ears, ptosis and hypertelorism. They also have short stature and chest and spinal deformities. There are presence of congenital heart defects like atrial septal defect and pulmonary valve stenosis [8].

The compression of the lungs might have resulted from the thoracic wall deformity, but the presence of two lobes on the right side indicates an intrauterine anomaly. The presence of different abnormalities with different aetiologies indicates the case to be a syndrome, so a differential diagnosis of Noonan syndrome can be proposed.

The variations like, compressed lungs and presence of only oblique fissures in both the lungs such as in this case distorts the landmarks and increase the challenge for the surgeon to operate. Awareness about these variations will definitely be of help to the thoracic surgeons and radiologists.

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