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Case Series

Hunchback with "S" Shape Spine: Cadaveric Case Series

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ABSTRACT

Introduction: Kyphosis is a sharp posterior angulation due to localized collapse or wedging of one or more vertebrae and scoliosis is the lateral curvature of the spine. These may occur due to a congenital defect, fracture, and sometimes pathological or spinal tuberculosis.

Materials and Methods: During routine cadaver dissection, four cases of kyphoscoliosis were identified. In the first case, a protrusion on the posterior surface of the trunk of the cadaver at L1 level was observed in a 60-year-old male. The lungs were collapsed and adherent with the pleura and thoracic wall. The posterior wall of the stomach was adherent to the pancreas and a depression was noted on the anterior surface of abdominal aorta. The T12 and L1 vertebrae were fused, and the spine was deviated to the left side. The anterior aspect of the body of L1 vertebrae was marginally eroded. The second case of kyphoscoliosis was from 80-year-old female cadaver. The descending and abdominal aorta had an abnormal course along the vertebral column, third and fourth cases were observed with slight protrusion and scoliosis but not much changes in the vertebral column.

Results: The cause of the deformity was confirmed by pathological examination of the tissues showed inter vertebral disc degeneration with calcification and ossification and osteopenia. The underlying etiology can be tuberculosis, injuries, or infections.

Conclusion: Severe kyphoscoliosis can affect wide range of systems in the body. It will affect not only the respiratory system also pressure on great vessels as a consequence of bony deformity.

KEY WORDS: Kyphosis, Cadaver Dissection, Kyphoscoliosis, Deformity, Calcification, Ossification.

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INTRODUCTION

The scoliosis is a lateral curvature of the spine [1]. It is congenital in 5% patients and can occur due to defects in the formation of vertebral bodies [2]. In kyphosis, there is a sharp posterior angulation due to localized collapse or necrosis of one or more vertebrae [1]. Some of the pathological bases of vertebral collapse are tuberculosis, ankylosing spondylitis, osteoporosis, and metastases [2]. The bulge on the back is the striking feature of patient of kyphosis. The condition is usually of such a slight degree that it does not affect the patient [3].There are reports of kyphoscoliosis in patients with Goldenhar syndrome, Klippel-Trenaunay syndrome, Cohen syndrome, and neurofibromatosis [4-7].

The Goldenhar syndrome can be called as OCULO-AURICULO-VERTEBRAL DYSPLASIA, it's an embryological defect of the derivatives of 1st and 2nd pharyngeal arches. So the ear, mandible and vertebrae are developing in abnormal way. This was explained by Dr. Maurice Goldenhar in 1952 [8]. The incidence of Goldenhar syndrome will occur in 1:35, 000–1:56,000 population, ratio of male to female gender 3:2.2 [9]. Neural crest cells and chromosomal defects, the habituation of alcohol by mother during pregnancy and also the environmental factors like, cocaine and retinoic acid are responsible [10].

4% of people in the population gets affected by the musculoskeletal defects is called Scoliosis. The vertebral column in scoliosis people shows a disproportionate lateral curvature of the vertebral column [11]. It's a neuromuscular, congenital or idiopathic disorder. The neuromuscular scoliosis will occur in the lesion of upper (or) lower motor neurons and in muscular dystrophy [12].

The abnormal segmentation of embryonic sclerotome masses [13] and their rotational displacement [12, 14, 15, 16] leads to the changes in the vertebral column. It may occur in early stage of embryonic life and forms the congenital scoliosis in 1:1000 [17].

In congenital scoliosis the lateral curvature can happen due to the hemivertebra or the unsegmental vertebral bars [18,19]. Here, we report four cases of kyphoscoliosis encountered during routine cadaveric dissection.

OBSERVATIONS

During routine anatomical dissection in the department of anatomy at Pondicherry Institute of Medical Sciences, gross morphological anomalies were observed in two cadavers, 60-year-old male (case 1) and 80-year-old female (case 2) in third (65 year male) and fourth (70 year male) cases showed small protrusion. In first and second cases had the posterior aspect of the trunk was protruded at the level of L1 vertebra (Fig.1), and the spine was deviated to the left side (Fig.4). Further gross dissection along with the radiological analysis was done. The bone from affected area and lung tissues were collected for histopathology.

CASE 1:

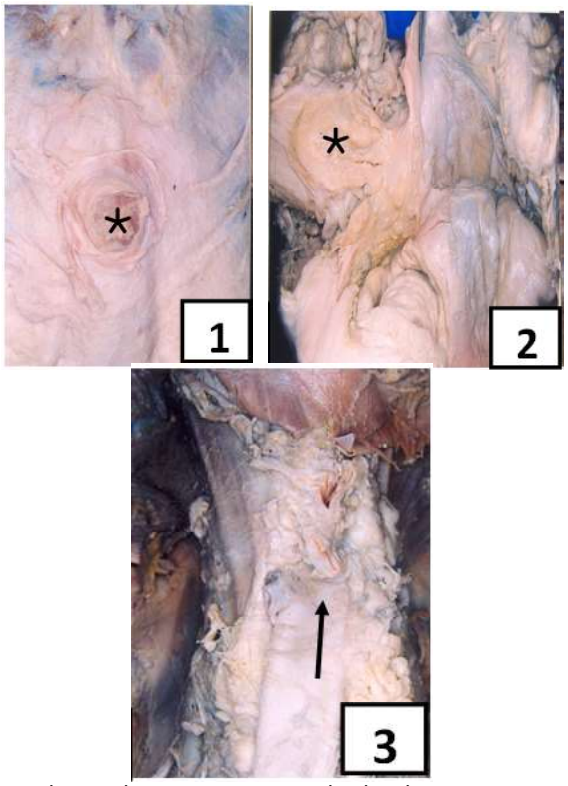

Fig.1: Shows the protrusion on the lumbar spine at L1.
Fig.2: The adherence of posterior wall of stomach with head and neck of pancreas.
Fig.3: Depression on the abdominal aorta at T12 and L1 vertebra level.

Gross Anatomical Finding: In the thoracic cavity, heart was normal in morphology. The lungs were collapsed, and adherent to diaphragm as well as thoracic wall. The posterior wall of stomach was adherent to the

head and neck of pancreas (Fig.2). As the pancreas was reflected upwards, a depression was noted on the anterior aspect of abdominal aorta, at the level of T12 and L1 vertebrae (Fig.3), due to a tumour growth from stomach and pancreas. Also, indentation on the body of the lumbar vertebrae was observed. There was no evident compression of the spinal meninges in the vertebral canal.

Radiological Finding: The spinal angulation on the left side with scoliosis was seen. The thoracic vertebrae showed osteoporotic changes. In the lateral view of radiograph, the T12 and L1 vertebrae were fused (Fig.5) and the body of L1 vertebrae had erosion (Fig.4) on its anterior surface. The posterior border of spine had a smooth appearance.

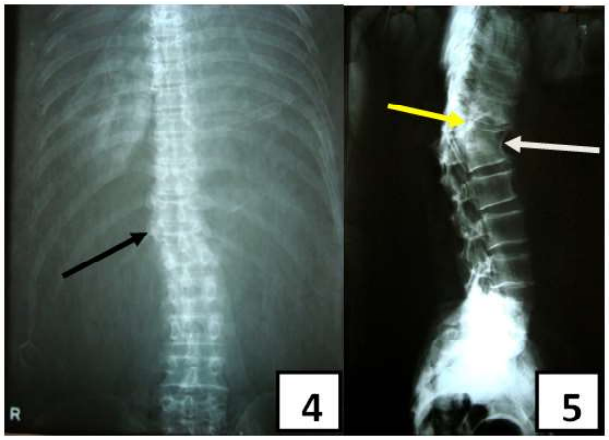

Fig. 4: Antero - posterior view of plane X-ray shows deviation of vertebral column at L1vertebra.
Fig. 5: White arrow shows the concavity on the anterior surface of body of L1 vertebrae and Yellow arrow shows the fusion of T12 and L1 vertebral spine.

Pathological Finding: A section of bone from T12 and L1 exhibited chronic osteomyelitis (Fig.6) with bone formation.

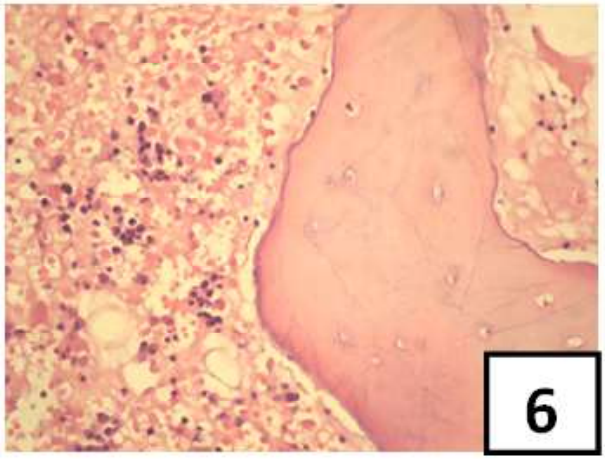

Fig.6: Section of bone tissue in Haematoxylin&Eosin stain shows (40x) chronic osteomyelitis.

CASE 2:

Gross Anatomical Finding: The descending thoracic and abdominal aorta had an abnormal course in relation with vertebral column (Fig.8&9). In the posterior mediastinum, the descending thoracic aorta made an "S" curve on the right-side vertebral column than the usual because of kyphosis. The abdominal aorta had a forward convexity and shifted the course to left side of vertebral column than the usual course due to scoliosis (Fig.8&9).

Radiological finding: The thoracic and lumbar vertebra showed kyphoscoliosis.

Pathological Finding: The section of lung tissue was autolyzed and bone tissue from thoracic and lumbar level indicated osteopenia.

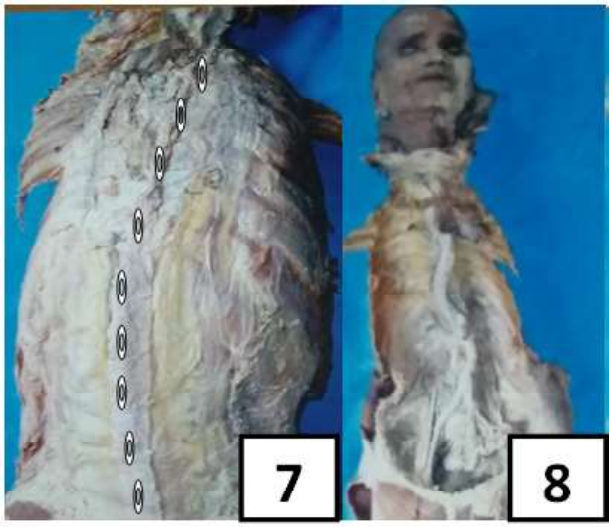

Fig.7: Shows the S - shape curve of vertebral spine.
Fig. 8 & 9: Shows the S - shape thoraco – andominal aorta.

Case three and four:

The third and fourth cases were observed with slight protrusion and scoliosis but not many changes in the vertebral column.

DISCUSSION

The normal thoracic curvature is concaving forwards or kyphotic. The prolonged bad posture or multiple pathological changes will amplify this curvature that appears as a deformity [2]. These could be the result of a congenital defect, fracture and sometimes pathological or spinal tuberculosis. The most common etiology of kyphoscoliosis is idiopathic [3].

The scoliosis can be structural when a curved spine is rotated as well. This is seen usually in adolescents, muscle dystrophies or neural tube defects like spina bifida. A limb length discrepancy or hip deformities cause pelvis to slant laterally causing compensatory scoliosis. A protective scoliosis is due to action of muscles attached to the spine [2].

The vertebrae form from sclerotome of somites. The abnormal fusion of adjacent sclerotomes will form fused or block vertebrae. Also, hemivertebrae is formed if vertebral body has more than one ossification centers out of which only one forms the bone. These developmental anomalies like hemivertebrae or block vertebrae can cause spine to deviate laterally [2]. In our report, two vertebrae in thoracolumbar spine were fused together (Fig.5) as this is a cadaver with unknown history, it could not be determined if this was congenital.

The kyphosis can infrequently occur due to fracture of surgically fused lumbar spine. Calderaro et al. reported a case of 16-year-old female with surgically corrected scoliosis. The patient suffered a traumatic vertebral fracture due to fall from stairs and subsequently had a thoracolumbar kyphoscoliosis [20].

Stevenson et al. dissected a cadaver with severe scoliosis. The spine was deviated to the right side and the muscles attached to spine were wasted on the left side. The bones wereosteoporotic and the cranial dissection revealed a Chiari type - 1 malformation [21].

The spine is the most common site of skeletal tuberculosis. The drug abuse, prolonged corticosteroid medication and AIDS lead to reduction in body defense mechanisms. Mycobacterium tuberculosis passes through the lung or the gut by swallowing infected milk products or through skin. Initially the bacilli are fixed in the nodes and no clinical illness is seen. But occasionally the response is excessive with enlargement of glands in the neck or abdomen. Within the nodes which apparently healed calcified bacilli may survive for many years. The body has been sensitized to the toxin and re infection occurs. The response is quite different, the lesion being a destructive one which spread by contiguity.

If resistance to the original infection is low, infection may spread via the blood stream and giving rise to miliary tuberculosis or meningitis and more often blood spread occurs months (or) years later and bacilli get deposited in extra pulmonary tissues. There is a predilection for the vertebral bodies and the larger synovial joints. Once the bacilli have gained a foothold they elicit a chronic inflammatory reaction.

The bone lesions tend to spread quite rapidly only in the vertebral bodies and more rarely in the greater trochanter of the femur or the small bones of the hands or feet and the infection persist as a pure osteomyelitis. If articular cartilage is damaged healing occurs by fibrosis and incomplete ankylosis with progressive joint deformity.

Blood borne infection usually settles in a vertebral body adjacent to the intervertebral disc. Bone destruction and caseation follow with infection spreading to the disc space and to the adjacent vertebrae. As the vertebral bodies collapse into each other, a sharp angulation or kyphos develops. Caseation and cold abscess formation may extend to neighboring vertebrae or escape into the paravertebral soft tissues [1]. The kyphoscoliosis in thoracolumbar region can be associated with craniofacial microsomia in Goldenhar syndrome. The simultaneous development of the first two branchial arches and vertebrae during early embryonic life can cause both facial defects and scoliosis [4,22]. Another

syndrome related to kyphoscoliosis though rarely is Klippel-Trenaunay. It is a congenital syndrome with varicose veins, capillary hemangioma, and limb enlargement [4].

In a congenital syndrome such as Cohen syndrome, kyphoscoliosis is not present at birth. It gradually appears and progresses in adult life due to poor muscle tone [20].

Also, around 10% cases with neurofibromatosis type 1 have spine deformities like kyphoscoliosis [7].

Though mostly mild, severe kyphoscoliosis can affect wide range of systems in the body. In the respiratory system structure of alveoli and blood vessels is adversely disrupted along with respiratory capacity. The right ventricular hypertrophy due to pulmonary hypertension and pressure on great vessels as a consequence of bony deformity can also occur [3] In this case, the aorta was deviated from usual course in thorax and abdomen due to vertebral curvature.

The spine and chest wall distortion will also limit the respiratory movements and thence the volume of thoracic cavity. If severe, this is likely to affect lung expansion and eventually leading to pulmonary hypertension [23].

The severe kyphoscoliosis can deleteriously affect the daily life activities of a patient[10]. The diagnosis and timely intervention for severe disease is vital for surgeons to effectively manage the kyphoscoliosis cases.

In the present case of Goldenhar syndrome, the scoliotic deformity was progressive but Cobb angle was not severe. Furthermore, the deformity was not congenital and no hemivertebra or vertebral abnormalities were detected. Thus, brace treatment was prescribed to prevent the advancement of scoliosis [24].

CONCLUSION

Severe kyphoscoliosis can affect wide range of systems in the body. It will affect not only the respiratory system also pressure on great vessels as a consequence of bony deformity. So the postural and pathological curvature of spine and vertebrae's of vertebral column can be diagnosed at the earliest with proper treatment to help to restore the normal structure.

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Conflicts of Interests: None

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