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Case Report

Isolated sternal hypoplasia: a rare cause of kyphoscoliosis

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ABSTRACT

In most cases, kyphoscoliosis is idiopathic. However, this is a diagnosis of exclusion and can be made only if no cause can be identified. Kyphoscoliosis can occur due to various causes. Isolated sternal anomalies may also cause kyphoscoliosis secondary to the bony deformity though this has not previously been reported in literature. We have reported a case of kyphoscoliosis secondary to isolated sternal hypoplasia with complete absence of bony and cartilaginous elements of the body and xiphoid process of the sternum without any associated deformities of mediastinal structures, lung parenchyma or soft tissues in a young male patient. Careful evaluation of patients with kyphoscoliosis can ensure timely diagnosis of unusual and potentially treatable causes for the same such as sternal anomalies. Addition of lateral chest radiographs to the imaging protocol for evaluation of kyphoscoliosis can play a major role in timely diagnosis of such cases.

Keywords: Isolated sternal hypoplasia, Kyphoscoliosis, Sternal anomalies, Sternal hypoplasia

INTRODUCTION

Kyphoscoliosis refers to an abnormal curvature of the spine in the coronal and sagittal planes. Idiopathic kyphoscoliosis constitutes majority of the cases and is essentially a diagnosis of exclusion.[1] Only when no cause for kyphoscoliosis is identified after complete evaluation can it be termed idiopathic. Various other causes for kyphoscoliosis include infections, degenerative changes, neuromuscular disorders and a variety of syndromes. In addition, some unusual causes of kyphoscoliosis have also been identified.

These causes of kyphoscoliosis are sometimes clinically apparent but, in many cases, they are not and are not suspected until detailed radiological evaluation of the patient is done. Congenital sternal anomalies such as sternal agenesis, sternal hypoplasia and cleft sternum are usually seen as part of syndromes and have been very rarely reported in isolation.[2] Isolated sternal anomalies may also cause kyphoscoliosis secondary to the bony deformity although this has not previously been reported in literature. Here authors report an unusual case of kyphoscoliosis secondary to isolated sternal hypoplasia in an adolescent male patient.

CASE REPORT

A 16-year-old male presented with complaints of progressive kyphoscoliosis over the past 6 years, predominantly involving the dorsal spine, which was accelerated during puberty. He had some physical disability due to the kyphoscoliosis leading to mild functional limitation. Slight reduction of respiratory reserve secondary to the spinal deformity was documented on pulmonary function tests. Neurological examination was normal. There was no clinical evidence of nerve root or spinal cord compression. Intellect and mental functions were normal.

He underwent radiographs of the dorsolumbar spine (anteroposterior and lateral views) for the assessment of the cause and extent of bony anomalies which showed kyphoscoliosis with convexity to the right extending from the fourth dorsal to the third lumbar vertebrae. Associated lateral wedging with osteopenia and reduction in the vertical height of the vertebrae of the fifth to eighth dorsal vertebral bodies within the scoliotic curvature was observed. The ribs also showed crowding with variable intercostal spacing. Additionally, in the anteroposterior view of the dorsal spine, the lower trachea, bronchi and dorsal spine were unduly prominent (Figure 1).

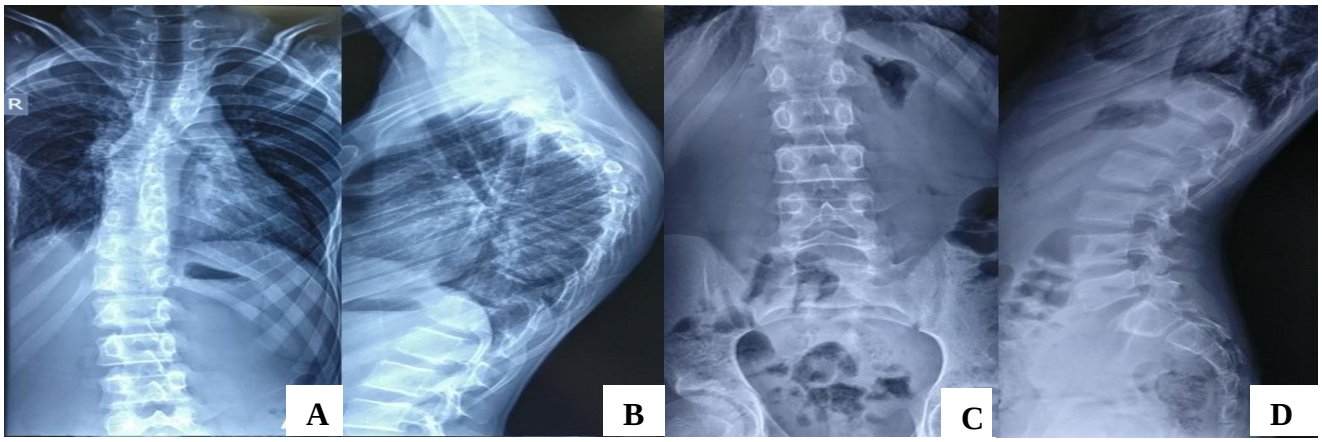

Figure 1: Dorsolumbar spine radiographs of the patient. (A): Dorsal spine AP view and (B): Dorsal spine lateral view show kyphoscoliosis with reduced vertical height and osteopenia of the involved vertebral bodies. The lower trachea, bronchi and dorsal vertebral bodies appear unduly prominent. (C): Lumbosacral spine AP view and (D): Lumbosacral spine lateral view show compensatory mildly increased lumbar lordosis.

In view of this finding, chest radiograph anteroposterior and lateral views were performed which additionally revealed the clavicles to be more obliquely and inferomedially directed than usual.

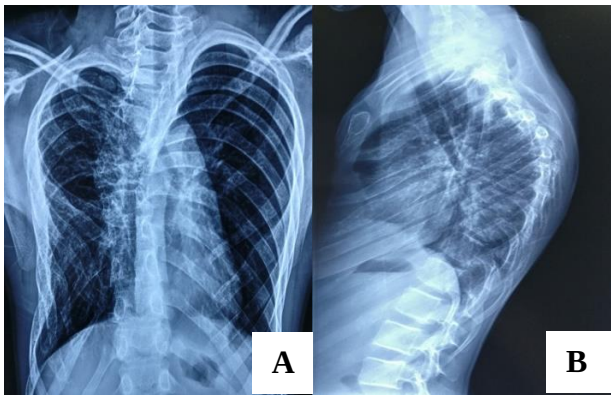

Figure 2: Chest radiographs of the patient. (A): PA view shows obliquely directed clavicles with crowded and variably spaced ribs. (B): Lateral view shows non-visualization of sternum below the level of the manubrium.

A careful assessment of the lateral radiograph then revealed complete absence of the body and xiphoid process of the sternum below the level of the visualized normal manubrium (Figure 2). Further evaluation was done with Non-Contrast Computed Tomography (NCCT) scan of the thorax using 6 slice SIEMENS multidetector computed tomography (MDCT) scanner for accurate assessment of bony anomalies and assessment of any associated intrathoracic abnormalities of the lungs, heart and mediastinum. The CT scan of the thorax allowed confirmation and detailed assessment of the osseous findings on the radiograph. The internal organs of the thorax and mediastinum as well as the lung parenchyma were normal (Figure 3). Magnetic resonance imaging (MRI) scan of the thorax using 1.5 Tesla Philips achieve MRI scanner was performed for cartilage and soft tissue assessment. In addition to corroboration of the findings on the CT scan, MRI of the thorax additionally revealed complete absence of any cartilaginous component of the sternum below the level of the manubrium. No significant abnormality could be identified in the other soft tissues of the chest wall. Spinal cord was also normal in the dorsal region (Figure 4). A complete evaluation of the patient was then done for any associated abnormalities. A skeletal survey was performed in order to look for any skeletal dysmorphisms and none were identified. Echocardiography was done for cardiac assessment and no abnormality was found. Ultrasound examination of the abdomen was performed for assessment of the abdominal viscera which turned out to be normal. Karyotyping was done for chromosomal anomalies which showed a normal pattern of chromosomes. Based on the clinical history, imaging findings and other investigations performed, a

final diagnosis of kyphoscoliosis secondary to isolated sternal hypoplasia was made. The patient was worked up for surgical correction of the deformity, but in view of the risks involved and the expected benefits of the surgical procedure at the time of diagnosis, the risk/benefit ratio was not expedient for surgical correction and the patient did not undergo surgery.

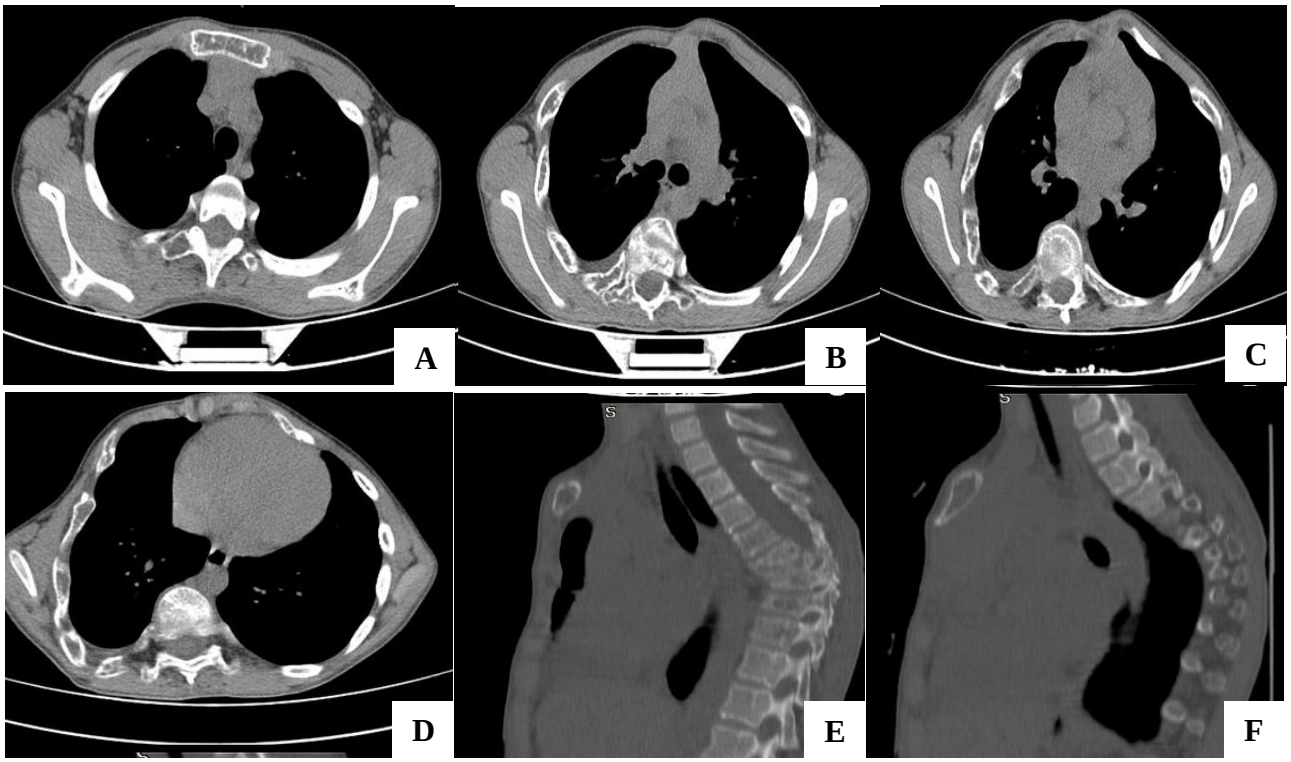

Figure 3: NCCT thorax of the patient. Axial scans at level of (A): aortic arch branches, (B): subcarinal region, (C): upper heart, (D): lower heart, and sagittal reconstructions at the level of (E): midline and (F): paramedian region show normal manubrium with complete absence of the body and xiphoid process of the sternum with kyphosis. Mediastinal structures appear normal.

DISCUSSION

The known causes of kyphoscoliosis can be classified broadly into congenital, neuromuscular, syndrome-related, and secondary causes, although in most cases a definite cause cannot be identified, and it is termed idiopathic. Congenital kyphoscoliosis includes those cases due to identifiable vertebral abnormalities such as hemivertebrae, butterfly vertebrae and unsegmented vertebral bar, causing mechanical deviation of the spinal curvature.

Neurological causes include cerebral palsy, nerve injuries and as sequelae to paralysis. Muscular abnormalities such as muscular dystrophies including Duchenne and Becker muscular dystrophies and facioscapulohumeral muscular dystrophy can sometimes cause kyphoscoliosis. Various syndromes such as Marfan syndrome, Ehlers-Danlos syndrome, Escobar syndrome, cleidocranial dysplasia, spondyloepiphyseal dysplasia and neurofibromatosis are also associated with kyphoscoliosis. In addition, kyphoscoliosis can also be secondary to other potentially correctable causes such pain, spinal cord abnormalities, tumors (both intraspinal and extraspinal) and infections.[3] Though uncommon, kyphoscoliosis can also be secondary to congenital sternal anomalies.[1] Significant deformity in such cases would be expected not at birth or in infancy, but rather during the period of rapid sternal and vertebral growth (particularly around puberty), as is seen in our case.

Congenital sternal anomalies are rare deformities and are frequently associated with other chest wall anomalies such as pectus carinatum. They can also be associated with various syndromes. Syndromic associations of congenital sternal anomalies include trisomy 7, 9, 12p or 18, pentalogy of cantrell, moebius syndrome, turner syndrome and noonan syndrome.[2]

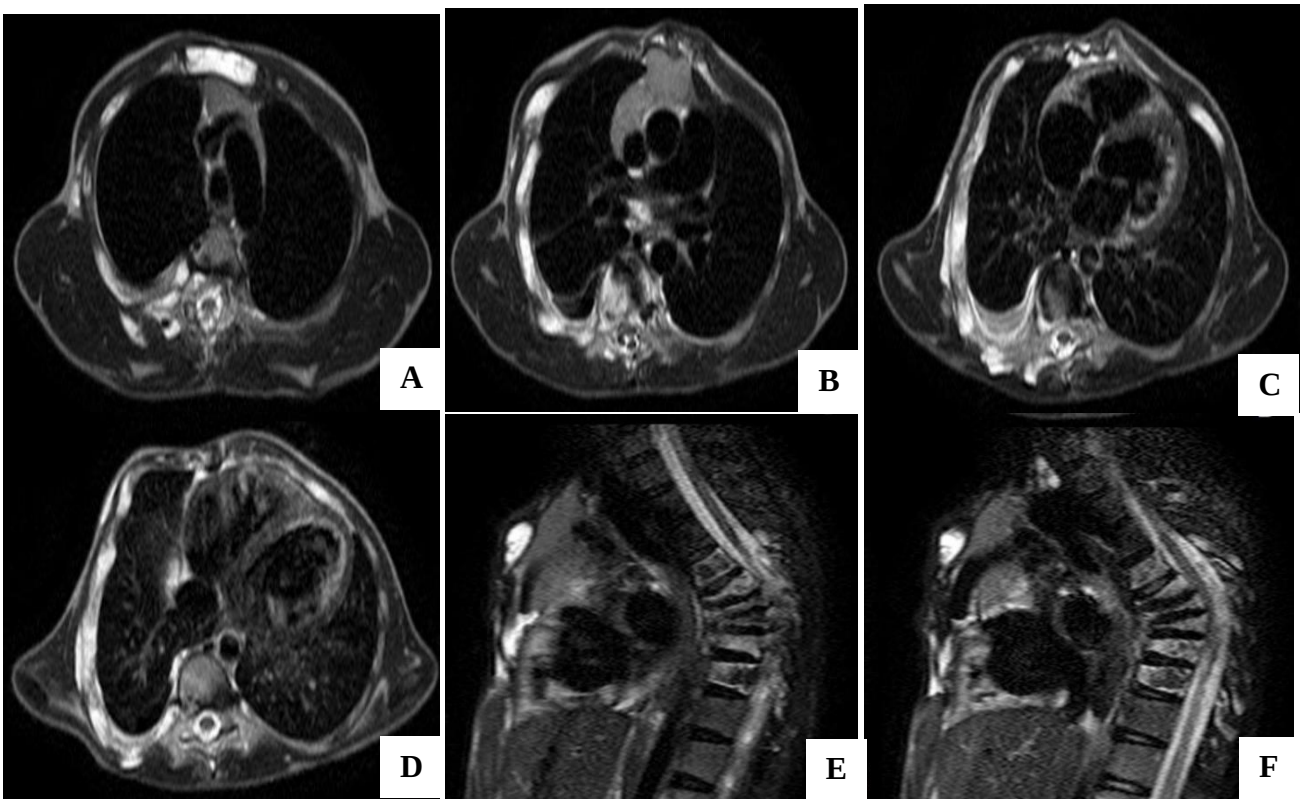

Figure 4: MRI thorax of the patient. T2W axial scans at the level of (A): aortic arch, (B): pulmonary trunk, (C): upper heart, (D): lower heart, and sagittal scans at the level of (E): paramedian region and (F): midline show normal manubrium with complete absence of any bony or cartilaginous component of body and xiphoid process of the sternum with kyphosis. Rest of the visualized soft tissues and spinal cord appear normal.

Isolated sternal anomalies such as sternal hypoplasia, sternal agenesis and cleft sternum are much rarer than syndromic sternal anomalies or sternal anomalies associated with other chest wall anomalies. If diagnosed early, these isolated anomalies are potentially correctable and have a better prognosis than if they are associated with other deformities.[2,4] In such isolated cases however, the etiology is not well understood. Various possible causes have been proposed based on animal models such as alcohol exposure, riboflavin and methyl cobalamin deficiency as well as genetic factors such as HOX4 gene disruption. But no definite associations have been reported in humans to date.[2,5]

The embryological development of the sternum is well known. The sternum develops from a pair of sternal bars which are longitudinal mesenchymal condensations that form in the ventrolateral body wall, i.e. in the somatic layer of the lateral plate mesoderm. As the growing cranial ribs meet them in the seventh week, the sternal bars meet along the midline and undergo progressive fusion, commencing at the cranial end and progressing caudally. Formation of the xiphoid process occurs in the ninth week which completes the embryological development of the sternum. This process ultimately forms the cartilaginous sternum. Ossification then occurs from six centers – one in the manubrium, four in the body of the sternum and one in the xiphoid. Sternal ossification centers appear before birth with the exception of the ossification center of the xiphoid process, which appears postnatally. Failure of fusion of the lateral sternal bands or an early intrauterine disturbance affecting the mesodermal structures of the midline between the sixth and ninth gestational week is a possible mechanism to explain the occurrence of sternal anomalies.[6]

In a case of dorsolumbar kyphoscoliosis, assessment of the deformity essentially relies on plain anteroposterior and lateral radiographs of the dorsolumbar spine for assessment of type and extent of deformity and measurement of various angles necessary for surgical correction. In cases where assessment of the sternum is to be done, lateral and oblique views of the sternum are necessary. The sternum is often difficult to assess and characterize completely on X-rays due to overlap of bony elements and the complex bony anatomy of the thoracic wall. Multiplanar and three-dimensional reconstructed MDCT images are useful in the evaluation of the

sternum. MDCT images are also useful for assessment of associated anomalies of the mediastinum, heart and lungs. MRI is usually not performed in a case of scoliosis unless there is an abnormality on neurological examination, when MRI will be necessary for evaluation of the spinal cord and nerve roots. Occasionally, when there is non-visualization of any of the bones in X-rays or MDCT as in our case, MRI will have to be performed for identification and characterization of cartilaginous components of the bone.[1,6]

Turturro F et al, reported a case of isolated asymptomatic short sternum in a healthy 13-year-old girl without any spinal deformities. She was diagnosed to have a hypoplastic sternum with non-visualization of the distal half of the body of the sternum. The manubrium was normal. It was the first case reported in literature of asymptomatic isolated sternal hypoplasia. Echocardiography had ruled out cardiac anomalies in the patient. No treatment was necessary as she was asymptomatic.[7] Another paleontological case report of relevance is the anomaly of Santa Rosa de Viterbo, an Italian mummy. A careful study of the well-preserved body had revealed isolated total agenesis of the sternum without any other identifiable abnormality.[8]

In our case of progressive kyphoscoliosis, the cause was found to be isolated sternal hypoplasia with complete absence of body and xiphoid process of the sternum which was not associated with any other anomalies of the chest wall, heart and great vessels, lung parenchyma, other mediastinal structures or soft tissues of the chest wall. To our knowledge, there been have no similar cases reported in literature to date. The exact cause of such isolated sternal anomalies, their functional implications as well as the rationale behind why they predispose to kyphoscoliosis only in some cases and not in others remains obscure. Further studies are necessary in order to answer these questions.

CONCLUSION

Kyphoscoliosis can be due to a variety of causes although most cases are idiopathic. Every effort should be made to identify any underlying cause early as many such causes may potentially be correctable if diagnosed early. Congenital sternal anomalies can be a cause of kyphoscoliosis in rare cases and when these occur as isolated anomalies, the diagnosis can be easily missed on the routine radiographs of the dorsolumbar spine due to incomplete visualization and the complex bony anatomy of the thoracic cage. This underlies the importance of careful assessment of patients with kyphoscoliosis for the timely diagnosis of unusual and potentially treatable causes for the same such as sternal anomalies. The addition of lateral chest radiographs to the imaging protocol for evaluation of kyphoscoliosis can play a major role in the timely diagnosis of such sternal anomalies as the cause of kyphoscoliosis.

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