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A Case Report on the Surgical Treatment of Scoliosis and Kyphosis Caused by Congenital Multiple Butterfly Vertebrae

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Abstract: ***Case: A 14-year-old adolescent patient presented with recurrent chest and lower back pain. Over a year ago, a kyphosis and a scoliotic deformity resembling a knife-back were detected at the thoracolumbar junction. Further examination revealed a diagnosis of hemivertebrae formation in the thoracic vertebrae, scoliosis, and kyphosis in the thoracolumbar region, caused by a rare developmental anomaly of multiple butterfly vertebrae. After confirmation of the diagnosis, the patient underwent surgical treatment, and the condition was effectively alleviated.***

Keywords: Butterfly vertebrae, Spine, Scoliosis, Deformity.

Abbreviations: BV=Butterfly Vertebrae

1. Introduction

Butterfly vertebra is a rare congenital anomaly, and due to its similarity to other diseases, it is easily misdiagnosed [1]. In literature, it is usually reported as an isolated finding. Therefore, multiple butterfly vertebrae occurring in the lower thoracic region is an extremely rare congenital spinal developmental deformity that may lead to kyphosis or scoliosis. Butterfly vertebrae is a deformity characterized by congenital failure of the vertebral body cartilage center fusion [2], it consists of two parts of the vertebral body separated by a sagittal cartilage septum or completely separated hemivertebrae. This abnormality is also referred to as sagittal cleft vertebra, anterior spinal cleft, or body cleft [3]. This developmental anomaly predominantly occurs in the lumbar vertebrae, and secondly in the middle thoracic vertebrae (T5-T9) [4, 5], It is rarely seen in the lower thoracic vertebrae. Butterfly vertebrae generally do not present with typical symptoms and are usually discovered incidentally or in the presence of back pain, which may be caused by changes in spinal biomechanics [6]. Case reports of multiple butterfly vertebrae occurring simultaneously in the lower thoracic vertebrae are also rare, and most butterfly vertebrae case reports focus on imaging results, with only a few papers discussing surgical treatment strategies [7, 8]. We report a case of a 14-year-old adolescent patient with chronic back pain associated with butterfly vertebrae at the T9-T11 level, presenting with kyphosis at the thoracolumbar junction and a knife-back-like scoliosis, which was corrected surgically.

2. Clinical Data:

The patient, a 14-year-old male, was admitted with a complaint of "recurrent chest and lower back pain for over 1 year." The patient was diagnosed with kyphosis at the thoracolumbar junction and a knife-back-like scoliosis (Figure 1) one year ago, following recurrent chest and lower back pain, The patient experienced dull, aching pain in the chest and lower back, occurring intermittently and tolerable, which worsened after prolonged desk work or bending. The skin showed no redness, swelling, ulcers, or sinus formation, and there was no morning stiffness or migratory joint pain. The patient visited the outpatient department of Fenggang County People's Hospital, where a DR examination revealed a scoliosis deformity, but no special treatment was given. The symptoms recurred, varying in intensity, and the patient sought further evaluation and treatment at our hospital, where surgery was recommended, and the patient was admitted under the diagnosis of "scoliosis".

Physical examination upon admission: Kyphosis and a knife-back-like scoliosis deformity were observed at the thoracolumbar junction. No obvious deformities were noted in both upper limbs, with normal sensation, normal muscle strength, and no increased muscle tone. Both Hoffmann's signs were negative. Tension and swelling of the paraspinal muscles on the left side of the thoracolumbar junction were noted, with localized tenderness. There was no redness, swelling, pigmentation, café-au-lait spots, or hair growth, and no ulceration or sinus formation. Skin temperature was normal. Forward flexion of the lumbar spine was slightly limited. The straight leg raise test (-) and the Bragard's test (-) were negative. No obvious abnormalities were found in the pelvic contour, and the compression separation test was negative. The abdominal reflex and cremasteric reflex were present, and there was no significant relaxation of the anal sphincter. Muscle tone in both lower limbs was normal. Muscle strength in the bilateral quadriceps, tibialis anterior, gastrocnemius, dorsiflexors of the toes, and plantar flexors was graded 5/5. Both sides of the Thomas sign and the "4" sign were negative. Patellar tendon and Achilles tendon reflexes were normal, and no patellar or ankle clonus was observed. The bilateral Babinski sign, Chadonk sign, Oppenheim sign, and Gordon sign were negative.

Radiological Supplementary Examination Data: Full-spine left-bending and anteroposterior/lateral X-rays indicate varying degrees of scoliosis in the cervical, thoracic, and lumbar spine, with a "C"-shaped kyphotic deformity centered at T10 and T11. (Figures 2d, 2e). MRI: 1. Cervical, thoracic,

and lumbar spine scoliosis of varying degrees with kyphotic deformity centered at T10 and T11. | MRI: 1. Scoliosis of varying degrees in the cervical, thoracic, and lumbar spine, with kyphotic deformity centered at T10 and T11.2. Flattening of the T9-T11 vertebral bodies with anterior bony discontinuity, forming a butterfly vertebra; C5-C6 and L3-L4 intervertebral disc space narrowing; hypertrophy of the left transverse process of L5 with pseudoarthrosis formation.3. Discontinuity of the lateral twelfth rib cortex; clinical correlation is recommended.4. Abnormal soft tissue signal behind the right inferior articular process of L5, hemangioma? Cyst? Contrast-enhanced scanning if necessary.CT: 1. Pneumonia in the right lower lung lobe; follow-up recommended after treatment.2. Axillary and mediastinal lymph nodes are visible.3. Cervical, thoracic, and lumbar spine scoliosis of varying degrees with kyphotic deformity centered at T10 and T11; T9-T11 vertebral bodies are flattened with anterior bony discontinuity; kyphotic Cobb angle = 79° (Figure 2f).4. Flattening of the T9-T11 vertebral bodies with anterior bony discontinuity, forming a butterfly vertebra (Figure 3g, 3h); narrowing of the C5-C6 and L3-L4 intervertebral disc spaces; hypertrophy of the left transverse process of L5 with pseudoarthrosis formation. No significant abnormalities were observed in the hematological assessment.

3. Surgical Methods

Surgical procedure: Thoracic vertebral hemivertebra resection, titanium cage bone graft fusion, pedicle screw internal fixation; scoliosis and kyphosis correction, thoracic deformity correction, and posterolateral bone grafting. Procedure: After the anesthesia took effect, the patient was placed in a prone position with the abdomen supported and undergoing electrophysiological monitoring, while an autotransfusion device was connected. Significant scoliosis and kyphosis deformity of the spine were observed intraoperatively. Preoperatively, the C-arm was used to locate the T12 vertebra, followed by routine disinfection and draping. A 25 cm midline incision centered at T12 was made, with skin and subcutaneous tissue dissected layer by layer to expose the thoracolumbar fascia. An electrocautery was used for incision and hemostasis, and dissection was performed along both sides of the spinous processes to expose the laminae from T6 to L2, extending to the level of the articular processes. During the procedure, scoliosis and kyphosis were noted in the lumbar spine, with T9-T11 being the primary deformity center. The positioning needle was accurately placed in the T12 vertebra, and C-arm fluoroscopy confirmed the accuracy of the positioning. Pedicle screws were placed on both sides of the T6, T7, T8, T12, L1, and L2 vertebrae, and C-arm fluoroscopy confirmed the screw position and length were as expected. A temporary fixation rod was connected to the right side. The right ribs were notably higher than the left, and a bone-biting forceps was used to remove the interspinous processes of T9-T11. An ultrasonic bone scalpel was applied to cut both sides of the T9 and T10 laminae, exposing the bilateral costovertebral joints and part of the ribs at T9 and T10. Using an ultrasonic bone scalpel, the left T10/T11 and right T10 costovertebral joints and approximately 5 cm of exposed ribs were resected to reconstruct the thoracic cage. An “eggshell” technique was used to remove the left T10 and T11 hemivertebrae and intervertebral discs, and the right side was similarly treated. After measuring the vertebral height, an appropriately sized titanium alloy intervertebral fusion cage was placed into the left osteotomy window, and the patient’s autologous bone was used as graft material. Anterior expansion correction was performed, the left fixation rod was connected, and posterior pressure was applied on the left side to gradually close the posterior osteotomy region and correct the kyphosis deformity. The right side was expanded and fixed, and spinal correction was performed. No spinal cord compression was observed during the correction, and no abnormal signals were detected during electrophysiological monitoring. Lateral bone grafting was performed on the right side, with C-arm fluoroscopy confirming the ideal position of the internal fixation. The scoliosis and kyphosis deformities in the thoracolumbar segment were effectively corrected, restoring normal physiological curvature. The incision was repeatedly irrigated with a large amount of saline during the procedure, and hemostasis was achieved with bipolar electrocautery. After confirming the instruments and gauze, a negative pressure drainage tube was placed, and the incision was closed layer by layer and dressed with sterile dressing. Anesthesia was effective, with an estimated blood loss of 2700 mL, 900 mL of autologous blood reinfused, and no transfusion reactions. The surgery was completed smoothly, with satisfactory anesthesia, and no abnormal waveforms were seen during intraoperative electrophysiological monitoring. The patient was transferred to the ICU for close observation postoperatively, with no adverse reactions or abnormal symptoms during follow-up at 1, 3, and 6 months, and 1 and 2 years.

4. Discussion

Butterfly vertebra (BV) occurring in the lower thoracic spine with multiple vertebrae affected simultaneously is an extremely rare congenital spinal developmental deformity that can lead to kyphosis or scoliosis. Butterfly vertebra is a congenital developmental abnormality in the early embryonic stage, primarily characterized by incomplete development on both sides of the vertebral body, usually with two developing ossification centers on each side of the vertebra. These ossification centers are supposed to fuse to form a complete vertebra during normal development. However, if this process is incomplete, it may result in incomplete fusion on both sides of the vertebral body, ultimately leading to the formation of a butterfly vertebra[9]. According to literature reports, a systematic review reveals that the age of onset for butterfly vertebra is mostly in young adults[10]. Congenital vertebral abnormalities are quite common, with a global prevalence of 0.5-1/1,000; however, BV anomalies presenting as sagittal cleft vertebrae are rare. In this case, the patient had developed kyphosis and scoliotic deformity in the thoracolumbar spine more than a year ago, with multiple butterfly vertebrae occurring in the lower thoracic spine, which is extremely rare. Abnormalities of the spine may be associated with congenital syndromes, such as VACTERL syndrome[11], but they can also occur without any related syndrome[12]. VACTERL syndrome is the most common vertebral developmental anomaly syndrome, and its diagnosis is typically based on the presence of three or more of the following manifestations: vertebral defects, anorectal malformations, cardiac defects, tracheoesophageal fistulas, renal abnormalities, and limb deformities[11]. This patient did not present with VACTERL syndrome. Butterfly vertebrae typically do not cause

neurological symptoms, but they may affect spinal stability and can be easily confused with pathological fractures, infections, and associated vertebral anomalies or syndromes[13]. For example, Alagille, Jarcho-Levin, Crouzon, and Pfeiffer syndromes, especially when confused with vertebral compression fractures[14], Therefore, imaging studies are important diagnostic tools. Further evaluation with CT, X-ray, and MRI for other spinal abnormalities (such as multiple lumbar vertebrae, spina bifida, myelomeningocele, and kyphosis) showed no abnormalities. Imaging revealed congenital developmental deformities at T9-T11 vertebrae, presenting as butterfly vertebrae, with no significant vertebral compression fractures. In this context, the patient did not show any clinical features of known associated syndromes.

Due to the progressive worsening of the kyphotic deformity and knife-back-like scoliosis, along with increasing back pain affecting the patient's quality of life, a surgical plan was made after good communication with the patient. The patient underwent general anesthesia for thoracic vertebra hemivertebra resection, titanium cage bone fusion, and pedicle screw internal fixation; spinal deformity correction, thoracic remodeling, and posterolateral bone grafting. This allowed for maximal resection of the pathological vertebra and correction of the kyphotic and scoliotic deformities. Postoperative follow-up anteroposterior and lateral X-rays show slight kyphosis centered at T10 and T11, with internal fixation in place and good fusion. **(Figure 3i).** Long-term follow-up revealed significant improvement in the postoperative spinal kyphosis, with no complications.

Authors' Contributions

XC, NZ, YJL and BWZ analyzed the data that we had collected and drafted the manuscript. ZC put forward the idea of this study and revised the manuscript. All the authors involved have read.

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Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

Competing interests

The authors have no conflicts of interest to disclose.

Figure legends

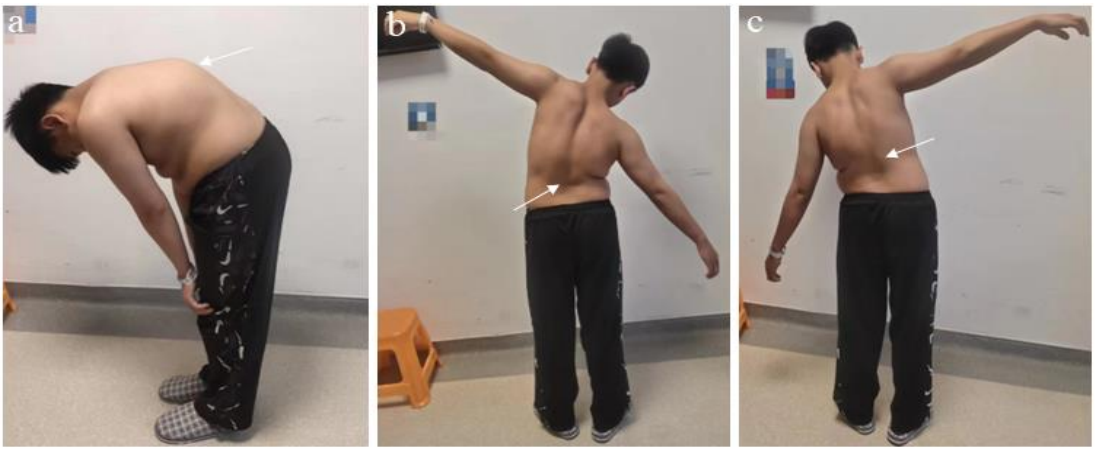

Figure 1: (a-c) Preoperative patient showed kyphosis at the thoracolumbar junction and a knife-back-like scoliosis deformity.

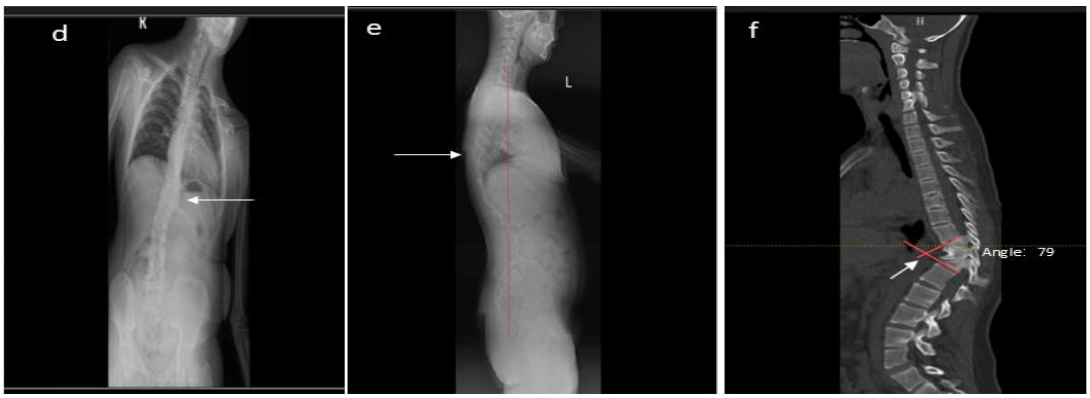

Figure 2: Figure 2. (d, e) Preoperative full-spine left-bending and anteroposterior/lateral X-rays show varying degrees of scoliosis in the cervical, thoracic, and lumbar spine, with a "C"-shaped kyphotic deformity centered at T10 and T11 (arrow).(f) Preoperative thoracolumbar CT indicates flattening of the T9-T11 vertebral bodies with anterior bony discontinuity, forming an angular deformity with posterior and rightward displacement centered at this region, with a thoracic kyphotic Cobb angle of approximately 79°.

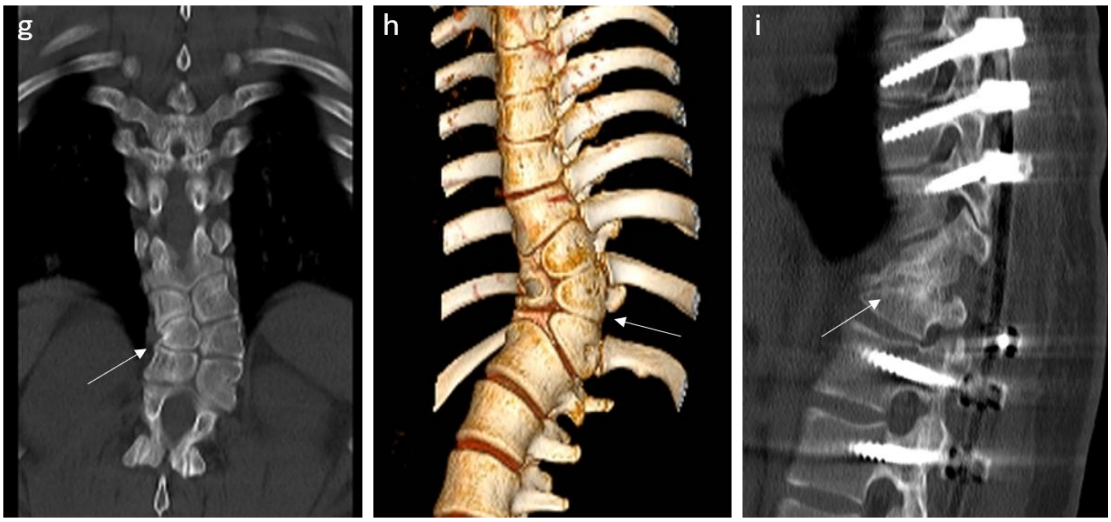

Figure 3: Figure 3. Preoperative CT (3D reconstruction) shows: (h) varying degrees of scoliosis in the cervical, thoracic, and lumbar spine, with kyphotic deformity centered at T10 and T11; (g) flattening of the T9-T11 vertebral bodies, with anterior bony discontinuity forming a butterfly vertebra (arrow).(i) Postoperative follow-up anteroposterior and lateral X-rays show mild kyphosis centered at T10 and T11, with internal fixation in place and good fusion.

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