

CASE REPORT

Anterior Endoscopy Combining with Modified Total En Block Spondylectomy for Synovial Sarcoma in Thoracic Paraspine Causing Neurological Deficits: Case Report and Literature Review

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Introduction

Synovial sarcoma is a rare and highly malignant tumor, accounting for 5%–10% of soft tissue sarcoma. Synovial sarcoma affects males more than it affects females. Synovial sarcoma often occurs in large joints of four extremities in young adults, especially in the knee joint. Synovial sarcoma seldom occurs in the spine, and it is rarer in the thoracic paraspinal. No report of thoracic paraspinal tumor causing canal invasion and spinal cord compression exists. Additionally, no standard surgical strategies exist for spinal synovial sarcoma. In the current paper, we have described a case with paraspinal synovial sarcoma treated in our department through posterior modified total en bloc spondylectomy combined with anterior endoscopy.

Case Report

A 16-year-old female patient with no family history of synovial sarcoma was presented with upper back pain for 40 days, with paraparesis, hypalgesia, bilateral lower limb numbness, and disabled walking for 7 days. No evident abnormality was observed on the skin of the whole spine region. Spinous process tenderness and percussion pain were evident in T7/8, and the pain increased at the standing position for a long time. The motor strengths of the left lower limb and the right lower limb were 3/5 and 2/5, respectively.

The bladder function reflex was intact. The deep tendon reflexes in the lower limbs were exaggerated, and the Hoffmann sign and the Babinski sign were positive. Blood tests revealed no evident abnormality. Plain films revealed T8 compression fractures and soft tissue mass shadow of the right-sided T7/8 (Figure 1A,B). MRI demonstrated T8 compression fractures causing spinal cord compression and revealed high T2WI density (Figure 1C) and low T1WI density (Figure 1D). A large soft mass on the right-sided paraspinal was observed (Figure 1E,F). CT revealed osteolytic changes of T7/8 vertebral body and right pedicles, especially the compression fractures of T8 (Figure 1G,I). Preoperative ECT and PET/CT were not abnormal.

Pathology

Using a C-arm machine, we performed a percutaneous needle biopsy of the T7 vertebral body on the right pedicles. The tumor was microscopically composed of spindle cells arranged in bundles or braids, ovoid or rod-shaped nucleus, and small nucleoli. Mitoses were significantly evident. The tumor was well vascularized in the profusely pink cytoplasm. Collagen fibers and degeneration of the local number were discovered. Immunohistochemistry staining was positive for Vimentin and Caldesmon, with a remarkably low proliferation index (Ki-67: 30%–40%, P53:

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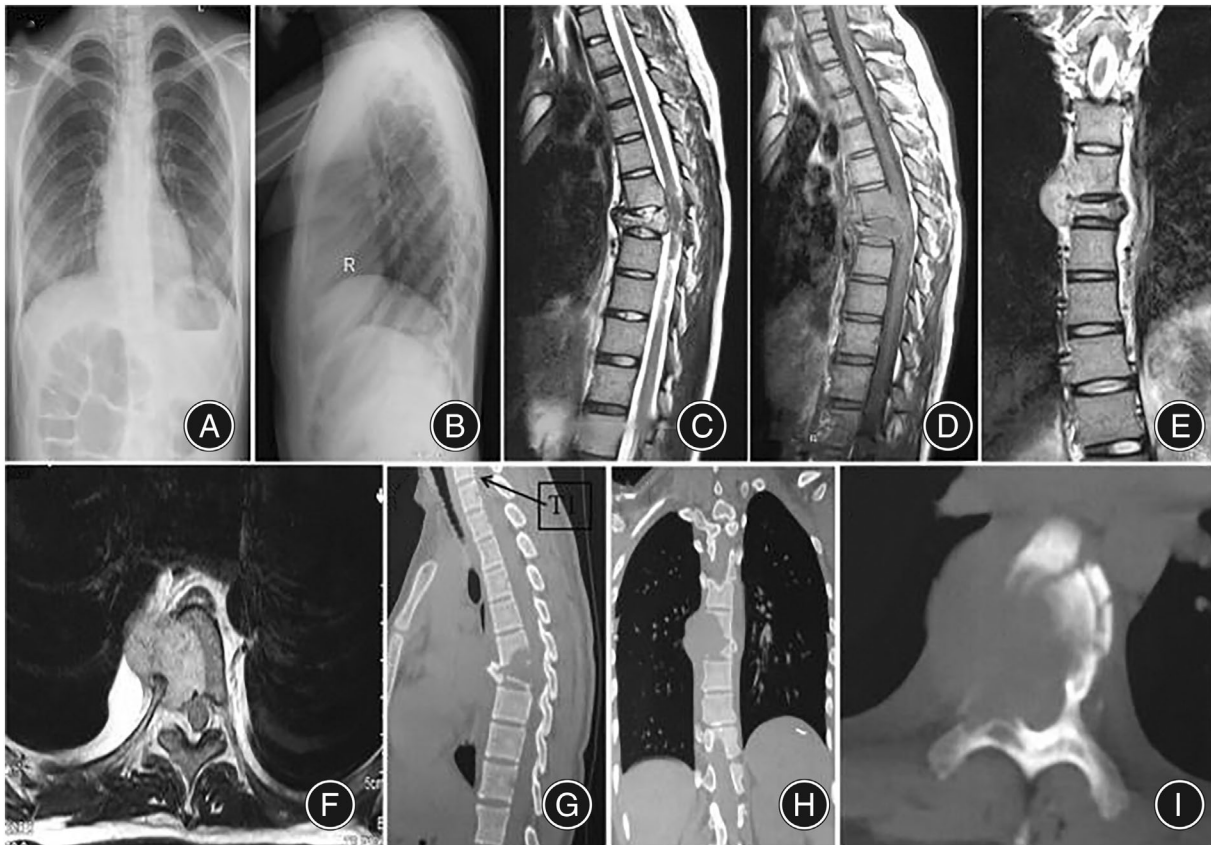


Fig. 1 Preoperative plain radiograph, magnetic resonance imaging and computed tomography. Plain films showed compression fractures of T8 in the anterior posterior (A) and lateral views (B); The MRI showed T8 compression fractures that causing dorsal spinal cord compression (T2 (C), T1 (D)) and soft mass in right-sided of paraspinal (E) and bony erosion on axial (F); CT showed compression fractures of T8 (G) and osteolytic changes of T7/8 vertebral body and right pedicles (H, I)

20%–30%). Microvascular endothelial cells were weakly positive in CD34, SMA, and Calponin. But It was negative for S-100, Bcl-2, CKP, EMA, CD99, CD117, Dog-1, Syn, CgA, and Desmin (Figure 2A–C). The patient received a diagnosis for monophasic synovial sarcoma. The patient was treated with chemotherapy using cyclophosphamide 40 mg every 2 weeks.

Surgery

Written informed consent was obtained from the patient before the surgery. A double-lumen endotracheal tube was used to control the patient's respiration. Thereafter, the patient was placed in a lateral decubitus position. The anterior and lateral components, including nerves, intercostal vessels, large blood vessels, and lungs were separated, ligated, or protected under thoracoscopy inserted through the right anterolateral sixth intercostal space (Figure 3A). The two-sided ribs head was resected at the involved segment level using a high-speed drill outside the costotransverse joint of approximately 3–4 cm.

The patient was placed in a prone position, and a dorsomedial skin incision was made from T5 to T10. Paraspinal

muscles and other soft tissues under the periosteum were stripped off. Four pedicle screws were inserted into the pedicles, two levels above T7 and two levels below T8. At the same time, a unilateral rod was installed to stabilize the spine.

The wire guide with a sleeve was passed through the intervertebral foramen and clung to the surface of the pedicle and lamina. Thereafter, the wire-saw was inserted into the sleeve of the wire guide. The wire-guide was removed, and the pedicles were sawed using the wire-saw placed on the superior articular and transverse processes below. After cutting off, the osteotomy surface was immediately sealed with bone wax, and the rear attachment was removed.

After that, an “S-shape” protective spatula with a sleeve was inserted from the posterior end to the anterior end through the interface between the anterior vertebral body and the anterior longitudinal ligament. A wire saw was inserted into the sleeve of the spatula, and one of the ends of the wire was extracted from the other side of the vertebral body. The spinal cord protective baffle (SCPB) was inserted through the interface between the posterior longitudinal ligament and posterior edge of the vertebral body and was lightly pulled backwards SCPB. The wire saw was

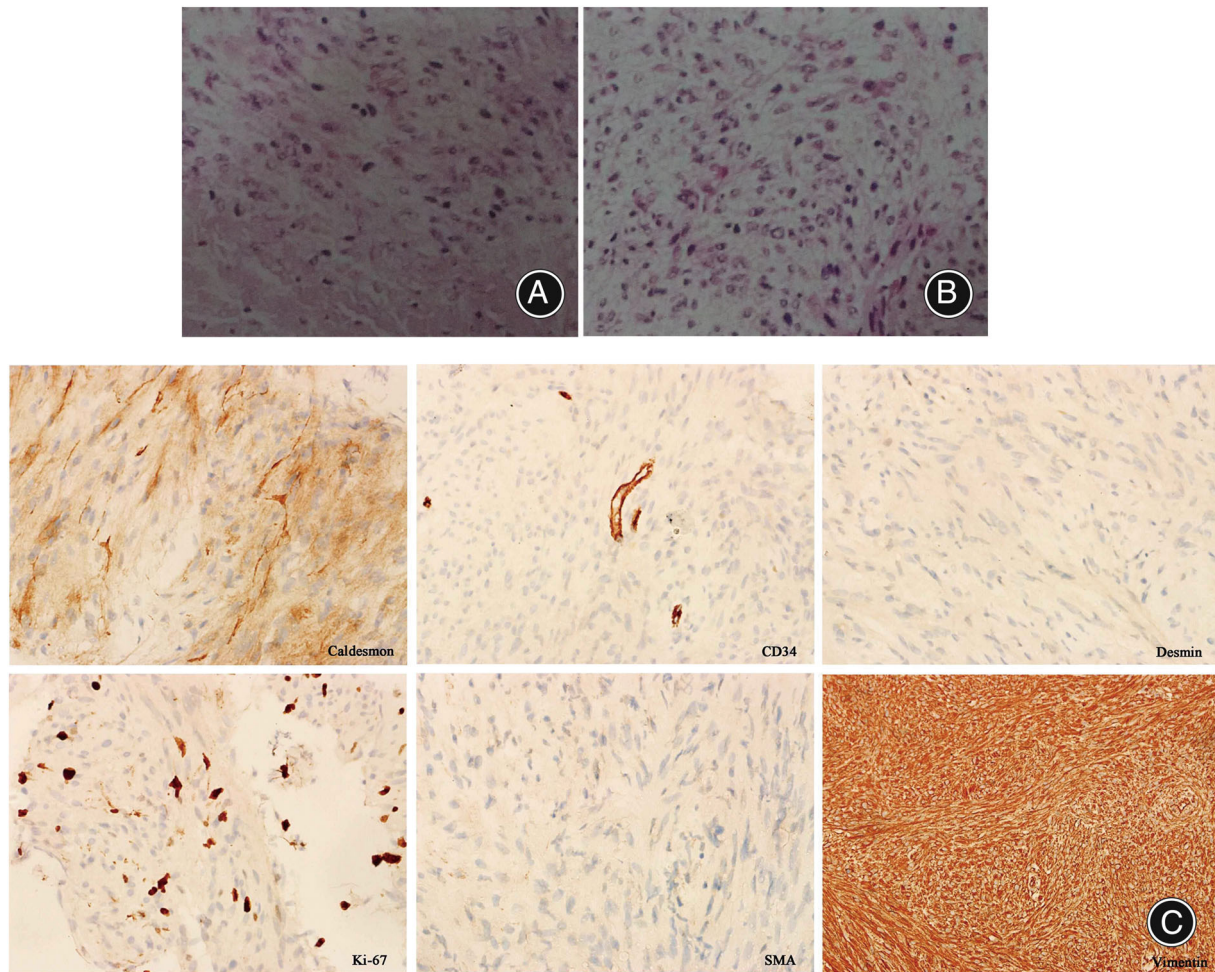


Fig. 2 Histopathology of the tumor. (A) The tumor mainly compresses short spindle cells with deeply stained nuclei. Few mitoses are observed (Hematoxylin and eosin [H&E]); (B) Some areas of vascular proliferation show larger vessels arranged in a “staghorn” pericytoma pattern; (C) Immunostaining for CD34 reveals many tumor cells with positive staining in the cytoplasm

inserted into SCPB (Figure 3B). Finally, both ends of the wire saw were on the spine's same side.

The anterior vertebral body was dissected using the wire saw reciprocating while keeping the SCPB stable. The affected vertebra was pushed approximately 5–10 mm forward and meticulously rotated along its longitudinal axis, achieving a spinal cord circumferential decompression and a segmental total en bloc corpectomy¹ (Figure 3C). Finally, dorsoventral reconstruction was carried out using titanium mesh filled with the ilium, and the rod on the other side was installed. The surgery took 240 min, and the estimated amount of bleeding during the operation was 900 ml. No neurological complications were observed.

Post-Operation

Motor and somatosensory revealed no change during the surgery. The pale red tumor was lying in the left front of the vertebral body, which was $3 \times 3 \times 3.5$ cm³, with adhesion, soft

and easy bleeding. Postoperative pathology and immunohistochemistry were consistent with preoperative pathology. The pathological examination of the upper and lower edges of the vertebral body was negative after the operation.

After the surgery, the patient stayed in bed for approximately 7 days and was treated with chemotherapy using cyclophosphamide three times. The dosage of cyclophosphamide was 40 mg every time. Additionally, the patient received external beam radiation therapy. The dosage was 200 Gy per day, and the treatment continued for 6 weeks, 5 days per week. While walking 2 weeks after the surgery, the patient wore a cervicothoracic brace. The paraparesis, hypalgesia, and numbness in the bilateral lower limb of the patient gradually improved. The motor strength of the patient of the left lower limb and the right lower limb 3 months after the surgery and pain palliation was 5/5 and 4/5, respectively. The patient performed routine life tasks for 8 months, although she was diagnosed

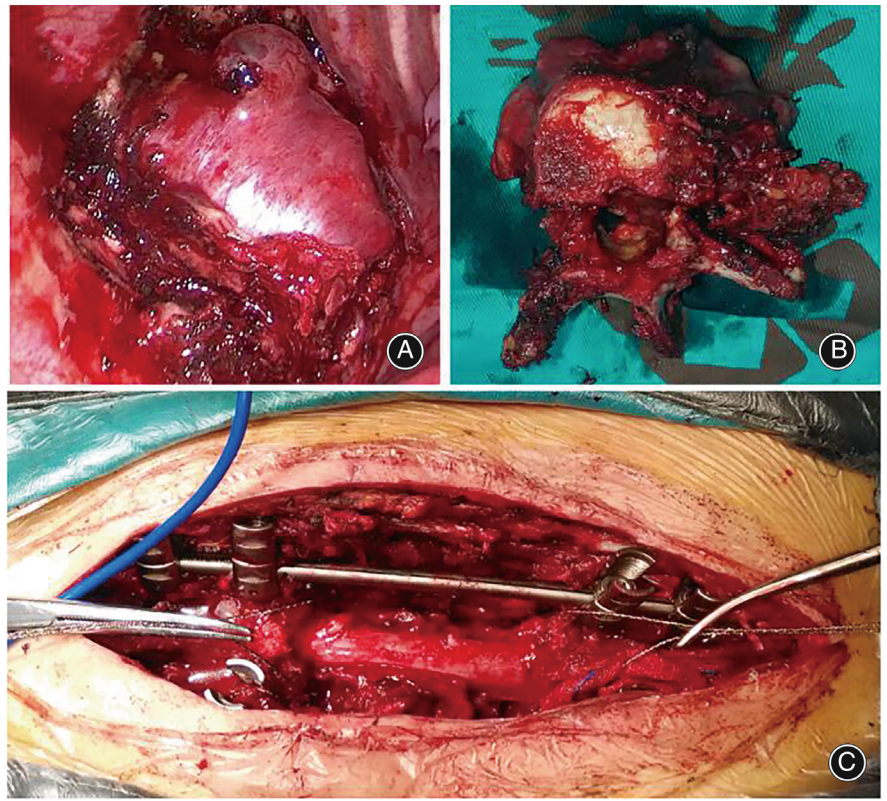


Fig. 3 Intraoperative photographs: Tumor tissue in front of vertebral body under thoracoscopy (A); wire saw was cutting off the anterior vertebral body (B); resected vertebra (C)

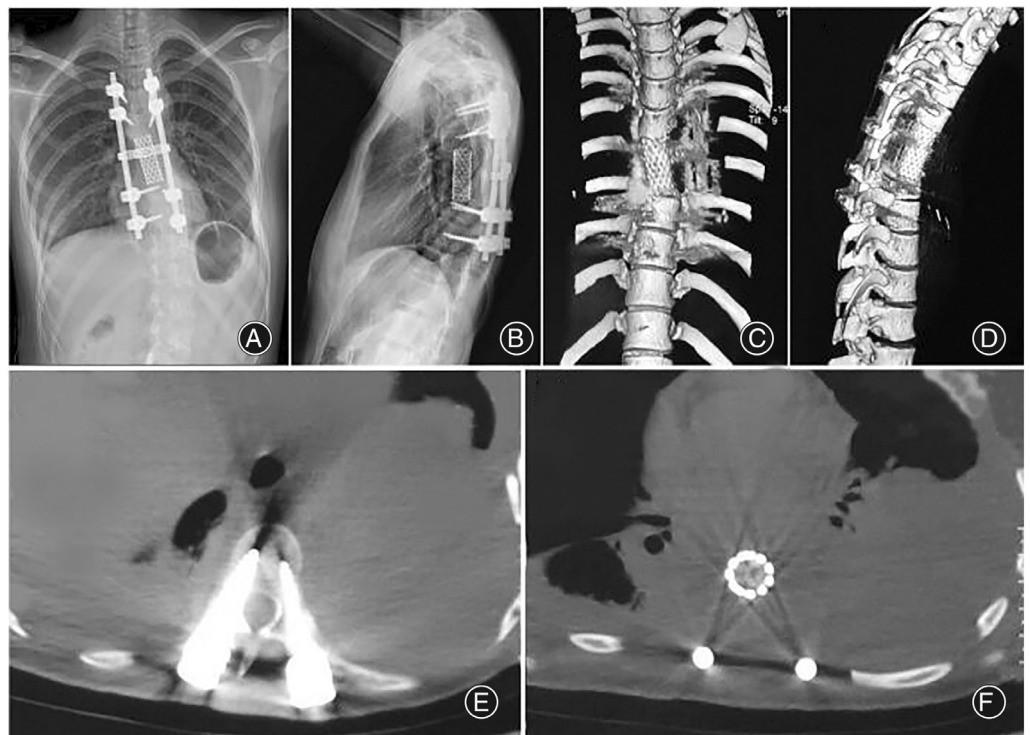


Fig. 4 Plain radiograph and computed photograph at 6 months after surgery: Original scoliosis increased in anteriorposterior (A) and lateral views (B); postoperative 3-D (C, D) and transverse sections (E, F) showed no failure in thoracic interfixation

with synovial sarcoma in thoracic paraspine. MRI and PET/CT illustrated no evidence of recurrence, but original scoliosis increased without affecting the patient's daily life (Figure 4).

Discussion

Clinical Features

Synovial sarcoma is an uncommon and highly malignant tumor accounting for 5%–10% of all soft tissue sarcoma. Synovial sarcomas are more common in males than females. It often occurs in large joints of four extremities in young adults—with an average age of 35 years, usually in the knee joint.² Synovial sarcoma seldom occurs in the spine, rarer in the thoracic spine. Cao and colleagues³ reported a patient with thoracic paravertebral synovial sarcoma originated from costovertebral joints and invaded the spinal canal through the intervertebral foramen. No report of thoracic paravertebral synovial sarcoma invading the spinal canal causing spinal cord compression has been reported. The current report is the case of a T7/8 paraspinous synovial sarcoma causing limb weakness, hypesthesia, and disabled walking.

Diagnosis

Radiological Features

No specific findings other than a soft tissue mass and bone destruction in the adjacent joint was observed on plain film.⁴ However, approximately 30% of the soft tissue tumor revealed calcification at the edge of the mass, whose reason is not very distinct. It is related to bleeding, infection, gangrene, and cartilage formation.⁵ T7/8 compression fractures of bone destruction and visible peripheral soft tissue masses in the current patient were observed.

Because of synovial sarcoma's varying appearance, plain films vary as well, with usually no apparent specificity. However, CT scans could make up for these deficiencies. The accuracy and sensitivity of CT scans can reveal subtle calcification and bone destruction, the relationship between soft tissue tumor and adjacent bone or other tissues, and the invasion of vertebral spinal cord volume that cannot be revealed on plain films.⁵ In the current case, both X-ray and CT revealed T8 compression fractures and soft tissue mass on the right side of the T7/8 paraspine. CT scan revealed bone destruction in T7/8 paravertebral body and right side pedicle of T8.

Evident patchy calcification at the edge of the vertebral body and spinal cord compression was observed.⁶ MRI is more sensitive and accurate than CT and plain films for diagnosis and staging purposes. A total of 40% of synovial sarcoma revealed hyperintense signal on T1WI and intermediate signal on T2WI, which may be related to bleeding.⁷ Synovial sarcoma can be demonstrated with "triple signal intensity": hyperintensity, iso intensity, hypointensity.⁸ In the current case, the mass performed T1 low signal and T2 high signal. Preoperative ECT and PET/CT examination can

determine metastasis,⁹ and the current patient exhibited no metastasis.

Pathological Features

Although most synovial sarcoma does not originate from the synovial tissue, the term arose from similar microscopic features to the developing synovium.⁷ Ultrastructural and immunohistochemistry examination revealed that the tumor has epithelial or spindle cells. Three histologic variations based on the combination of spindle and epithelial tumor cells exist, the classic biphasic synovial sarcomas, the monophasic, and the poorly differentiated subtype.¹⁰ The biphasic synovial sarcomas are only composed of epithelial or spindle, but the monophasic synovial sarcomas are characterized by epithelial cells mixed with spindle cells.¹¹

Many markers can be expressed in the immunohistochemical assay of synovial sarcomas, such as CD34, SMA, Calponin, and Bcl-2, S-100. Recently, the molecular study of synovial sarcoma has achieved preliminary progress. miRNAs play an essential role in tumor gene diagnosis, but miRNAs have some limitations in identifying muscle tumors and synovial sarcoma.¹² *t*(X;18) (p11.2;q11.2) is specific in the diagnosis of synovial sarcoma, and more than 90% synovial sarcoma is present in this gene.¹²

Treatment

The best treatment for synovial sarcoma is radical resection with negative margins combined with sequential radiotherapy and chemotherapy after the surgery.¹³ Radiotherapy is considered an effective adjuvant therapy for synovial sarcoma that can reduce the local recurrence and prolong the survival period of patients, but it can also improve the quality of the patient's life. Chemotherapy has no evident effects on synovial sarcoma compared with radiotherapy. Chemotherapy can be effective in combination with radiotherapy. The most commonly used chemotherapy regimen is cyclophosphamide combined with adriamycin. However, en bloc resection is the ideal treatment modality, which is the basis of chemotherapy.

Because of the complexity and particularity of anatomical structure around the thoracic spine and the synovial sarcoma's unique characteristics, it is difficult to operate the sarcoma completely. Ross *et al.* reported a patient with thoracic synovial sarcoma treated by en bloc resection. Although the operations mentioned above can reduce the recurrence rate and mortality, both block resection and transthoracic are intralesional resections and surgical trauma, leading to more bleeding, so that patients need more recovery time and higher local recurrence.

Advantages of our Surgical Method

According to the tumor surgical staging system of Tomita, the patient was identified as type VI and had undergone a posterior modified total en bloc spondylectomy combined with anterior endoscopy. Compared with traditional resection, this technique has five advantages as follows:

(1) The large vessels, segmental vessels, nerves and other soft tissue or organs in front of the vertebral body were ligated, separated, and protected. This significantly reduced the amount of bleeding during surgery. In this report, the operation time was 280 min, shorter than that of Melcher *et al.*,¹⁴ who reported at 9.2 ± 3.1 h (range 5.3–16.4 h), and the intraoperative bleeding was 900 ml, which was shorter than that of 2000 ml reported by Demura and colleagues¹⁵; (2) We detached the paraspinous muscles under periosteum that not only significantly reduced intraoperative bleeding and blood transfusion but also avoided the massive hemorrhage and blood transfusion that could induce internal disorders and diffusive intravascular coagulation. The current patient exhibited no such complications; (3) Modified total en bloc resection was applied to cut the anterior vertebral body, which was a simple and ingenious design to avoid the complications of spinal cord injury; (4) Motor and somatosensory evoked potential was used during the operation to detect the traction and injury of the spinal cord time, and reduce the recovery time; (5) A double-lumen endotracheal tube was used to control respiration, and can be good to deal with the emergency during operation.

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None.

Authorship Contribution

Zengping Wang conducted statistical analysis and drafted the manuscript. Jie Wen and Chunzhen Ren participated in the statistical analysis and the writing of the manuscript. Lin Liu conceived the research, participated in the research design and coordination. Wen Xue and Yuxin Song participated in clinical research and surgical treatment. All authors read and approved the final manuscript.

Conflict of Interest

The authors declare that they have no competing interests.

Ethics Statement

The study procedure conformed to the ethical guidelines of the Declaration of Helsinki, and the approval for the study was obtained from the Medical Ethics Committee of Department of Orthopaedics, Gansu provincial Hospital. A written informed consent was obtained from patients enrolled.

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