Successful Treatment for Rhabdomyosarcoma by Total Spondylectomy in a Child

Takeshi Saito, Toshimi Aizawa, Osamu Kashimoto, Tetsuro Sato and Shoichi Kokubun

Department of Orthopaedic Surgery, Tohoku University School of Medicine, Sendai 980-8574

Saito, T., Aizawa, T., Kashimoto, O., Sato, T. and Kokubun, S. Successful Treatment for Rhabdomyosarcoma by Total Spondylectomy in a Child. Tohoku J. Exp. Med., 2002, 198 (4), 251–258 — A 7-year-old girl with a retroperitoneal rhabdomyosarcoma having invaded the L3 vertebra was treated by combination therapy consisting of chemotherapy, surgical resection and intraoperative radiation. Surgically, total spondylectomy was performed through a combined anterior and posterior procedures, and the spine was reconstructed with fibula bone grafts using a pedicle screw system made of titanium alloy. Transverse junction plates were not used at operation because of their bulkiness for the patient. Five days postoperatively, the rods dislodged from the screws and the grafted bones leaned. A Chance fracture-like transverse fracture of the L4 vertebra occurred during the revisional operation. Transverse plates and sublaminar wires were used to reduce the fracture and achieved stability. Six years postoperatively, the spine was completely fused and no local recurrence or metastasis is detected. —— rhabdomyosarcoma; total spondylectomy; child

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Rhabdomyosarcoma is one of the most common soft tissue sarcoma in children under 15 years of age (Raney et al. 2001; Weiss and Goldblum 2001) and is histologically classified into four categories; embryonal, botryoid-type embryonal, alveolar and pleomorphic (Weiss and Goldblum 2001). The embryonal type accounts for approximately 50% and mostly affects children younger than 10 years old (Raney et al. 2001; Weiss and Goldblum 2001).

Paraspinal rhabdomyosarcomas generally arise in the retroperitoneal space and could invade the vertebrae (Heller and Pedlow 1997). Total spondylectomy followed by spinal reconstruction for the malignant tumors in the spine generally prevents local recurrence and provides a long-term survival (Lièvre et al. 1968; Stener 1971; Magerl and Coscia 1988; Sundaresan et al. 1988; Tomita et al. 1997). To our knowledge, however, only two cases of total spondylectomy have been reported in children less than eight years old, which had been performed

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Address for reprints: Toshimi Aizawa, M.D., Ph.D., Department of Orthopaedic Surgery, Tohoku University School of Medicine, 1-1 Seiryo-machi, Aoba-ku, Sendai 980-8574, Japan.
e-mail: HYPERLINK mailto:toshi-7@ra2.so-net.ne.jp

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only for rhabdomyosarcoma and described in Japanese (Takeda et al. 1992).

We report a patient of 7-year-old girl sustained by retroperitoneal rhabdomyosarcoma having invading the L3 vertebra. Total spondylectomy and spinal reconstruction through combined anterior and posterior procedures were performed in combination with chemotherapy and intraoperative radiotherapy. The spinal fusion completed and the patient was free from the disorder without local recurrence at the latest follow-up of six years postoperatively.

**CASE REPORT**

A 7-year-old girl presented with low back pain and a right curved scoliosis and came to our hospital in November 1993. On physical examination, an elastic soft tumor was palpated with tenderness in the left abdomen but no abnormal neurologic findings were detected excepting the weakness of the left iliopsoas muscle.

A plain radiograph showed a 25° right convex scoliosis caused by wedging destruction of the left half of the L3 vertebral body (Fig. 1). Magnetic resonance (MR) imaging revealed a tumor mass which was located in the left retroperitoneal region expanding the spinal level of from L2 to the sacrum and had invaded the L3 vertebral body and the epidural space (Fig. 2). Aspiration biopsy disclosed that the tumor was composed of small round or spindle shaped cells with deeply eosinophilic cytoplasm and nuclei of various size and shape. It was diagnosed as an embryonal rhabdomyosarcoma.

Chemotherapy, consisting of adriamycin (doxorubicin), clophosphamid, cisplatin (CPPD), CPPD plus etoposide (VP-16) and daunomycin, was given according to the IRS protocol (Maurer et al. 1993; Crist et al. 1995), thereby resulting in marked reduction of the tumor size on MR images (Fig. 3). Excision of the retroperitoneal tumor and total spondylectomy of L3 were performed in February 1994 in collaboration of pediatric surgeons.

*En bloc* laminectomy of L3 and partial laminectomy of L2 and L4 was performed on prone position. The tumor was found to have invaded the left pedicle and lamina of L3, but no tumor mass was found in the epidural space. Titanium alloy tulip-headed pedicle screws of J.B.S. SPINE System® with 5 mm in diameter were inserted into the L2 and L4 pedicles with the diameter of 5.6 mm and 6 mm, respectively (Marnay and Hupper 1996). The cortices of the pedicles were obviously thin in CT scans indicating osteoporosis. The posterior longitudinal ligament (PLL) and the posterior halves of the intervertebral discs at L2/3 and L3/4 were cut transversely. The rods were assembled to the screws. Posterolateral fusion was conducted with iliac bone grafts. No transverse junction plates were used because it looked too bulky for the patient. After the patient's position was changed to supine, the retroperitoneal part of the tumor was removed through an extraperitoneal approach and intraoperative radiation of 12 Gy was perfor-

![Fig. 1. A plain radiograph showing the destructive change of the L3 vertebra and the right convex scoliosis.](image-url)
med. Subsequently, the L3 body was resected en bloc by incising the anterior longitudinal ligament (ALL) and adjacent intervertebral discs. Disc materials and cartilaginous end plates of the L2 and L4 bodies were removed. Two fibula struts were grafted between the bodies (Fig. 4). Histological examination revealed that the removed tumor tissue was

Fig. 2. Gd-EDTA enhanced MR images on admission. a: Coronal view. A paravertebral tumor locates from the L2 vertebra to the sacrum and pushes up the left kidney. b: Axial view. The tumor spreads over the left retroperitoneal space and invades into L3 body and the epidural space.

Fig. 3. Gd-EDTA enhanced MR images after the chemotherapy. a: Coronal view. The tumor has significantly reduced its size so that it can be detected only around the L3 body. b: Axial view. The tumor locates at the left L3 pedicle and paravertebra.
Fig. 4. A plain radiograph after total spondylectomy of L3 showing spinal reconstruction between L2 and L4 with two fibula bone grafts instrumented with pedicle screws and rods of J.B.S. SPINE system.

Fig. 5. An AP radiograph five days postoperatively showing both rods off from the pedicle screws and the grafting bones leaning rightward.

Fig. 6. An AP view of radiograph after the second operation. The transverse junction plates are used. Sublaminar wirings are added between the L4 lamina and the transverse plate because a transverse vertebral fracture was occurred just below the L4 pedicle screws to the lamina.

totally necrotic.

Five days postoperatively, it was found on a radiograph that the rods had bilaterally dislodged from the screws at L4 and the grafted struts been leaning to the right (Fig. 5). Revision was performed immediately with the patient in the prone position. After partial removal of the L4 superior articular processes, two transverse junction plates were used in order to achieve a secure construct. But the L4 pedicles were fractured transversally just below the screws like a Chance fracture by a large twist exerted when a nut was driven home to the screw head over the rod. Sublaminar wirings were added between the L4 lamina and the junction plate so that the fracture was reduced and solid stability was obtained between the L2 and L4 vertebralae (Fig. 6).

Additional chemotherapy according to the IRS protocol and autologous peripheral blood
stem cell transplantation was performed by pediatricians (Maurer et al. 1993; Crist et al. 1995; Sato et al. 1998). Four months after the re-operation, the patient started walking with a brace. Six years postoperatively, the L2 and L4 bodies were completely united with lumbar lordosis although a 10° right curved scoliosis was remained (Fig. 7). No back pain, no local recurrence or metastasis was detected.

**DISCUSSION**

The clinical status of rhabdomyosarcoma is classified by Intergroup Rhabdomyosarcoma Study (IRS) into four groups according to the extent of disease and type of surgery performed: Group I—completely removal of localized disease with negative regional nodes; Group II—grossly resected regional disease with microscopic residual tumor, with or without regional lymph node involvement, or completely resected regional disease and resected positive nodes without microscopic residual tumor; Group III—incomplete resection with gross residual disease; and Group IV—metastatic disease at diagnosis (Maurer et al. 1988, 1993; Raney et al. 2001; Weiss and Goldblum 2001). This classification is valuable for predicting the effectiveness of the therapy (Maurer et al. 1988, 1993; Raney et al. 2001; Weiss and Goldblum 2001). Eighty-eight and 85%, 5-year survival were reported for Group I and II respectively by combination therapy consisting of surgical removal, radiation therapy and multiagent chemotherapy (Raney et al. 2001; Weiss and Goldblum 2001). Since the tumor had invaded the left pedicle and seemed unremovable without microscopic residues by any techniques of total spondylectomy (Tomita et al. 1997), the tumor in the present case belonged to Group II. As a long-term survival was expected by the combination therapy, total spondylectomy was performed.

Total spondylectomy was first reported by Lièvre et al. (1968) for a giant cell tumor in the lumbar spine, which was a two-staged operation through anterior and posterior approaches.
Thereafter, excellent results of total spondylectomy were independently reported by Stener (1971), Magerl and Coscia (1988), Sundaresan et al. (1988), and Tomita et al. (1997). Magerl and Coscia (1988) first described a total vertebrectomy via a single posterior approach. Tomita et al. (1997) named their technique as a total en bloc spondylectomy, which is performed using a threadwire saw through also a single posterior approach. It might be the most ideal procedure in terms of preventing contamination from tumor cells, while the other total spondylectomy techniques involve piecemeal resection of the tumor. However, as the authors themselves pointed out (Tomita et al. 1997), the technique has many risks, such as contamination by tumor cells during pediculotomy and injury of the major vessels during blunt dissection of the anterior aspect of the vertebral bodies. Albeit the surgical approach is more invasive than the single posterior approach, the combined anterior and posterior approach is the only procedure which allows to reach the vertebral body with direct visualization. In the present case, the tumor mass could be removed only partially through the posterior approach, since it was widely spread in the retroperitoneal space. Therefore, we selected the combined anterior and posterior approaches.

Spinal instrumentation with pedicle screws in children has more difficulties than that in adults. There were few spinal implants applicable to children on the market in Japan at that time. We chose J.B.S. SPINE system® which had implants made of titanium alloy suitable for postoperative MR imaging and had pedicle screws of 5 mm in diameter, which were smaller than the diameters of the pedicles of the patient (Marnay and Hupper 1986). However, the mechanism joining the rods to the screw heads are rather delicate and it is hard to obtain a feed back of complete tightening because the titanium alloy had a two time higher elasticity than stainless steel (Gotman 1997). Accordingly, the tightening of the rods to the tulip-headed pedicle screws may not have been enough and the dislodgement of the rods occurred postoperatively, as in a case described by Edmunds et al. (1993). In addition, no transverse approximating device of the system was used as they were too bulky. Attachment of the transverse junction plates might have prevented the instrument failure as they increases the rigidity of the construct, particularly against torsional stress (Gurr et al. 1988; Abumi et al. 1989).

Various intraoperative complications associated with pedicle screws have been reported: screw misplacement, pedicle fractures, cerebrospinal fluid leakage, and vessel injuries (Esses et al. 1993). A transverse fracture of the L4 body occurred through the pedicles like a Chance fracture at the re-operation in the present case. The most plausible cause of this fracture is the excessive occupancy of the screws in the fractured pedicles. It has been reported by several authors that the incidents of pedicle fracture were from 0.3% to 2.3% and that pedicle fracture or screw cut out tends to occur when the screw thread diameter is larger than 80% of the measured outer cortical diameter of the pedicle (Misenhimer et al. 1989; Esses et al. 1993; Lonstein et al. 1999). In the present case, the screw used had a diameter of 5 mm which is more than 80% of the diameter of the L4 pedicles fractured. The second possible cause is the weakness of the pedicles. The cortical bone of pedicles is thinner in children than in adults. In addition, the spine had become osteoporotic because of the preoperative chemotherapy and long recumbency.

Total spondylectomy with spinal reconstruction for children has more difficulties than for adults because of the technical and implants' problems. But in the present case, total spondylectomy in combination with chemotherapy and radiotherapy has led to no local recurrence and good spinal fusion although there were a couple of complications after and during operations. After completing
the spinal fusion, no back pain has been detected and good lordotic curve has been kept six years postoperatively. Total spondylectomy for children should be useful to prevent local recurrence and bring a long-term survival.

References


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