5-Level Spondylectomy for En Bloc Resection of Thoracic Chordoma: Case Report

Daniel M. Sciubba, MD*
Ziya L. Gokaslan, MD*
James H. Black, III, MD‡
Oliver Simmons, MD§
Ian Suk, BSc, BMC*
Timothy F. Witham, MD*
Ali Bydon, MD*
Jean-Paul Wolinsky, MD*

Departments of *Neurosurgery, ‡General Surgery, and §Plastic Surgery, Johns Hopkins University, Baltimore, Maryland

Correspondence:
Jean-Paul Wolinsky, MD,
Department of Neurosurgery,
Johns Hopkins University,
600 North Wolfe Street,
Meyer 7-109,
Baltimore, MD 21287.
E-mail: Jwolins3@jhmi.edu.

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BACKGROUND AND IMPORTANCE: Primary tumors of the spine are considered for en bloc resection to improve local control and even obtain cure. Anatomic restrictions often prohibit extensive resections with negative margins that are safe and feasible. We report the first case involving a patient with a large chordoma of the thoracic spine who underwent a successful 5-level spondylectomy with bilateral chest wall resection for en bloc resection without neurologic compromise.

CLINICAL PRESENTATION: A 26-year-old woman with a chest mass was found to have a T1-5 chordoma via a percutaneous biopsy. En bloc resection of the mass was thought to be the best option for long-term local control and possible cure. She presented without neurologic or pulmonary dysfunction. The patient underwent a 3-stage procedure. The first stage involved a posterior C2-T8 exposure, allowing release of posterior elements from C7 to T6 and instrumented stabilization from C2 to T8. T1-5 ribs were cut bilaterally, and 2 wire saws were placed ventral to the thecal sac at the C7-T1 and T5-6 disc levels. The second stage involved a right-sided thoracotomy, and the T5-6 wire saw was used to complete the lower osteotomy. The third stage involved completion of the C7-T1 osteotomy with the wire saw, delivery of the tumor specimen en bloc, ventral reconstruction of the spine with a titanium mesh cage, and bilateral thoracoplasty.

CONCLUSION: This is the first case report of a 5-level spondylectomy for en bloc resection of an extensive thoracic chordoma via a bilateral thoractomy without neurologic compromise.

KEY WORDS: Chordoma, En bloc, Malignant, Spine, Spondylectomy, Thoracic, Tumor

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En bloc spondylectomy is often proposed to treat primary malignant tumors of the spine in an effort to improve local control and sometimes offer cure.[1-6] However, successful technical resection may be limited by crucial neighboring anatomy that may prohibit safe tumor resection. Vascular, visceral, or neurologic morbidity may be especially high in the upper thoracic spine where there is a confluence of the heart and great vessels with organs of the mediastinum, all intimately juxtaposed to the spinal canal.

In this technical case report, we discuss the successful en bloc resection of a large thoracic chordoma that involved T1-5 vertebral bodies and the chest wall. The tumor was removed in 1 piece with negative margins, and the patient experienced no neurologic or surgical morbidity. She has survived more than 24 months without local tumor recurrence, distant metastasis, or instrumentation failure. The specific surgical technique used is described for each step of the approach and resection. In addition, important fundamental aspects of this case are discussed, including staging the procedure into an initial posterior approach, a stage 2 right-sided thoractomy, and a stage 3 combined left-sided thoracotomy and posterior approach, spinal osteotomies using wire saws, delivery of the specimen, reconstruction of the spine, and creation of an artificial spinal canal to protect the spinal cord from repetitive trauma by the lungs during normal pulmonary excursion. In the process, we seek to elucidate some of our technical insights for removing large tumors of the upper thoracic spine in an en bloc fashion.

CLINICAL PRESENTATION

A 26-year-old woman presented to the neurosurgery service after a large chest mass was detected on a chest radiograph obtained as part of a routine health screening (Figure 1A). Neurologically she was intact and was generally asymptomatic from this mass. Additional imaging studies were obtained, including a computed tomography scan of the chest abdomen and pelvis (Figure 1B) and magnetic resonance imaging of the cervical, thoracic, and lumbar spine (Figure 1, C, D). These imaging studies demonstrated a solitary mass originating from the T3 vertebral body and extending rostrally and caudally to T1 and T5. The mass extended into both sides of the chest and displaced the mediastinal contents ventrally. No other lesions were detected. She underwent a percutaneous computed tomography–guided biopsy that demonstrated the mass to be consistent with a chordoma. Given the diagnosis, an attempt at en bloc resection was recommended. To achieve an en bloc resection, the operation was planned to be undertaken in 3 stages over 2 separate operating days with a surgical team consisting of neurosurgery, vascular surgery, and plastic surgery. The operations were planned and performed under the control of the senior author (Z.L.G.).

INTERVENTION AND TECHNIQUE

Stage I: Posterior Approach

The patient was positioned prone on the OSI Jackson table (Mizuhosi, San Francisco, California). Her head was secured to the table in a neutral neck position via a Mayfield 3-point headholder. Her cervical thoracic region was cleaned, prepped, and draped in the usual sterile fashion and preoperative antibiotics were given (Figure 2A). An incision was made from C2 to T8. This incision was carried down through the fascia of the trapezius and rhomboid muscles, disconnecting their attachments to the spine on either side. On the left side, the trapezius and the rhomboid muscles were then elevated as a myocutaneous flap and retracted laterally, allowing mobilization of the left scapula. The left scapula was then rotated and elevated, exposing the posterior chest wall from the T1 through T8 ribs. On the right side, the trapezius myocutaneous flap was elevated exposing the paraspinous muscle from T1 to T8. Using a subperiosteal technique, the C2-T8 spinous process, lamina, facets, and transverse processes were exposed. The paraspinous muscles on either side of the spine were elevated such that they could be mobilized either medially or laterally to expose the ribs or spine, respectively (Figure 2B). Self-retaining retractors were placed to maintain exposure.

After the exposure was completed, instrumentation was then placed. Using normal anatomic landmarks for the entry points and trajectories, pedicle screws were placed bilaterally into the pedicles of C2. Lateral mass screws were placed into the lateral masses of C3-6 bilaterally, and pedicle screws were placed in the pedicles of T6-8. Two separate 3.5- to 6.0-mm tapered rods were contoured such that they would span from the C2 through the T8 instrumentation. These were retained for later assembly.

Once the instrumentation was in the proper position, laminectomies were performed from C7 through T6 exposing the thecal sac. The T2-6 nerve roots were identified bilaterally. These nerve roots were doubly ligated with 2-0 silk suture and sectioned proximal to the dorsal root ganglion. The proximal 3 cm of the T6 ribs and rib heads were resected bilaterally. The T6 segmental vessels were identified, and then the plane between the vertebral body and the segmental vessel was dissected open with blunt dissection. A 0.54-mm Tomita saw was then placed ventral to the thecal sac, posterior to the disc space, at the T5-6 level. The ends

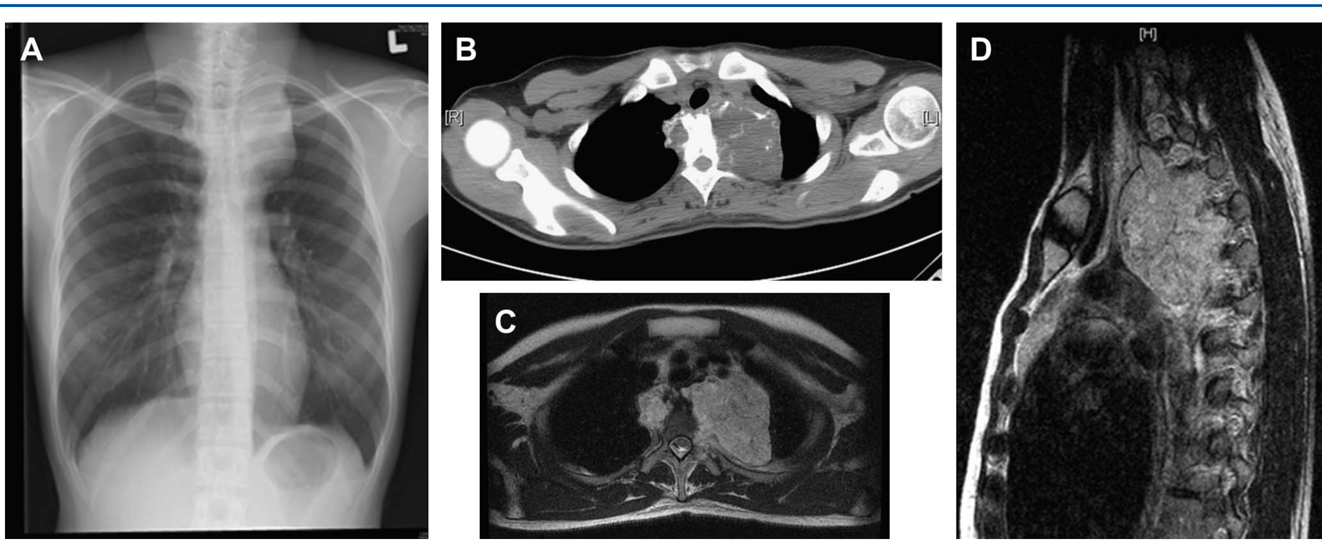

FIGURE 1. *Images of the patient on initial presentation including chest radiograph (**A**), computed tomography of the chest (**B**), magnetic resonance imaging of thoracic spine in axial (**C**) and sagittal (**D**) planes.*

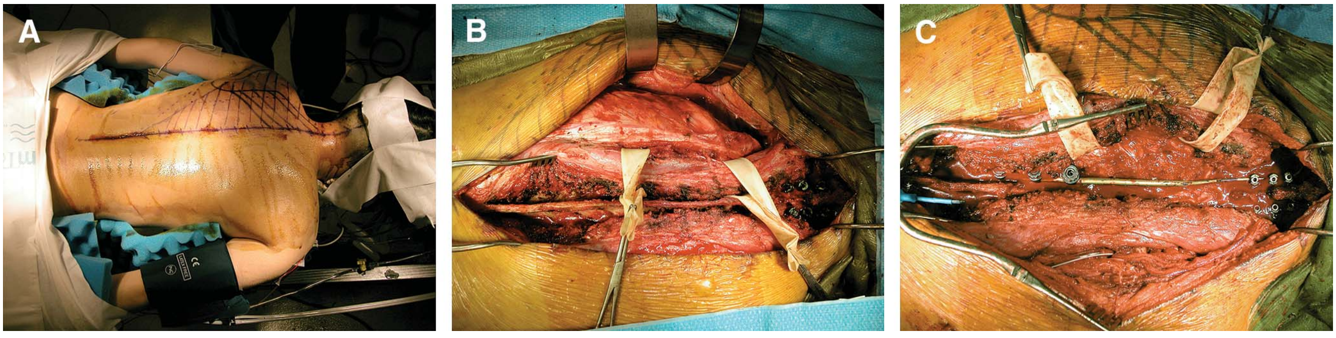

FIGURE 2. *Operative photographs: positioning and planned incision (**A**), soft-tissue exposure (**B**), and placement of posterior instrumentation (**C**).*

of the saw were then placed lateral to the vertebral bodies on either side.

Attention was then turned to preparing the most rostral osteotomy through the C7-T1 disc space. The lateral masses of C7 were bilaterally resected using a high-speed diamond burr giving exposure to the C8 nerve root bilaterally. The C8 nerve root was then dissected laterally to the point where the C8 and T1 nerve roots come together to form the lower trunk of the brachial plexus. The T1 rib head was then resected using a high-speed diamond burr, thus allowing visualization of the T1 nerve as it exits the foramen until where it joins the C8 root at the lower trunk of the brachial plexus. Working ventral to the thecal sac, between the C8 and T1 nerve roots, the C7-T1 disc space was then visualized. Using a no. 15 blade scalpel, a down-angled curette, and a pituitary rongeur, the majority of the C7-T1 disc was removed. Laterally, the posterior two thirds of the annulus was resected with a narrow Leksell rongeur. A 0.81-mm diameter Tomita saw was then placed ventral to the thecal sac at the C7-T1 disc level. The ends of the saw were tucked lateral to the vertebral bodies between the exiting C8 and T1 nerve roots, just proximal to the formation of the lower trunk. The remainder of the osteotomy through the ventral one third of the disc and annulus would be completed during the third stage of the operation.

The T1-5 ribs were then cut lateral to the extent of the tumor on both sides. On the right side, this cut was approximately 3 cm lateral to the transverse process, and on the left, this was approximately 8 cm lateral to the transverse process. A 1-cm portion of rib was removed lateral to these rib cuts at each rib to give better ventral access during the second and third stages of the operation. At this point in the operation, the 2 contoured rods were assembled to the instrumentation and locked in place (Figure 2C). The wound was closed in a multilayered fashion, completing the first stage of the operation.

Stage II: Right-Sided Thoracotomy

The patient was placed in the left lateral decubitus position and prepped and draped in the usual sterile fashion (Figure 3, A-C). A right-sided thoracotomy was fashioned at the T5-6 rib space. This thoracotomy was connected to the previously made cuts in the T2, T3, T4, and T5 ribs. A chest spreader was placed, and the right chest was opened. The right lung was displaced, and the spine was exposed. The pleura was cut along the border between the tumor and the esophagus. The azygous vein was identified and dissected away from the tumor capsule. The plane between the tumor and the esophagus was opened. At this point, all right-sided soft tissues abutting the tumor were mobilized. The Tomita saw that had been placed at T5-6 disc space was retrieved and placed ventral to the disc space, posterior to the aorta and inferior vena cava, and into the left chest. The right thoracotomy was then closed, and the patient was repositioned for the third stage.

Stage III: Left-Sided Thoracotomy

The patient was repositioned in the right lateral decubitus position. Her head was fixed in position via the Mayfield 3-point headholder, and her posterior cervicothoracic region and left chest were cleaned, prepped, and draped. The original incision from C2 to T8 was reopened. The previously prepared latissimus dorsi and rhomboid myocutaneous flap was elevated. This allowed retraction of the entire skin and muscle coverage as well as the scapula over the left chest to be retracted laterally, exposing the left chest wall. A thoracotomy was performed through the left fifth and sixth rib space, and the thoracotomy was extended from the spine to just beyond the rib cuts in ribs 2 to 5. This allowed the left chest to be opened in a "trap door" fashion, giving exposure of the chest from the lower brachial plexus and subclavian artery to below the level of the heart (Figure 4A).

The pleura was incised along the fifth and sixth interspace from the level of the spine medially to beyond the left border of the tumor laterally. This incision was taken rostrally, beyond the apical aspect of the tumor at the first rib. Medially, the pleura was opened at the lateral border of the aorta. Dissection then proceeded posterior to the aorta and ventral to the tumor, maintaining the integrity of the tumor capsule.

The Tomita saw that had been placed at the T5-6 disc level was retrieved from the right chest cavity and pulled into the left chest cavity, ventral to the disc space. Using the 2 ends of the Tomita saw through the left chest, the osteotomy through the T5-6 disc was completed. Then attention was directed to the Tomita saw at the

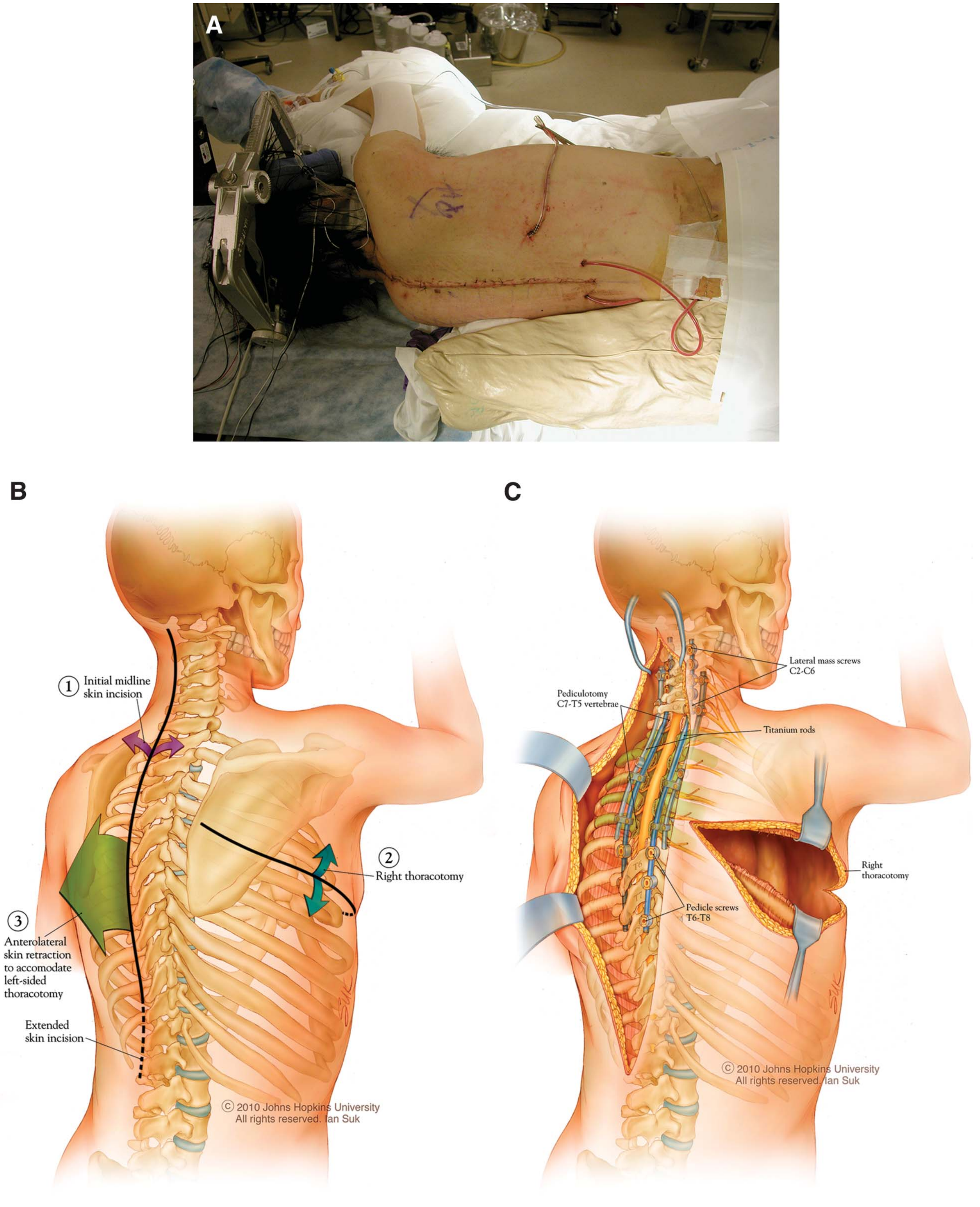

FIGURE 3. *Stage 2 operation involving right-sided thoracotomy. Operative positioning (***A***); posterior and right incisions planned (***B***) and opened (***C***).*

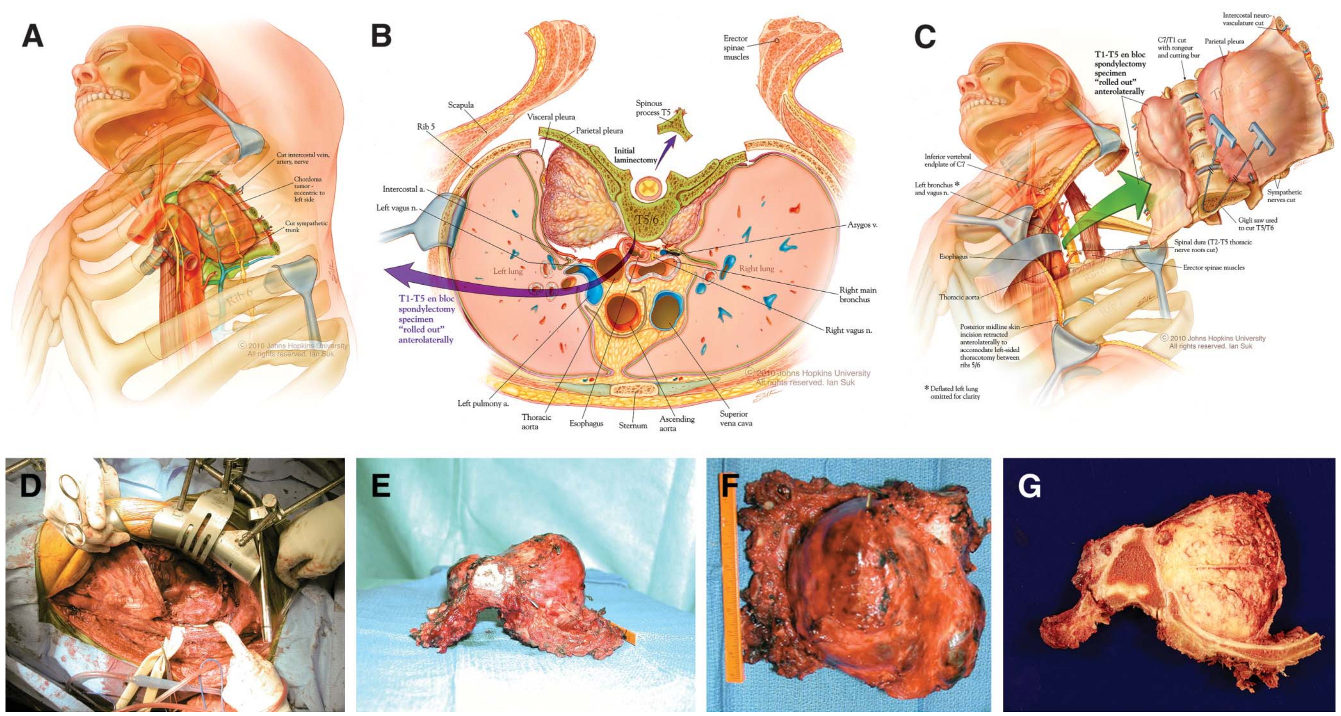

FIGURE 4. *Stage 3 operation involving left-sided thoracotomy, removal of mass, and anterior reconstruction. Left thoractomy with trap-door opening (**A**), removal of tumor (**B**, **C**), thoractomy access with tumor in situ (**D**), removed specimen (**E-G**).*

C7-T1 level. This saw had been placed during the first stage of the operation, and during the second stage, this saw was retrieved from ventral to the C7-T1 disc and was passed to the left chest. At this point, the saw was retrieved, and the lower trunk and C8 and T1 nerve roots were protected. Using the Tomita saw, the rostral osteotomy at C7-T1 was then completed. The specimen was now completely mobile. The specimen was pushed ventrally, away from the spinal cord, and then rotated and delivered out through the left side of the chest (Figure 4, B-G). After the specimen had been delivered, malleable retractors were used to keep the lungs from herniating into the spinal cord during ventilation.

The 5-level spondylectomy defect was reconstructed ventrally by using a contoured titanium mesh cage (Figure 5A). This cage was cut to the length of the defect, but the anterior portion of the cage was left long so it would overlap the C7 and T6 vertebral bodies. The cage was filled with demineralized bone matrix and allograft and placed from C7 to T6 for the anterior reconstruction and arthrodesis. Compression was placed on the cage by compressing on the rod-screw construct posteriorly. A second set of 5.5-mm titanium rods were contoured and placed next to the 3.5- to 6-mm tapered rods. These were connected to the tapered rods using side-by-side domino connectors to reinforce the posterior construct (Figure 5B). Two titanium cables were passed around the rods posteriorly and passed ventrally through the cage. The cables were tightened, thus pulling the ventral portion of the cage against the C7 and T6 vertebral bodies, buttressing the cage. An alloderm graft was then wrapped around the cage and rod construct, reconstituting a spinal canal, and thus creating a barrier to prevent the lungs from herniating into the spinal canal and causing spinal cord compression (Figure 5, C, D). A thoracoplasty was then performed with titanium mesh and methylmethacrylate reconstructing the chest wall defect. The wound was then closed in a multilayered fashion.

Clinical Outcome

The patient was neurologically intact after surgery. Margins of the pathologic specimen were negative for tumor. Postoperatively, chest tubes were managed and removed after a few days. The computed tomography scan showed maintenance of normal cervicothoracic alignment (Figure 6). No perioperative complications occurred, and the patient was sent to inpatient rehabilitation in stable condition, and she has returned to work in her full capacity. The patient is now 25 months out from surgery and has shown no signs of tumor recurrence. In addition, she has had no chest or instrumentation complications.

DISCUSSION

Primary malignant tumors of the spine can lead to neural compression and spinal instability locally, but can also affect

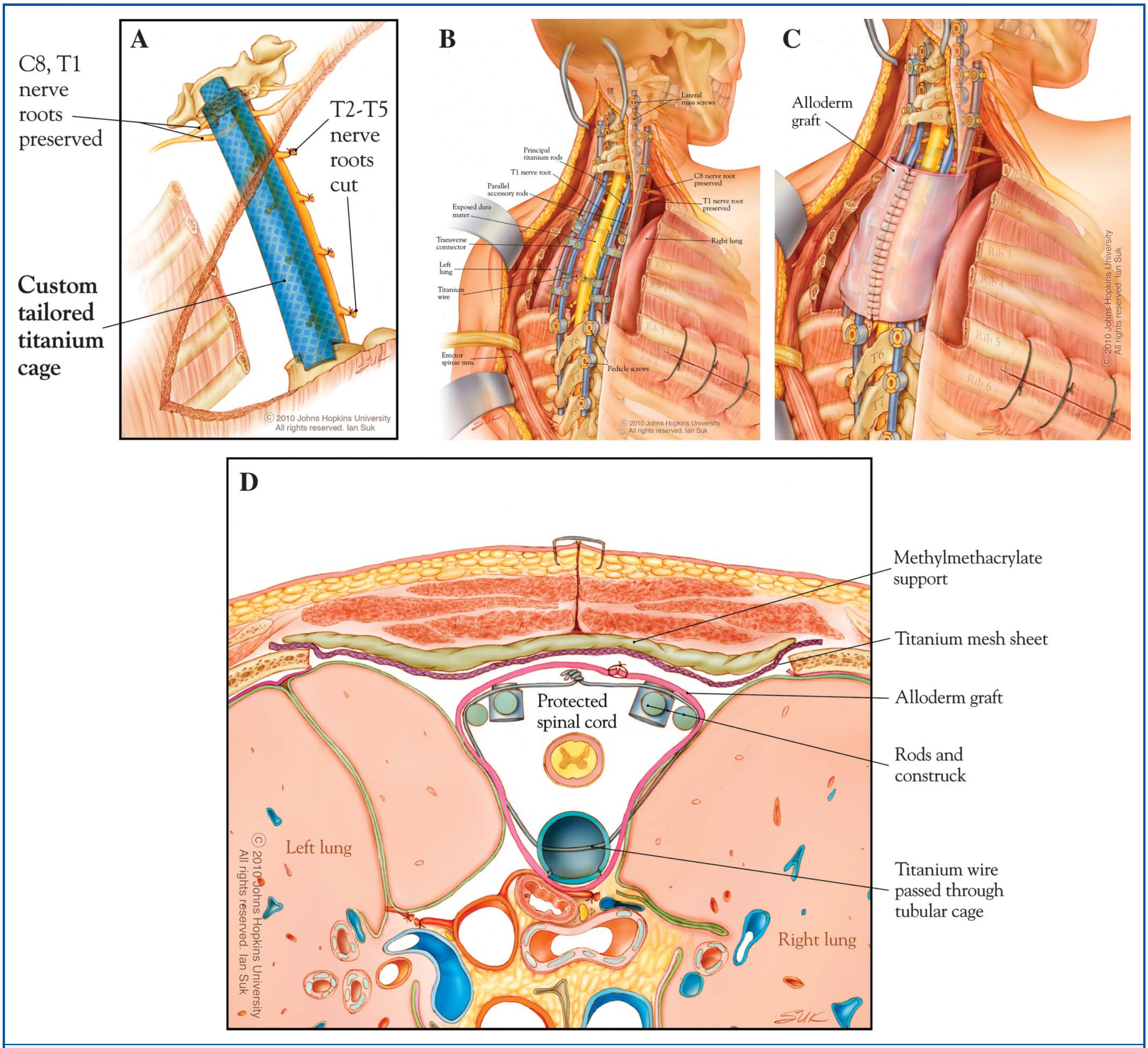

FIGURE 5. *Anterior reconstruction with cage (**A**), posterior instrumentation (**B**), and circumferential reconstruction using alloderm (**C**, **D**).*

survival via aggressive local growth and distant metastasis. Long-term local control and even cure can be attempted with en bloc resection in which negative margins are obtained around all aspects of the tumor.[1-6] The largest experience published regarding treatment of chordomas of the mobile spine was published by Boriani et al,[4] who reviewed their experience over a 50-year period. Although only 1 of the 48 patients included presented with a chordoma involving the upper or mid-thoracic spine (T1-8), they concluded that margin-free en bloc resection, when compared with intralesional resection with or without radiation treatment, is the only treatment that allows for long-term disease-free survival (>50 months on average). In addition, they commented that when chordomas of the mobile spine were deemed unsuitable for en bloc resection and were treated with a combination of surgery, radiation, and medical treatments, early recurrence was the most common outcome. Unfortunately, tumors of the spinal column, unlike tumors of the appendicular skeleton, often involve anatomic structures (eg, spinal cord, great vessels, visceral organs) that cannot be sacrificed with a radical resection. For this reason, complex resection strategies have been used to

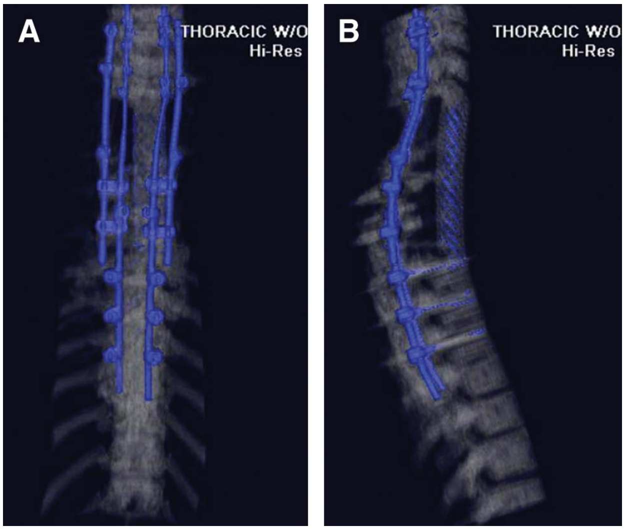

FIGURE 6. *Three-dimensional computed tomography reconstructions of final construct from an anteroposterior view (***A***) and lateral view (***B***).*

remove such tumors en bloc depending on the geometry of the tumor and the neighboring anatomy.

In the mobile spine, multiple authors have attempted to provide anatomic insights that may assist surgeons during en bloc resections. Boriani et al[7] developed a system in which the tumor location in the vertebral body is first localized within 12 radiating segments, and then various osteotomy techniques are suggested to remove the tumor without breaching the tumor capsule, the so-called WBB staging system. Unfortunately, such a system is limited when multiple spinal segments are involved, neighboring regional anatomy is restrictive, and reconstruction strategies are complex. As a result, various case reports have served as the basis for fundamental approaches to complex primary spine tumor resections throughout the mobile spine and sacrum.[8-10] Despite such examples, however, the upper thoracic spine remains an extremely difficult region to approach surgically because of the proximity of the mediastinal contents (eg, trachea, esophagus), the heart, and the great vessels. This technical case report outlines some of the techniques that can be used to remove a large-diameter tumor en bloc that involves multiple spinal segments in the upper thoracic spine.

As is commonly suggested for any en bloc spondylectomy, placement of instrumentation is usually done early in the procedure because of the severely destabilizing effects of removing the entire vertebral body. Although the upper thoracic spine generally has additional stabilizing effects of the rib cage and sternum, the so-called fourth column, screws should be placed, with or without temporary rods, to maintain the stability of the bony spine around the spinal cord during surgical manipulation of the involved segment(s). In this case, instrumentation was extended well above (C2) and below (T8) the area to be resected. The Mayfield head fixation device in conjunction with the rib cage allowed decompression to proceed without temporary rods.

After placement of instrumentation, resection of the posterior elements was conducted to provide initial release of the spinal cord. In this case, release involved laminectomies, facectomies, rib resection, and discectomies. However, complete osteotomy of the diseased spinal segments from the parent spine could not be accomplished safely from posteriorly because of the large extent of tumor into the thoracic cavity. Therefore, Tomita wire saws were placed ventral to the thecal sac so that they could be retrieved anteriorly and used to complete the cuts. Importantly, during these latter cuts, the saws could be pulled so that tension was directed away from the spinal canal, theoretically reducing the likelihood of spinal cord injury. Rigid stabilization was then obtained with screw-rod fixation. In subsequent stages, bilateral thoracotomies allowed dissection of the tumor from the surrounding soft tissues, delivery of the tumor, and anterior spinal reconstruction.

Important technical aspects of this operation are essential for successful removal of the tumor in 1 piece without resultant neurologic decline. First, we consistently use motor evoked potential and somatosensory evoked potential monitoring in all aspects of the operation to aid us in preventing unintentional compression, distraction, or twisting of the spinal cord during spinal manipulation. If signals become abnormal, we run through a checklist to determine the cause of the change in potentials. Immediately we attempt to reverse any manipulation of the spinal column or cord recently done (eg, retraction, distraction, hardware placement). If nothing is easily identified, we check with anesthesia and monitoring personnel to identify any abnormalities that may interfere with ideal monitoring (eg, anesthesia, technical aspects of monitoring). In general, if we are concerned that the changes represent potential injury to the spinal cord, we consider halting the surgery and waking the patient, with the plan of returning to attempt the en bloc resection later when the patient is stable. We do not recommend changing the type of oncologic procedure based on the monitoring, such that a planned en bloc resection is converted to an intralesional resection based on monitoring abnormalities.

A second important component of a successful resection involves sectioning of spinal roots to allow for circumferential en bloc resection in the thoracic spine. Sectioning of such roots can be done in the thoracic spine without clinically significant weakness or sensory loss. However, there is concern that ligation of multiple segmental arteries in succession or ligation of the artery of Adamkiewicz may lead to spinal cord ischemia. Tomita et al[11] showed that bilateral ligation of greater than 4 segmental arteries in a canine animal model increases the chance for spinal cord ischemia. Performing a preoperative spinal angiogram is an option to help understand vascular supply to the tumor and the spinal cord, especially for tumors that are expected to be well vascularized (eg, aneurysmal bone cysts, renal cell carcinoma). In such patients, preoperative embolization may help to decrease intraoperative blood loss. However, there are currently no reliable factors obtained from a spinal angiogram that have been shown to be predictive of spinal cord ischemia after en bloc spinal tumor resection.

Third, thoracic spondylectomies with pleura resection place the spinal cord at risk of dynamic compression from the lungs, which may distort the spinal cord with each respiration. For this reason, reconstruction of the spinal canal, not just the spinal column, may be necessary in such cases. It is for this reason that an alloderm patch was placed around the instrumentation, effectively recreating the spinal canal. Fourth, because of the large amount of shear forces acting at the ends of the cage strut, we recommend that such cages do not merely engage in the neighboring vertebral bodies via a mortised approach, but rather that the cage is buttressed into the neighboring bone. In this way, the cage acts both like a strut and an anterior stabilizing plate, potentially reducing the risk of bone-instrumentation interface failure.

This type of surgery must only be attempted after careful consideration of the surgical risks to the patient. Discussion of all possible risks in this setting is beyond the scope of this discussion, but it must be noted that such a procedure should only be attempted by a multidisciplinary team, including, but not limited to, neurosurgery, thoracic surgery, plastic surgery, and cardiac anesthesia. In addition, staging the procedure likely limits the cumulative risk to the patient by effectively segmenting a larger surgery into multiple smaller ones. Such staging has been shown to increase surgical safety in other surgeries that would normally last longer than 12 hours.[12]

CONCLUSION

We have illustrated successful en bloc resection of a chordoma of the thoracic spine involving 5 levels without neurologic compromise. Long-term follow-up has shown no tumor recurrence of instrumentation failure. For optimal outcomes after such procedures, we recommend a multidisciplinary approach and a carefully planned staged procedure.

Disclosures

Dr Sciubba receives honorarium from Depuy Spine Ziya Gokaslan, has stock ownership in Spinal Kinetics (10 000 shares, 5%) and U.S. Spinal Tech (20 000 shares, 5%), and has speaking and/or teaching arrangements with Stryker. Dr Witham has speaking and/or teaching arrangements with Stryker Spine and receives research support (investigator salary) from Integra LifeSciences. Dr Bydon receives research support from Depuy Spine. Dr Wolinsky has speaking and/or teaching arrangements with Stryker ENT. The other authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENTS

Dan Sciubba and the immeasurably talented group at Johns Hopkins very elegantly present the technical nuances of a 5-level en bloc resection of an upper thoracic chordoma involving the T1-5 vertebral bodies and bilateral chest wall with extension to the mediastinum. The surgery was performed as a 3-staged procedure and resulted in no neurologic morbidity, no significant surgical complications, and no evidence of recurrence at 24 months. The expertise brought to bear on this complicated problem is truly an operative tour de force. I would add only 1 technical consideration: the potential use of preoperative angiography to identify the risk of taking a major radicuolomedullary feeder before 5-level bilateral rhizotomies.

The only remaining question is whether this extensive surgery will ultimately cure the patient or even result in long-term local tumor control. Although margins were reported as negative, the resection appeared to be marginal based on anatomic constraints of mediastinal and pleural involvement. The question with a chordoma remains whether a marginal resection is adequate because these tumors frequently demonstrate local or marginal recurrences after en bloc resection, even with negative margins. The question, particularly for large multicompartmental tumors, is whether surgery is ever truly curative or whether we should be using neoadjuvant radiation in an attempt to not only treat the gross tumor volume, but also the at-risk margins. This may lessen the risk of marginal recurrence and, if one can create significant tumor necrosis, may allow one to perform intralesional en bloc or possibly even curettage resection (with less chance of local recurrence or seeding). Delaney et al[1] recently reported the use of neoadjuvant radiation in a phase 2 clinical trial of primary spine tumors including chordomas and sarcomas. The treatment paradigm was 50.4 Gy RBE in 1.8 Gy per fraction of photon therapy delivered preoperatively with surgery at 5 weeks post-radiation. Postoperatively patients were given 19.8 Gy RBE in 1.8 Gy RBE per fraction to a total dose of 70.2 RBE for negative margins or 77.4 Gy RBE for gross residual disease. The radiation dose was delivered to not only the gross volume identified, but to areas of suspected microscopic disease, such as the piriformis muscles for sacral disease. Radiation was well tolerated with local disease control demonstrated in 78% at 5-year follow-up. Wu et al[2] reported a case report of a patient who underwent definitive radiation 24-Gy single fraction to an L4 chordoma. Radiographic changes consistent with progression prompted resection at 4 months and demonstrated 99% necrosis of the tumor.

Although surgical techniques, such as the one demonstrated in this case report, are important in the resection of a chordoma, anatomic constraints and microscopic disease outside the gross tumor volume may limit the usefulness of surgery as the sole treatment for a chordoma. More work and long-term follow-up are critically important to assess the role of neoadjuvant therapy, but may represent a promising advance in the treatment of this disease.

Mark H. Bilsky
New York, New York

1. Delaney TF, Liebsch NJ, Pedlow FX, et al. Phase II study of high-dose photon/proton radiotherapy in the management of spine sarcomas. *Int J Radiat Oncol Biol Phys*. 2009;74(3):732-739.
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This is an excellent and extremely well documented article of surgical technique. It should be considered at the same level of the unsurpassed articles of surgical oncology of spinal tumors written by Bertil Stener[1] on his experience that started more than 40 years ago.

Today, in the era of the minimally invasive approach, we are faced with the dilemma: is it better to treat a chordoma with en bloc resection, possibly including the sacrifice of important anatomic elements, or with a less aggressive intralesional excision combined with radiation and drugs in a multidisciplinar approach? Is it better to look at the final prognosis or be more concerned on the immediate evolution? In other words, is it worthwhile to expose the patient to such an aggressive and morbid[2] surgery?

Based on my personal experience of more than 20 years of surgical activity on chordomas, confirmed by a recent systematic literature review, I think that the answer is yes.

The data reported in our article published some years ago reporting on 58 cases of chordomas[3] stressed that en bloc resection with marginal or wide margin is associated with long-term local and systemic control. Three recurrences occurred at 5, 7, and 8 years after surgery, making all articles on chordomas with follow-up shorter than 2 or 3 years very limited in their usefulness in predicting outcome. These data were confirmed by a more recent systematic review.[4]

Any other different approach, including variable combinations of less aggressive surgery, radiotherapy, medical treatment, sometimes required by anatomic constraints or by the extension of the tumor, will keep the patient burdened by the need for frequent controls, of different treatments, with anxious waiting for "something new" to remove the residual and progressive tumor that can sometimes produce very painful courses. The tumor remains mostly in site, latent, or slowly growing, ending in painful huge masses that are extremely difficult to treat. Everyone who has experience in operating on a recurrent chordoma knows how difficult it is to excise because it is infiltrating surrounding tissue or adherent or even permeating the dura.

In every spinal tumor center, several of such patients are under observation. They report severe, sometimes untreatable, pain, asking for heroic surgery rather than going on with such a very poor quality of life.

The patient should be warned that less aggressive approaches can result in such a difficult condition, and en bloc resection should be always considered and discussed, if feasible, because it is the treatment that will keep the patient free of any trouble for a very long time, even forever.

I strongly believe that the first treatment affects the final prognosis. Once the first treatment is unable to control the chordoma, no treatment to date is able to stop the slow but relentless evolution.

En bloc resection, even in such a difficult cases like the one here presented, should be considered the gold standard in the treatment of a chordoma until some different treatment or combination of treatments will prove to be more effective in terms of local recurrence, quality of life, pain, number of procedures or treatments, survival at the same long-term follow-up.

Stefano Boriani
Bologna, Italy

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