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Original Article

Multicompartmental Primary Spinal Extramedullary Tumors: Value of an Interdisciplinary Approach

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Abstract

Background: Primary extramedullary tumors involving multiple compartments around the spine are a technically demanding group of tumors whose extent traverses beyond the normal confines of those anatomical regions which fall in the common domain of neurosurgeons. In the following series, we present 12 patients who were diagnosed with primary spinal extramedullary tumors with multicompartmental extension, and whose surgical management was facilitated by a combined multidisciplinary approach involving surgeons of other superspecialties. This multidisciplinary assistance from the inception to the culmination of surgical management helped in achieving a better surgical removal, thereby resulting in better surgical outcomes. **Materials and Methods:** Twelve consecutive patients who fulfilled the inclusion criteria in the 5-year period (January 2010 to January 2015) were included in the series based on the radiological and pathological characteristics of the tumor. Depending on the site of the spine involved by the lesion, radiologists and surgeons were involved from the planning phase of the surgical management, and their assistance in procedures such as preoperative embolization/ureteric stenting was sought whenever was deemed necessary. The extent of resection and total blood loss was recorded meticulously. Regular follow-up (3, 6, and 12 months and 2 and 5 years) of the patients was done after the initial follow-up at 6 weeks and their disability scores were recorded. **Results:** Of the 12 cases (6 males and 6 females), sacrum was the most common location of the tumors (6). Histopathologically, giant-cell tumors, schwannomas, and chondrosarcomas (3 each) were most common followed by Ewing's sarcoma (2) and malignant peripheral nerve sheath tumor (1). Eight patients had functional status of McCormick scale 1 and two patients had a functional status of 2. One patient was lost to follow-up and one patient died during surgery. **Conclusion:** Judicious involvement of access surgeons and adjunct therapies along with careful preoperative planning can help in improving surgical outcome in multicompartmental spinal tumors.

Keywords: *Access surgeon, adjunctive therapy, extramedullary spinal tumors, giant spinal tumors, McCormick grade, multicompartmental, surgery*

Introduction

A special subset of primary extramedullary spinal tumors pose a particular challenge by virtue of their size, local spread in multiple compartments around the spine and their proximity to vital neurovascular structures. As these tumors extend beyond the limits of well-defined natural fascial planes, their anatomical delineation by a single conventional surgical approach is often not feasible. Moreover, their excision mandates a multidisciplinary team, each of whom plays an important part in a particular aspect of its surgical management. In this series, we focus on the surgical excision of 12 cases of giant multicompartmental primary spinal extramedullary tumors and highlight the contribution by different subspecialties in achieving their successful surgical extirpation and better outcome.

Materials and Methods

In this prospective, descriptive series of 5-year duration (from January 2010 to January 2015), twelve consecutive patients were included in the series after fulfilling the inclusion criteria based on radiological and pathological characteristics of the tumor. Depending on the site of the spine involved by the lesion, radiologists and surgeons belonging to other superspecialties were involved from the planning phase of the surgical management, and their assistance in procedures such as preoperative embolization/ureteric stenting (UTS) was sought whenever was

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deemed necessary. The extent of resection (ER) and total blood loss was recorded meticulously. Regular follow-up (3, 6, and 12 months and 2 and 5 years) of the patients was done after the initial follow-up at 6 weeks and their disability scores were recorded.

The inclusion criteria are as follows:

Primary spinal extramedullary tumors of size more than 5 cm × 5 cm, multicompartmental involvement, nonmetastatic lesion on fine-needle aspiration cytology (FNAC)/trucut biopsy, access surgeon involvement, recurrent/residual lesions operated elsewhere but fulfilling the above criteria.

Radiological protocol

The size, extent, and multicompartmental nature was defined by a senior radiologist at our institute with the help of multiplanar plain and contrast magnetic resonance imaging (MRI).

Trucut biopsy

The trucut biopsy/FNAC was done at our institute and those cases who were operated outside/biopsied outside were reviewed at our institute and reconfirmed.

Access surgeons

Access surgeons were those who belong to nonneurosurgical specialties whose inputs were taken in preoperative radiology study and/or preoperative planning and/or intra- and post-operative management of the patients.

- Cervical cases – neuro-otologists and gastrosurgeons
- Cervico dorsal – cardiothoracic and vascular surgeons (CTVS), neuro-otologists, and gastrosurgeons
- Dorsal – CTVS surgeons,
- Dorsolumbar – CTVS surgeons and urologists
- Lumbar – urologists and gastrosurgeons
- Lumbosacral and sacral – urologists and gastrosurgeons.

Other supportive investigations

MR angiography/digital subtraction angiography/therapeutically embolization; computed tomography (CT) scan with bone windows; intravenous pyelogram, UTS; and CT thorax and/or bone scan were the additional investigations performed to rule out secondary spread of aggressive tumors.

Disability assessment and follow-up protocol

Preoperative neurological status was recorded and their disability was graded according to the McCormick grading scheme.[1] Postoperative contrast-enhanced MRI at 6 weeks was used to confirm the extent of excision. The extent of excision was labeled as near-total (NT) if no visible tumor residue was present after excision, subtotal if some part of the tumor was left behind (>50% tumor removed), and partial (if <50% removed). The minimal follow-up with radiology was done at the end of 6 weeks.

Results

Demographic, clinical profile, and tumor characteristics

The demographic, clinical profile, and tumor characteristics are shown in Table 1. Twelve patients were included in the present series which consisted of equal number of males and females (six each). The patients' mean age was 34.8 years (range from 7 to 51 years). Local pain was the earliest clinical symptom experienced by 11 patients (91.66%), and the mean duration between appearance of pain and presentation was 10.9 months (range 2–24 months). Neurological deficits (paraplegia, foot drop, and Kyphoscoliosis) were present in a minority of patients (n = 3, 25%). Motor and sensory symptoms were of equal distribution in the patients (n = 8). Five of the patients (41.2%) suffered from bladder/bowel involvement. Six patients had vascular involvement (internal iliac vessels) bilaterally. Sacroiliac joint was involved in five cases bilaterally. The system of Enneking *et al*.[2] was used for the classification of giant-cell tumors (GCTs).

Tumor characteristics

The most common anatomical location of the tumors was at the sacrum (n = 6, 50%) followed by the dorsal vertebrae (n = 4, 33.3%). One patient had cervicodorsal involvement whereas the other patients had a purely lumbar spinal mass. The mean tumor size was 5 cm × 10.9 cm. MRI was the preferred mode of radiological investigation as compared to CT (10 vs. 6). Six patients (50%) had heterointensities on T1- and T2-Weighted images with heterogeneous contrast enhancement on MRI suggestive of cystic/necrotic changes within the tumor. CT was used as an additional modality for better delineation of the bony involvement in six patients. Six patients had tumor extension into the sacroiliac joint, four patients had posterior mediastinal involvement, and eight patients had retroperitoneal extension. Compression over the retroperitoneal structures and urinary bladder was observed in nine patients.

Histopathology and surgical approaches

The various histopathologies, preoperative interventions, details of the surgical procedure, ER, and blood loss are shown in Table 2. Nine patients underwent NT resection of the tumor; three patients had subtotal resection.

Outcome and follow-up

The outcome and follow-up of patients are summarized in Table 3. All patients were followed up at 3, 6, and 12 months and 2 and 5 years, except for one patient. In the follow-up period, the functional status was assessed on the McCormick grading scale. There was one mortality.

Discussion

Etiopathogenesis of late presentation and giant size

Primary tumors of the spine both benign and malignant are usually asymptomatic and go unnoticed until they attain

Table 1: Demographic, clinical profile, and tumor characteristics

Patient	Age/sex	Spinal level	Compartments	Size (cm)	DOS (months)	Neurological symptoms/deficit
1	22/female	S1-S3 + sacroiliac joint	RUP + VUP + RVP + IIV B/L + SC + PVS	6×8×6	24	Paraplegia
2	35/female	S1-S3 + sacroiliac joint	RUP + VUP + RVP + IIV B/L + SC + PVS	7×9×7	12	Lp + incontinence
3	30/male	S1-S3 + sacroiliac joint	RVP + IIV B/L + SC + PVS	5×6×8	6	Lp + incontinence
4	21/male	D12-L4	Left kidney and ureter + SC	8×10×8	-	Kyphoscoliosis
5	42/male	S1-S3 + sacroiliac joint	RVP + IIV B/L	6×7×9	15	Lp + Incontinence
6	16/male	C3-D2	PVS + PRG + PSS	5×6×9	4	Lp
7	18/female	L4-L5	PSS + spinous process	5×5.5×8	3	Right foot drop
8	51/male	L5-S2	RVP + IIV B/L + PVS	8×10×8	12	Lp + incontinence
9	25/female	D3	PSS + SC + PVS	6×7×10	12	Lp
10	7/male	D8-D9	SC + PSS + PVS	5×7×9	2	Lp
11	31/female	S1-Co + sacroiliac joint	RUP + VUP + RVP + IIV B/L + SC + PVS	8×10×10	18	Lp + incontinence
12	49/female	D3-D4	PSS + PVS + compression on the aorta	10×10×10	12	Lp

RUP – Rectouterine pouch; VUP – Vesicouterine pouch; RVP – Rectovesical space; IIV – Internal iliac vessel; PVS – Paravertebral space; PSS – Paraspinal space; SC – Spinal canal; PRG – Parapharyngeal space; DOS – Duration of symptom; Lp – Local pain; B/L – Bilateral

Table 2: Histopathology and surgical approaches

Patient number	Histopathology	Preoperative intervention	Conventional approach	Contribution by other surgeons	Details of procedure conducted	ER	Blood loss
1	GCT sacrum	-	Ds	Urologist	Intralesional decompression	NT	4500
2	GCT sacrum	B/L UTS	Combined dorsal and ventral approach	Urologist	*En bloc* resection	NT	1800
3	Chondrosarcoma	B/L UTS	Ds	Urologist	Intralesional decompression	NT	1500
4	Neurofibroma	EMB + B/L UTS	Combined dorsal and ventral approach	Urologist/ radiologist	*En bloc* resection	NT	1500
5	Chondrosarcoma	B/L UTS	Ventral approach	Urologist	Intralesional decompression	ST	2000
6	Ewing's sarcoma	-	Ventral approach	ENT surgeon	Intralesional decompression	NT	500
7	Ewing's sarcoma	-	Dorsal approach	Urologist	*En bloc* resection	NT	500
8	GCT sacrum	EMB + B/L UTS	Ventral	Urologist	Intralesional decompression	ST	800
9	Neurofibroma	-	D3 Pt	CTVS surgeon	*En bloc* resection	NT	500
10	MPNST	-	D8 Pt	CTVS surgeon	*En bloc* resection	NT	300
11	Chondrosarcoma	B/L UTS	Dorsal	Urologist	Intralesional decompression	ST	1500
			Dorsal	Urologist	Intralesional decompression	ST	1500
12	Neurofibroma	-	D3 Pt (right)	CTVS surgeon	Intralesional decompression	ST	1000
			D3 Pt (left)	CTVS surgeon	*En bloc* resection	NT	1000

MPNST – Malignant peripheral nerve sheath tumor; GCT – Giant-cell tumor; ER – Extent of resection; NT – Near total; ST – Subtotal; PT – Partial; UTS – Ureteric stenting; EMB – Embolization; Ds – Dorsal sacrectomy; Pt – Posterolateral thoracotomy; CTVS – Cardiothoracic and vascular surgery; ENT – Ear, nose, and throat; B/L – Bilateral

large dimensions and produce mass effect or invade the adjacent structures. Multicompartmental nature and their large size could be attributed to extracanalicular origin, large anatomical spaces around the lesion[3] and origin from relatively less mobile segment of spine (cervicodorsal, dorsal spine, and sacrum), and proliferative potency.

Role of trucut biopsy

The predefinitive surgery biopsy was done in all the cases at our institute. Most needle biopsies are performed under fluoroscopic or CT control[4] with accuracy ranging from 80% to 90%, but it is nondiagnostic in 25% of patients.[5] Trucut biopsy proved useful for the categorization of benign and malignant lesions preoperatively and then relevant investigations were done to rule out metastasis. In cases of preoperative biopsy proven GCT (cases 1, 2, and 8), in addition, CT chest was done to rule out benign lung metastasis.[6] There was no disparity in preoperative biopsy and postoperative biopsy in any of our cases.

Operative strategies and role of access surgeon and adjuvant treatment

Cervical/cervicodorsal lesions

In cervical lesions, special considerations such as use of fiberoptic intubation, flexometallic endotracheal tube, preoperative discussion with neuro-otologist/gastrosurgeons to avoid injury to esophagus and CTVS surgeons to address the upper dorsal tumor.

Table 3: Outcome and follow-up

Patient	Preoperative grade	Postoperative grade	Follow-up grade at 3, 6, 12, 24, 60 months					Adjuvant treatment	Recurrence
1	4	Death due to exsanguinating hemorrhage and shock	-	-	-			-	-
2	1	1	1	1	1	1	1	RT	-
3	1	2	1	1	1	1	2	RT	-
4	2	3	2	2	2	2	2	-	-
5	1	1	1	1	1	3	4	RT	-
6	2	3	2	2	2	2	2	RT + chemotherapy	-
7	2	1	1	1	1	1	1	RT + chemotherapy	-
8	1	1	1	1	1	2	2	RT	-
9	1	1	1	1	1	1	1	-	-
10	1	1	1	1	1	1	1	RT	-
11	1	1	2	4	*	*	*	RT	Present
12	1	1	1	1	1	1	1	-	-

*Lost to follow-up. RT – Radiotherapy

Dorsal spine tumors

Prior discussion with the CTVS surgeons/cardiac anesthetist, positioning, need for single-lung ventilation, preoperative embolization, appropriate corridor, and also single or staged surgeries were planned.

Dorsolumbar/lumbar tumors

special considerations such as positioning, preoperative stenting of ureter, access to the vascular pedicle, preoperative embolization, gastro surgery, and urology opinion on the surgical approach and anatomical delineation due to the large size of these tumors.

Sacral tumors

Required special attention on the relation with the ureter, access to the vascular pedicle, adjacent bladder, bowel and involved pelvic splanchnic nerves, and the sacroiliac joint.

Adjunct procedures

Embolization

Preoperative embolization was done in two patients (cases 4 and 8) that decreased tumor blush by 10%–15% in both cases [case 4/ Figure 1a-e]. Following embolization, surgery was done within the next 72 h in both cases.

Ureteric stenting

Due to the disturbed anatomical planes in large tumors with retroperitoneal and pelvic extension, the possibility of ureteric injury seeks consideration in the preoperative planning. Bilateral UTS was done at least 24 h before the surgery in (cases 1, 2, 3, 5, 8, and 11) as the position for surgery would not allow intraoperative stenting. In our series, no ureteric injury was encountered and the stent was removed within 48 h of surgery.

In case 8, bladder insufflation with saline, Foley placement in rectum and intraoperative pelvic nerve monitoring was done along with spinal stabilization (L4-S1 sacral screw fixation with connector) [Figure 2e].

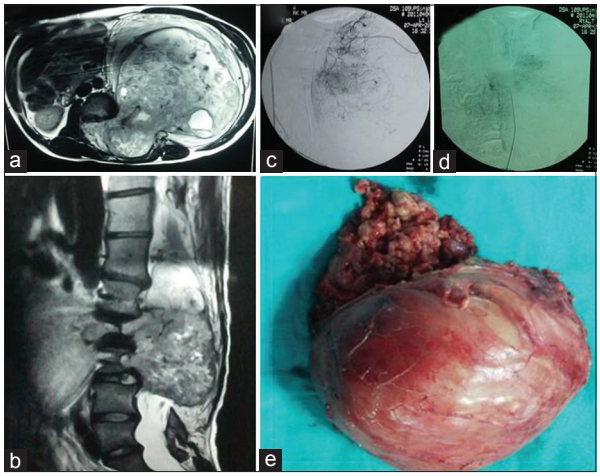

Figure 1: (a) Axial image showing heterointense lesion with foraminal extension. (b) Sagittal image showing lesion extending from d12 to dl4 level with extension towards the left side. (c) Digital subtraction angiography showing tumor blush after injection form the anterior spinal artery at d12 level. (d) Digital subtraction angiography showing decrease in the tumor blush after onyx injection. (e) Complete excised specimen

Evaluation of cases with respect to histopathological examination

Giant-cell tumor: Cases 1/2/8

All cases of GCT in our series were Grade 3 of the Enneking *et al.*'s surgical staging system.[2] Recurrence rate of spinal GCT following *en bloc* surgical excision is in the range of 11%–50%.[7,8] However, in sacrum, this cannot be applied as *en bloc* resection results in significant morbidity. In sacral lesions, due to the extensive disease at presentation and inability to carry out *en bloc* resection without neurological deficit, intralesional resection is planned which has a recurrence rate in the range of 0%–71% [case 8, Figure 2a-c]. Hence, in sacral lesions when *en bloc* resections are prohibited due to the risk

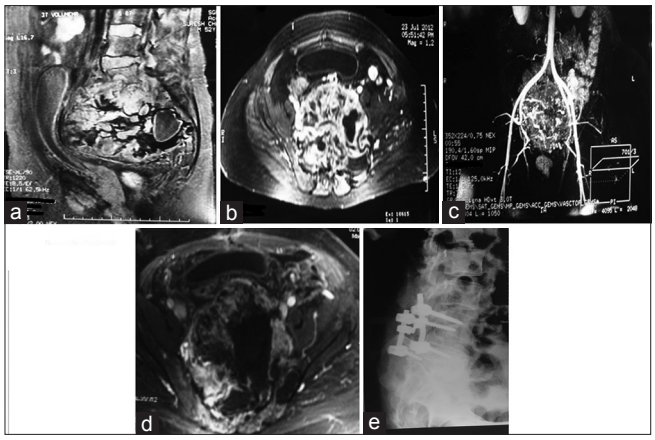

Figure 2: (a) Sagittal image showing heterogeneous contrast enhancing lesion involving the sacrum and extension into pelvis. (b) Coronal image showing heterogeneous contrast enhancing lesion involving the sacrum and extension into sacroiliac joint. (c) Magnetic resonance angiography showing extensive involvement of the bilateral iliac vessels. (d) Postoperative image showing subtotal resection of the lesion. (e) Postoperative lateral X-ray shows good cortical purchase of the sacral screws (L4-S1 pedicle screw fixation)

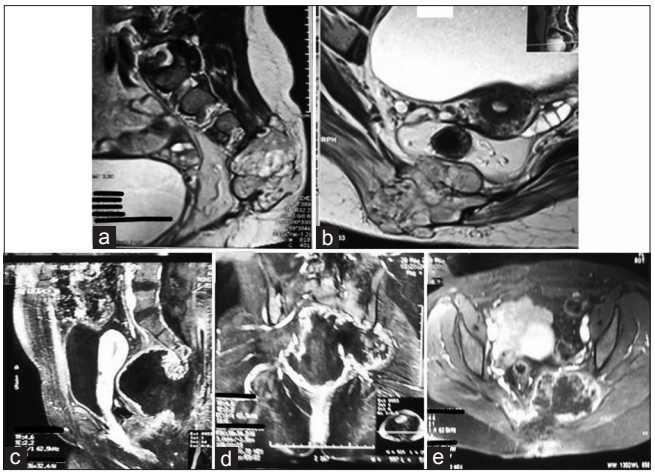

Figure 3: (a) T1 sagittal showing destruction of the sacrum and extending to rectouterine pouch, paravertebral space. (b) Tumor extension into the paraspinal space, paravertebral space. (c) Postoperative sagittal contrast image showing subtotal resection of the lesion. (d) Postoperative coronal contrast image showing subtotal resection of the lesion. (e) Recurrence of the tumor and extending anteriorly to the pelvis

of postoperative morbidity, preoperative embolization followed by intraregional resections has been used with varied success for adequate control of local burden followed by adjuvant radiation therapy. The surgical corridors employed proved to have a significant influence on the outcome. With preoperative planning, the blood loss drastically reduced (cases 2 and 3 – 1800 and 800 ml, respectively) and hemostasis was easily achieved. No stabilization was required due to lack of instability.

Postoperative scan showed no evidence of recurrence in both cases [Figure 2d].

Chondrosarcoma: Cases 3/5/11

In our series, three patients complained of perianal numbness, bladder, and bowel disturbances. The standard regimen is complete *en bloc* resection to prevent recurrence as they are generally resistant to conventional radiotherapy and chemotherapy. However, *en bloc* resection often requires spinal reconstruction involving a multidisciplinary team, and when *en bloc* resection is not possible, partial removal followed by radiotherapy may provide palliation of pain and improve neurological deficits.[9-13] Since the tumor was histopathologically of low grade in case 3, preoperative planning for intralesional curettage was done and about 60%–70% of the tumor was removed followed by adjuvant radiotherapy. In case 5, bilateral UTS was done preoperatively and then a two-staged surgery was performed. First stage was by the posterior approach and the 2nd stage by the left flank incision and anteromedial approach. NT excision was done and there was a loss of 2000 ml of blood [case 11, Figure 3a and b]. In case 11, posterior midline approach was undertaken, NT excision was done with 1500 ml blood loss and subjected to radiotherapy [Figure 3c and d]. At the end of 3-month follow-up, patient's grade deteriorated from Grade 1–2 and showed local recurrence [Figure 3e]. Hence, the patient was reoperated but at the end of next 3 months suffered massive recurrence. This time, the patient was offered *en bloc* excision and stabilization but patient deferred.

Ewing's sarcoma: Cases 6/7

We encountered two adolescent patients (1 male/1 female) of vertebral ES, one in the cervical and the other in the lumbar spine. Radiological signs of ES are late to appear, the most common finding is the lytic bone destruction involving the vertebrae.[14] The lytic changes vary from focal to complete flattening of the vertebral body (vertebra plana).[15] Our patients had lytic lesions causing destruction of mainly the posterior elements and the vertebral body with paraspinal and intraspinal extension. The cervical disease was resected *en bloc* with the help of combined supra and the infraclavicular approach. The lumbar mass was tackled entirely by the neurosurgical team and *en bloc* resection was done in both cases. Postoperative MRI revealed no residual or recurrent lesion.

Neurofibroma: Cases 4/9/12

Giant intrathoracic neurofibromas are very rare tumors with only two case reports found in literature.[16] Surgery was planned for staged excision in case 12 and single stage in case 9. Two-staged surgery was done after a period of 3 months in case 12. Both patients had significant relief of their complaints and uneventful recovery and are now in routine follow-up.

Case 4 was a giant neurofibroma in a 21-year-old male who presented with weakness and thinning of the lower limbs (right > left), with radiculopathy in the right L5 distribution and sensory loss below T10 of 9-month duration [Figure 1a and b].

Intraoperatively, after taking control of the vascular pedicle around the tumor, it was removed in two piecemeal. The main bulk of the tumor and the residual tumor passing through the intervertebral foramina were excised after preserving the nerve roots, and the blood loss was 300 ml [Figure 1e].

Life-threatening intraoperative bleeding before complete excision of the mass has been previously reported.[17] Such complications can be avoided with judicious preoperative/intraoperative planning with access surgeons. Diathermy is of limited use in such cases as the tissue is very friable.[18] A number of authors have reported significant blood loss during surgery requiring high-volume transfusion; several management strategies, including argon beam coagulation, have also been suggested.[18]

Malignant peripheral nerve sheath tumors: Case 10

Malignant peripheral nerve sheath tumor is a rare variety of soft-tissue sarcoma of ectomesenchymal origin.[19,20] Around 5%–42% have association with neurofibromatosis Type 1.[21-27] Our patient was a 7-year-old male child who was operated in 2006, was referred to radiotherapy but was lost to follow-up. In January 2012, the patient presented with a dumbbell-shaped extramedullary/intradural lesion at the level of D7/D8 vertebral body with paravertebral extension across the left neural foramina of D6 into the adjacent posterior mediastinum [Figure 4a and b]. D6 left posterolateral thoracotomy with NT excision was done with the blood loss of 300 ml [Figure 4e]. Postoperative scans revealed complete tumor removal [Figure 4c and d]. The patient received radiotherapy for 6 weeks.

Limitations

We understand that the study is limited due to its small sample size, varied histopathologies, and different location of the tumors. We also understand that the follow-up period is too small to warrant any relevant recommendation or conclusion. All the patients will be strictly followed up to monitor progress and recurrence of the tumor.

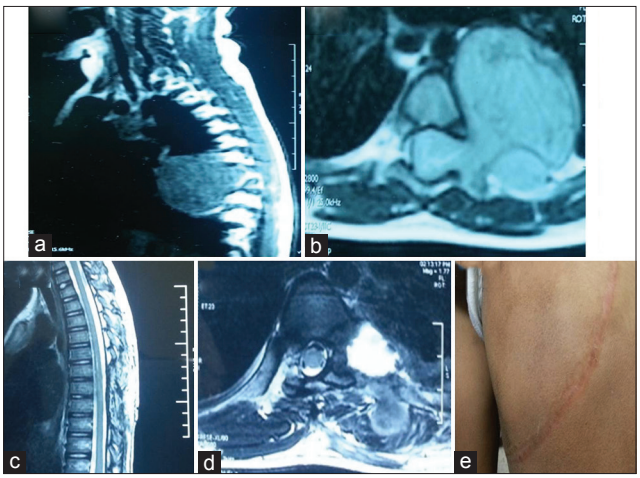

Figure 4: (a) Sagittal images showing tumor extending from D8 to D9 level with paraspinal extension. (b) Axial image showing foraminal extension of the lesion. (c) Postoperative sagittal scan showing complete excision of tumor. (d) Postoperative axial scan showing complete excision of the tumor. (e) The operative scar of posterolateral thoracotomy on the left side

Conclusion

With rising life expectancy, we are more likely to encounter complex and sizeable spinal tumors in general neurosurgical practice. In continuation of the present trend of increased involvement of neurosurgeons in the management of spinal conditions, the neurosurgical community must shed its inertia in singlehandedly treating these cases. We have demonstrated in this series that with the judicious use of access surgeons, careful preoperative planning, and adjuvant measures, satisfactory outcomes are possible in these patients with minimal morbidity and blood loss.

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Conflicts of interest

There are no conflicts of interest.

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