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Survival and Functional Outcomes in Patients with Thoracic Spinal Epidural Lymphoma with Spinal Cord Compromise: Three Cases Requiring Emergent Surgical Management

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Abstract

Spinal epidural lymphoma is a rare pathological entity that usually responds well to chemotherapy. However, it occasionally requires immediate surgical treatment for progressive neurological symptoms caused by a bulky epidural mass and bone destruction. In the management of spinal epidural lymphoma, the role of surgical interventions has not yet been fully defined, and few reports on long-term functional outcomes are available. We report 3 cases of thoracic spinal epidural lymphoma that required emergent surgical management for neural decompression and were followed postoperatively over 2 years or until death. Two patients (cases 1 and 2) underwent spinal instrumentation to prevent subsequent spinal deformity, whereas instrumentation was avoided in case 3 due to atopic dermatitis, which increases the risk of surgical site infection. During the 2-year postoperative period, 2 patients (cases 1 and 3) maintained disease-free survival and achieved sufficient neurological recovery to remain ambulant, while the other patient (case 2) died due to spinal epidural lymphoma relapse. Given the high treatment responsiveness and the potential for long-term disease-free survival in spinal epidural lymphoma, treatment should aim to maximize functional outcomes. Prompt surgical decompression is mandatory in patients with neurological deficits. Although spinal instrumentation is typically recommended to prevent spinal deformity, it should be withheld if it poses a risk of delaying chemotherapy, such as in cases with a high risk of surgical site infection. Therefore, spinal surgeons play several essential roles in managing spinal epidural lymphoma from the acute to the chronic phase, aiming to improve patients' prognosis and quality of life.

Keywords: thoracic spinal epidural lymphoma, tissue diagnosis, spinal decompression, instrumented surgery

Introduction

Spinal epidural lymphoma (SEL) is a rare pathological entity, accounting for 0.1%-3.3% of all lymphomas, yet it represents approximately 10% of spinal epidural tumors and is the second most common type following metastatic tumors.[1-3] SELs can occur at any age, with a median age of 45 years. The thoracic spine is the most commonly affected site, followed by the lumbar and cervical regions. Histologically, diffuse large B-cell lymphoma (DLBCL) is the most common type of SEL, although low-grade B-cell or T-cell types are also observed.[4] SEL should always be considered in the differential diagnosis of epidural lesions because it lacks imaging features distinct from those of other diseases, such as metastases, meningiomas, multiple myeloma, and inflammatory lesions.[5] A correct diagnosis can be made based on histopathological examination.

As with other lymphomas, chemotherapy (CTh) is the

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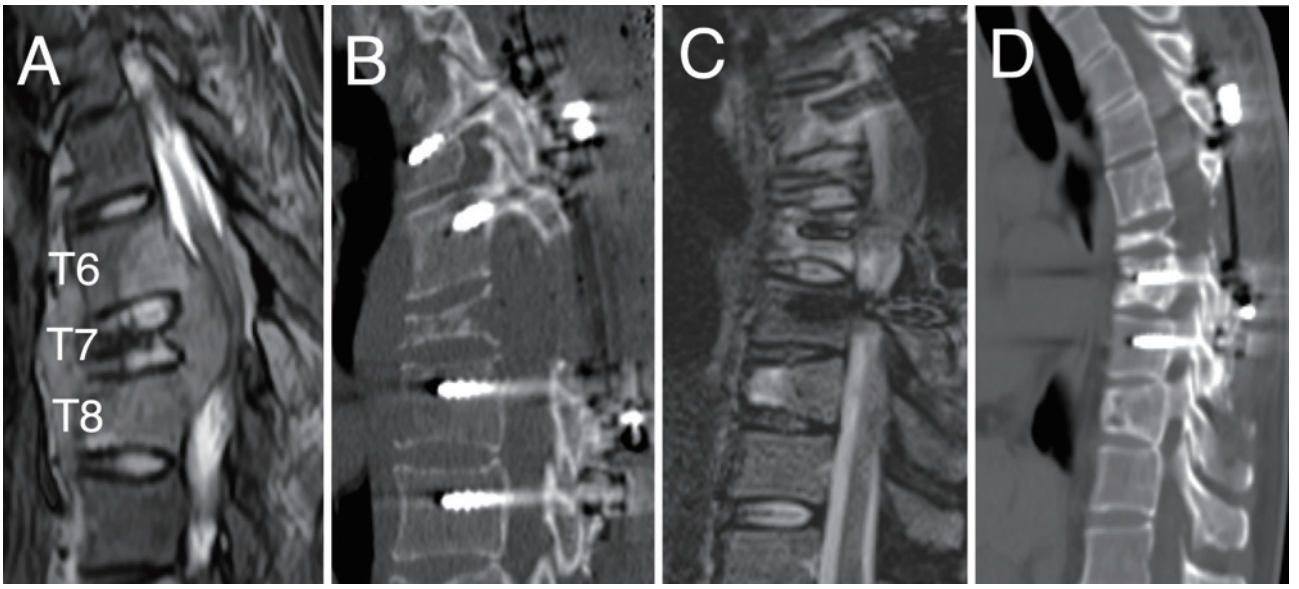

Fig. 1 Preoperative and postoperative imaging studies of case 1.
Preoperative sagittal T2-weighted MRI (A) showing an extradural tumor involving the T6-8 vertebral bodies with severe spinal cord compression. Sagittal CT scan immediately after surgery (B), revealing posterior decompression with T7 laminectomy and spinal stabilization with posterior instrumentation. Sagittal T2-weighted MRI at 3 months (C) showing CR after CTh. Sagittal CT at 23 months (D) revealed that spinal alignment was maintained, albeit with slight progression of the T7 vertebral fracture.
CR: complete remission; CT: computed tomography; CTh: chemotherapy; MRI: magnetic resonance imaging

gold-standard treatment for SEL, with a relatively good prognosis expected in more than half of patients (5-year overall survival rate of 69%).[6] However, in most cases, the condition is complicated by (1) myelopathy or radiculopathy caused by a bulky epidural mass and (2) spinal instability, mainly due to bone structure destruction.[7] The significance of surgical interventions in such SEL cases has been debated, particularly regarding tissue diagnosis and spinal decompression; however, few reports are available on the indications for instrumented surgery and its long-term functional outcomes. Herein, we present 3 cases of thoracic SEL requiring emergent surgical management for neural decompression, the functional outcomes of which were followed postoperatively over 2 years or until death.

Case Report

Case 1

A 31-year-old woman presented with a 1-month history of fever of unknown origin and swelling of the cervical lymph nodes. A definitive diagnosis was not obtained despite undergoing a lymph node biopsy at another hospital. Subsequently, the patient developed progressive paralysis, and spinal computed tomography (CT) scans revealed vertebral body collapse at T7, leading to a referral to our hospital for admission. The initial neurological evaluation revealed severe paralysis and bladder-bowel dysfunction, classified as grade B2 on the modified Frankel Scale (MFS). Magnetic resonance imaging (MRI) showed an extradural mass (T6-8) causing ventral compression of the thoracic spinal cord and a pathological compression fracture of the T7 vertebral body, with a Spine Instability Neoplastic Score (SINS) of 16 points (Fig. 1A).[8]

Urgent decompressive surgery was performed for functional recovery on hospitalization day 3. First, the spinous processes, laminae, and transverse processes were exposed from T5 to T9, and the exposed spinous processes were fully removed. Normal local bone was harvested for grafting. Intraoperative findings showed that tumor infiltration extended to the entire T7 lamina, and thoracic instability was present specifically at the T7 level; therefore, posterior instrumentation from T5 to T9 was performed before tumor debulking. The bilateral T7 laminae and T8 superior articular processes were resected, and T6 and T8 laminae were partially resected for sufficient neural decompression. The extradural tumor on the ventral side of the dural sac and the infiltrated parts of the vertebral body at the T7 level were resected piecemeal via the bilateral T7 pedicles (Fig. 1B). Decompression was completed by confirming the pulsation of the dural sac. T7 spinous process and extradural tumor specimens were subjected to histological examination, and the diagnosis of DLBCL was subsequently confirmed.

Six cycles of combination therapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) were promptly administered by hematologists. Follow-up postoperative images at 3 months demonstrated radiologic complete remission (CR) and preservation of

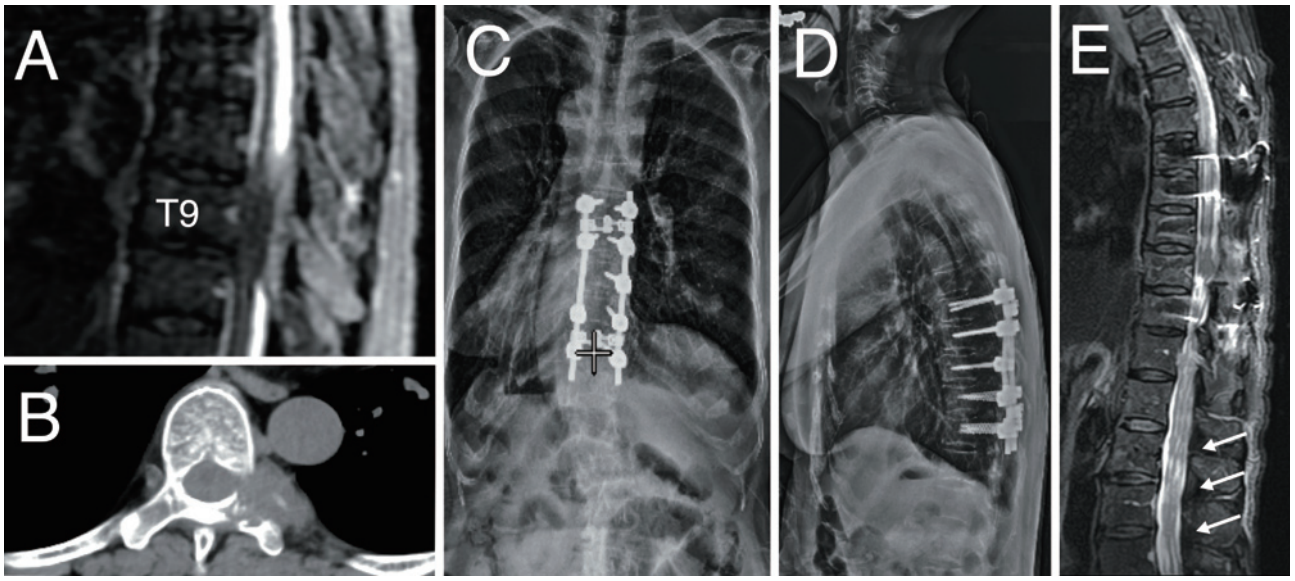

Fig. 2 Preoperative and postoperative imaging studies of case 2.
Preoperative sagittal T2-weighted MRI (A) and axial CT scan image at the T9 level (B) showing an extradural tumor located posterolaterally to the spinal cord at the T8-10 levels with osteolysis of the left pedicle of the T9 vertebral arch. Postoperative plain radiographs (C, D) showing spinal stabilization using posterior instrumentation from T7 to T11. The T9 vertebra was stabilized using only a right pedicle screw. Sagittal T2-weighted MRI at 11 months (E), illustrating dissemination to the conus medullaris and cauda equina (white arrows) without recurrence in the primary area.
CT: computed tomography; MRI: magnetic resonance imaging

spinal alignment, as shown in Fig. 1C and D. With continued rehabilitation, her paraplegia and bladder-bowel dysfunction improved, and the MFS recovered to grade D2. Twenty-four months have passed since treatment initiation without relapse.

Case 2

A 79-year-old woman was referred to our hospital with rapidly progressing paraparesis. MRI revealed an extradural tumor (T8-10) causing ventral compression of the thoracic spinal cord, as well as a diffuse osteolytic lesion in the T9 lamina and left pedicle (Fig. 2A and B), with a SINS of 9 points. Upon initial examination, the patient exhibited paraparesis and bowel-bladder dysfunction classified as grade B2 on the MFS. Whole-body CT revealed mediastinal lymphadenopathy.

One-stage posterior decompression and fusion were semi-urgently performed on hospitalization day 5. Specifically, T9 laminectomy, T8 and T10 doming, and posterior instrumented fusion from T7 to T11 were performed with autologous bone grafting (Fig. 2C and D). After sufficient spinal decompression, the resected lamina at T9 and the extradural tumor were subjected to histopathological diagnosis, confirming the diagnosis of DLBCL.

Postoperatively, 6 courses of the rituximab, pirarubicin, cyclophosphamide, vincristine, and prednisolone (R-THP-COP) regimen were promptly administered, leading to a partial response; however, the patient relapsed 7 months after diagnosis. Therefore, 6 cycles of the polatuzumab vedotin plus bendamustine and rituximab (Pola-BR) regimen for relapsed DLBCL were added; however, metastasis to the conus medullaris and cauda equina occurred 11 months after surgery (Fig. 2E). Despite multidisciplinary treatment for SEL and rehabilitation, the patient failed to achieve neurological recovery, remained classified as grade C1 on the MFS, and died 12 months after treatment initiation, soon after the second relapse.

Case 3

A 73-year-old woman with a comorbidity of severe atopic dermatitis presented to our hospital with fever and progressive paralysis. Three weeks before her hospital visit, the patient had also been brought to another hospital because of chest pain; however, she was discharged after cardiovascular events were ruled out. On initial neurological evaluation at our hospital, there was severe paralysis and bladder-bowel dysfunction, classified as modified Frankel Grade B1. MRI scans revealed an extradural tumor from T 4 to T6, causing spinal cord compression and pathological compression fractures of the T4 and T5 vertebral bodies, with a SINS of 16 points, compatible with the "instability" group (Fig. 3A).[8] Whole-body CT screening revealed no lesions other than spinal lesions.

Urgent decompressive surgery was performed for functional salvage on hospitalization day 1. The spinous processes, laminae, and transverse processes were exposed

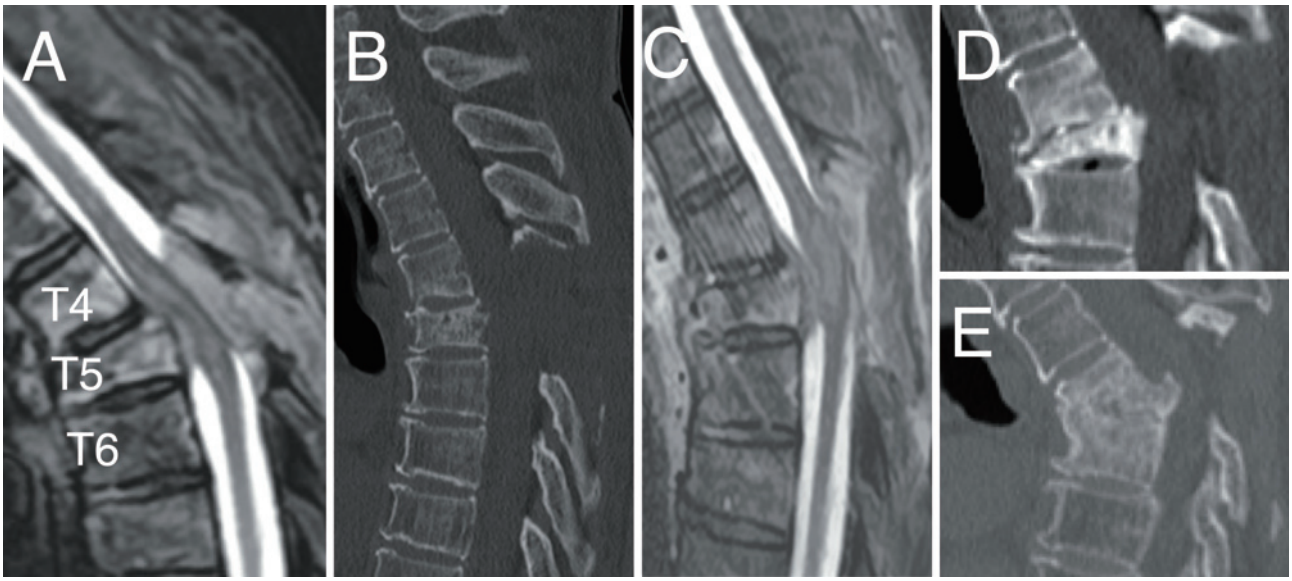

Fig. 3 Preoperative and postoperative imaging studies of case 3.
Preoperative sagittal T2-weighted MRI (A) showing an extradural tumor located behind the spinal cord at the T4-6 levels with a pathological compression fracture of the T4 and T5 vertebral bodies. CT scan (B) and T2-weighted MRI images (C) immediately after surgery, revealing posterior decompression obtained through tumor resection via T4-6 laminectomy. A CT scan at 6 months (D) depicts the progression of T5 vertebral collapse and slight local kyphosis. A CT scan at 26 months (E) demonstrated fused vertebral bodies between T4 and T5.
CT: computed tomography; MRI: magnetic resonance imaging

from T4 to T6, and the exposed spinous processes were fully removed. Due to tumor infiltration, the bilateral facet joints at T5 were dislocated, resulting in spinal instability. However, spinal instrumentation was intentionally avoided, considering the patient's medical history of severe atopic dermatitis on her back. It was anticipated that surgical site infection (SSI) could occur after instrumentation and hinder the continuation of CTh. While T4-5 laminectomy and T6 doming were performed, the extradural tumor and its infiltration into the vertebral body at T5 were partially resected piecemeal through the bilateral T5 pedicles (Fig. 3 B and C). After sufficient spinal decompression, the resected T5 lamina and extradural tumor were submitted for histopathological diagnosis, confirming the diagnosis of DLBCL.

R-CHOP CTh was administered postoperatively; however, it was discontinued after 3 cycles due to recurrent urinary tract infections (UTIs). Subsequently, field radiation therapy (RT) at 20 Gy was administered, which led to CR. Recurrent UTIs delayed ambulation for several months after treatment, and the patient remained at grade C1 on the MFS for 6 months. Although MRI showed postoperative progression of the T5 vertebral collapse and kyphosis deformity (Fig. 3D), her paraplegia and bladder-bowel dysfunction steadily improved. After 26 months of treatment, there has been no relapse, and in-home walking independence was finally achieved, with recovery to grade D1 on the MFS. As shown in Fig. 3E, conservative treatment achieved fusion of the T4 and T5 vertebral bodies.

Discussion

Herein, we report 3 patients with thoracic SEL who required emergent surgical management for aggressive disease progression. Only the patient in case 2, who had no extra-spinal lesion, corresponds to primary SEL. Two of them (cases 1 and 2) underwent spinal instrumentation to prevent subsequent spinal deformity and for spinal decompression, whereas in case 3, instrumentation was purposefully avoided due to a severe skin disorder. Since whole-body screening revealed no other organ lesions except for lymphadenopathy (Table 1), SEL rather than metastatic tumors was the primary diagnosis. Therefore, we collaborated preoperatively with hematologists in all 3 cases to initiate systemic treatment as early as possible. In the 2-year postoperative period, 2 patients (cases 1 and 3) maintained disease-free survival. They achieved favorable improvements in MFS, while the patient in case 2 had a poor prognosis due to SEL relapse. This survival discrepancy might be attributed to the advanced age (≥75 years) in case 2.[9,10] This study includes only 3 cases of SEL from a single center and lacks the ability to generalize the treatment strategy for SEL. However, this case series underscores the importance of prioritizing CTh as the primary treatment modality for this rare disease. Our observations suggest that early diagnosis followed by prompt initiation

Table 1 Summary of Patient Characteristics and Therapeutic Decisions

Case No.		1	2	3
Age / sex		31/F	79/F	73/F
Clinical manifestation		Paraplegia, BBD	Paraplegia, BBD	Paraplegia, BBD
Lesion level		T6-8	T8-10	T4-5
Spine Instability Neoplastic Score		16	9	16
Other than spinal lesion		Cervical LAP	No lesion	Mediastinal LAP
sIL-2R (U/mL)		2168	1848	1603
LDH (U/mL)		248	368	366
Surgery		PSD, PSIF	PSD, PSIF	PSD
Histopathological diagnosis		DLBCL	DLBCL	DLBCL
Denosumab		Yes	Yes	Yes
CTh	1st-line	R-CHOP, 6 cycles	R-CHOP, 6 cycles	R-CHOP, 3 cycles
	2nd-line	N/A	Pola-BR, 3 cycles	N/A
Radiation therapy		N/A	N/A	20 Gy, 5 Fr*
Response criteria		CR	PD	PR -> CR
Survival outcome		Alive	Dead	Alive
F/U period after surgery (months)		24	12	26
Modified Frankel Scale		B2 -> D2	B2 -> C1	B1 -> D1

BBD: bladder and bowel dysfunction; CR; complete remission; CTh: chemotherapy; DLBCL: diffuse large B-cell lymphoma; F/U: follow-up; LDH: lactate dehydrogenase; LAP: lymphadenopathy; PD: progressive disease; Pola-BR: polatuzumab vedotin plus bendamustine and rituximab; PSD: posterior spinal decompression; PSIF: posterior instrumented fusion; R-CHOP: rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone; sIL-2R: soluble interleukin-2 receptor
*Consolidative RT.

of CTh can substantially improve survival outcomes. Furthermore, as illustrated in case 3, effective CTh combined with timely neural decompression surgery, even in the absence of spinal instrumentation, can result in favorable functional recovery. These findings collectively highlight the central role of CTh in achieving both prolonged survival and neurological improvement in patients with SEL.

Hereafter, we discuss the role of spinal surgeons in the multidisciplinary treatment programs of SEL. First, spinal surgeons must suspect SEL when encountering an epidural mass lesion; subsequently, a rapid and accurate histological diagnosis is considered. The serum levels of soluble interleukin-2 (sIL-2) receptor proved to be a good indicator of SEL activity in our case series. However, preoperative ascertainment of sIL-2 might be difficult, especially if emergent surgery is required, since it takes several days to obtain results in most medical settings. In asymptomatic or slow-growing cases, nonsurgical tissue sampling may be an option to confirm the diagnosis, such as needle biopsy or superficial lymph node excision. However, these methods might lack diagnostic accuracy due to insufficient quantity or quality of specimens, as was the case with the patient in case 1.[11] In particular, when a bulky epidural mass severely compresses the spinal cord with rapid progression, emergent spinal decompression and definitive histological diagnosis are concomitantly warranted.[12] Given the lack of evidence on the prognostic impact of the extent of resection, the aim of surgery should be adequate tissue sampling and sufficient neural decompression, but not radical tumor resection.[6,7,13] Since there are concerns that preoperative use of steroids might affect diagnostic accuracy, they should be administered, to the extent possible, after surgery. Palliative RT for spinal cord compromise can be an alternative to surgical decompression,[14] but should be administered with caution, particularly in younger patients, in light of late radiation effects.[15] Currently, limited evidence supports the use of consolidative RT in patients who have undergone surgical decompression; it may be a practical option for SEL patients who require a reduced dose intensity of CTh due to toxicity or poor treatment tolerance, as was the case with the patient in case 3.[16]

In many patients with spinal instability owing to SEL, instrumented surgery with autologous bone grafting should be indicated until bone union is achieved, as more than half of these patients are expected to be disease-free long-term survivors.[6,17] The patient in case 1 represents those who benefit from instrumented surgery. However, because the prognosis of elderly patients with DLBCL is inferior to that of younger patients, instrumental management

should be carefully determined, particularly in patients aged 75 years or older.[9,10] It should be noted that SSIs after instrumentation can cause severe complications for SEL patients, leading to CTh discontinuation. Risk factors for SSIs after instrumentation, such as diabetes, obesity, malnutrition, and skin disorders, should be surveyed preoperatively.[18,19] In case 3, which was deemed to be at high risk for SSI, spinal instrumentation was not purposefully utilized, despite the risk of kyphotic deformity. However, bone synthesis and spinal stability were observed after successful chemoradiation therapy. Consequently, neurological deficits improved. The results of this case suggest that early introduction of CTh is crucial in patients with SEL and that our decision not to utilize spinal instrumentation was appropriate in case 3. While neural decompression alone may be a reasonable option for SEL patients with a high risk of SSI, it is important to note that this approach requires trade-offs, such as the need for rigid external immobilization during the course of CTh.

Conclusion

Patients with SEL show better chemosensitivity and promise for long-term disease-free survival. However, it remains a fact that some cases carry a poor prognosis, and early diagnosis and prompt treatment initiation must be prioritized above all to improve survival outcomes. Once CR is achieved, these patients have a good chance of sufficient recovery even after severe spinal cord compromise. Therefore, surgical treatment strategies should also aim to optimize functional outcomes. Prompt surgical decompression is mandatory in patients with neurological deficits. Although spinal instrumentation may be added to achieve bone union and long-term spinal stability, indications for instrumental management should be carefully determined in older adults and high-risk patients with SSIs. Spinal surgeons play several vital roles in managing SEL from the acute to the chronic phase, aiming to improve the prognosis and quality of life of patients with SEL.

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Disclaimer

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Conflicts of Interest Disclosure

All authors have no conflict of interest.

Ethical Approval

This study was approved by the Ethics Institutional Review Board of the Hanwa Memorial Hospital (reference number 2023-06). Informed consent regarding the use of medical information was obtained from patients.

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