Posterior Instrumentation without Curettage Promotes Rapid Restoration of Adult Spinal Langerhans Cell Histiocytosis

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Abstract:

Introduction: Adult spinal Langerhans cell histiocytosis (LCH) presents a treatment challenge due to ongoing controversies. Traditional approaches such as curettage with bone grafting and internal fixation are preferred for severe cases involving mechanical instability, neurological deficits, or deformity. This study aimed to explore the efficacy of a customized approach involving simple posterior instrumentation without curettage or bone grafting in treating adult spinal LCH.

Methods: This retrospective study analyzed a prospectively maintained database of all spine surgeries conducted at our institute from April 2013 to December 2020. Adult patients (age≥20) diagnosed with LCH were included. We assessed surgical methods, adjuvant therapy, and clinical results, such as perioperative progression of disease, symptoms, and recurrence.

Results: Four male patients aged between 21 and 28, each with a single spinal LCH lesion (T6, T5, and C5) except one case (T5 and T7), were treated. Diagnoses were confirmed via biopsy (two open, two needle biopsies). Whole-body computed tomography or bone scintigraphy revealed no additional LCH lesions in any patient, except in one patient with a small lung nodule. All patients presented with severe back or neck pain and pathological fractures at the affected vertebra. Thoracic LCH cases received percutaneous pedicle screw fixation, while the cervical case was managed with conventional posterior instrumentation using lateral mass screws. After surgery, all patients experienced significant pain relief, halted bone lysis, and rapid new bone formation. One patient underwent chemotherapy postsurgery. Over 3 years of follow-up, imaging studies revealed no recurrences of the disease.

Conclusions: Posterior instrumentation, without the need for curettage or bone grafting, is a promising surgical treatment for adult spinal LCH. This method may effectively halt lesion progression, prevent spinal deformity, and avert neurological deficits in the patients with progressive spine lesion where conservative treatment may not adequately prevent vertebral fractures.

Keywords:

Langerhans cell histiocytosis (LCH), Adult spinal LCH, Posterior instrumentation, Curettage, Spine tumor, Percutaneous pedicle screw

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Introduction

Langerhans cell histiocytosis (LCH) is a rare disease that is particularly prevalent in children, characterized by the accumulation of CD1a+/Langerin+ LCH cells[1)]. The clinical spectrum ranges from single-system LCH, which may spontaneously remit, to multiple-system LCH, treated with chemotherapy[1)]. In children, spinal LCH is usually treated non-surgically and resolves spontaneously with the collapse of the affected vertebra[2,3)]. Remodeling of the affected vertebra is observed, particularly in children[2-5)]. However, adult spinal LCH often presents with neurological symptoms and vertebral destruction, necessitating surgical intervention in most cases[6,7)]. Traditional surgical treatment typically involves lesion curettage with bone grafting and instrumentation, yielding satisfactory clinical results[8-11)]. However, these surgeries

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Table 1. Summary of the Clinical Data of the Patients.

	Case 1	Case 2	Case 3	Case 4
Age	27	26	21	28
Sex	Male	Male	Male	Male
Smoking	Current	Former	Current	No
Concomitant disease	None	None	None	None
Symptoms	Severe back pain	Severe back pain	Severe neck pain	Severe back pain and anterior chest pain
Neurological deficit	Mild numbness in bilateral lower limbs	None	None	None
Diagnostic methods	Two needle biopsies Open biopsy	Needle biopsy	Open biopsy	Needle biopsy
Location of the lesion	T6	T5	C5	T5 and T7
Other LCH lesions	None (examined with bone scintigraphy and whole-body CT)	Small nodule in lung (examined with whole-body CT)	None (examined with whole-body CT)	None (examined with whole-body CT and brain MRI)
Duration from symptom onset to surgery	9 weeks	6 weeks	20 weeks	9 weeks
Surgery	PPS fixation (T4, T5, T7, and T8)	PPS fixation (T4 and T6)	Open fixation with lateral mass screw (C4, C5, and C6)	PPS fixation (T4, T6, and T8)
Adjuvant therapy	None	Chemotherapy	None	None
Follow-up periods (years)	5	3	3	3
Implant removal	No	2 years postoperatively	2 years postoperatively	2 years postoperatively
Remission of LCH	Yes	Yes	Yes	Yes
Neurological deficit at final follow-up	No	No	No	No

LCH, Langerhans cell histiocytosis; PPS, percutaneous pedicle screw

are invasive, involving significant tissue damage, bone harvesting, losing mobile segments, and potential future complications such as adjacent segment disease[12].

In contrast, we have been treating adult spinal LCH cases with severe vertebral bone lysis using simple posterior fixation without curettage or bone grafting. This study aims to evaluate the effectiveness of our treatment approach for adult spinal LCH.

Materials and Methods

This study was a retrospective observational case series and was approved by the Institutional Ethics Committee.

Patients

We retrospectively reviewed patients who underwent surgery for histologically diagnosed spinal LCH, extracted from a prospectively maintained database of all spine surgeries at our institute from April 2013 to December 2020. Patients below 20 years old were excluded.

Outcomes

We investigated whether remission of spinal LCH lesions was achieved and examined the presence of spinal deformities and neurological deficits at the time of the final follow-up.

Moreover, we recorded demographic and other clinical data, such as imaging studies, comorbidities, symptom duration, surgical methods, and adjuvant therapy.

Results

Four male patients, aged between 21 and 28, underwent surgical treatment. Three patients had single spinal lesion each (T6, T5, and C5), while one patient had multiple lesions (T5 and T7). Whole-body Computed tomography (CT), bone scintigraphy, and/or brain Magnetic resonance imaging (MRI) revealed no involvement of other organs in any patients, except for one patient with small lung nodules. Table 1 summarizes the demographic and clinical details of all four patients.

The initial symptoms were back or neck pain, localized around the affected lesion, with no preceding trauma or specific incident reported. Notably, one patient (Case 4) had received a COVID-19 vaccine 2 weeks before symptom onset. All patients exhibited substantial and progressive osteolysis in the affected vertebral body. All patients were diagnosed with LCH either by needle biopsy (Cases 2 and 4) or open

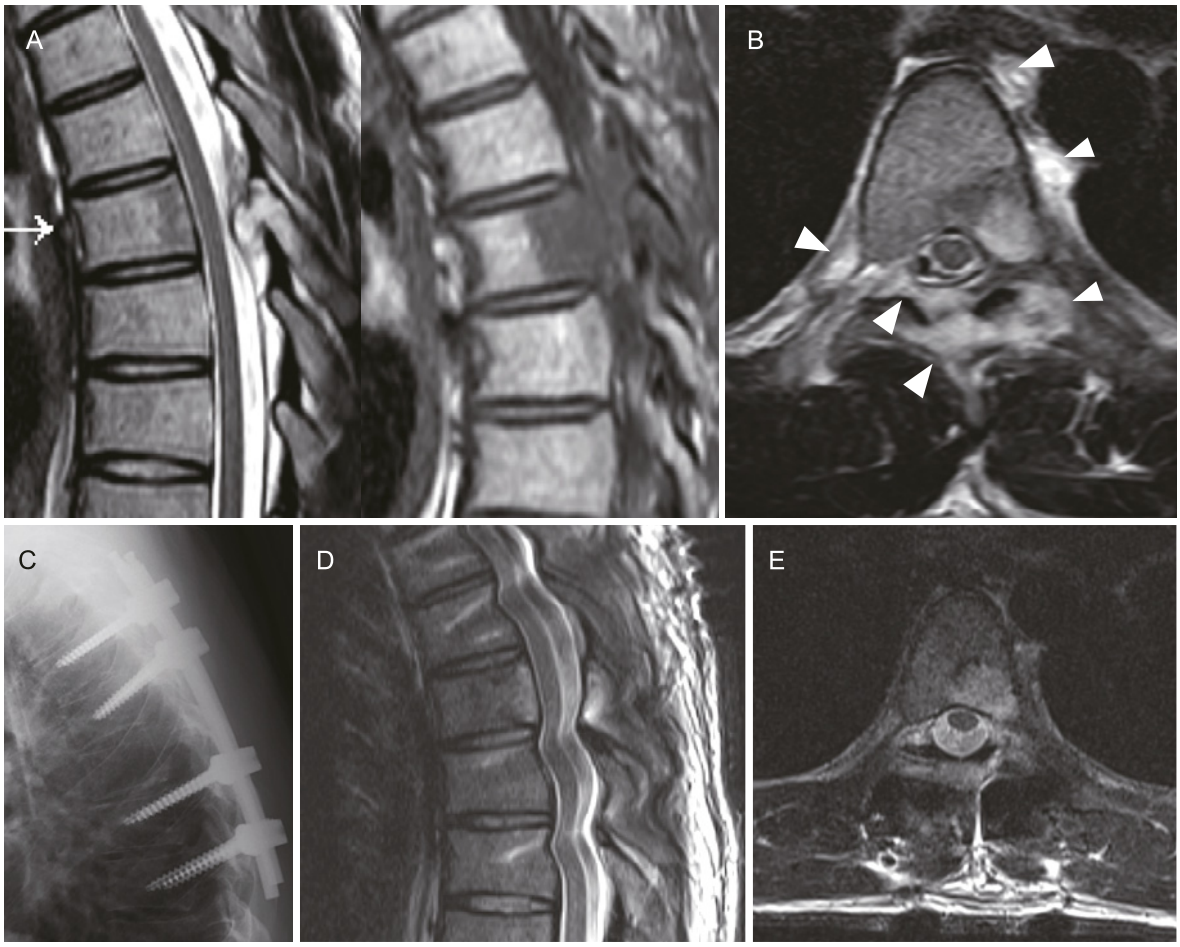

Figure 1. Imaging study of Case 1: A, T2-weighted (left) and T1-weighted (right) magnetic resonance imaging (MRI) displaying an abnormal lesion in the T6 vertebral body, lamina, and epidural space. B, T2-weighted axial MRI highlighting the abnormal lesion extending into the epidural and perivertebral spaces (indicated by white arrowheads). C, Lateral X-ray postfixation surgery, showing the surgical outcome. D and E, T2-weighted MRI scans taken 7 days postsurgery, revealing a significant reduction in the size of the epidural lesion.

biopsy (Cases 1 and 3). Specifically, Case 1 underwent an open biopsy along with percutaneous pedicle screw (PPS) fixation after two needle biopsies failed to clarify the diagnosis. Case 3 underwent an open biopsy under general anesthesia due to the location of the lesion in the cervical vertebral body. The duration from symptom onset to surgery was 6-20 weeks.

Postoperatively, all patients experienced significant pain relief. CT scans demonstrated cessation of bone lysis and rapid new bone formation, confirmed within 2 months after surgery. Pathological fracture or deformity of the vertebral shape did not progress after the index surgery, and the shape of the vertebrae was reconstructed to the same form as before surgery. However, deformities caused by fractures that had already occurred before surgery were not repaired even after remodeling (Case 2).

All patients, except one, consented to implant removal, which was successfully performed without complications. One patient (Case 2) underwent chemotherapy 6 months postsurgery at another hospital, although the lung lesion had diminished. Over a 3-year follow-up period, imaging studies revealed no recurrences of the disease.

Case Presentation

Case 1

A 27-year-old male presented at our hospital with severe back pain and bilateral lower limb numbness. MRI revealed an abnormal lesion in the T6 vertebra and posterior epidural space (Fig. 1A and B). CT confirmed a large lytic lesion in T6 (Fig. 2). Two CT-guided needle biopsies failed to provide a definitive diagnosis. Due to the rapid progression of vertebral lysis (Fig. 2), we opted for posterior instrumentation using a PPS system combined with an open biopsy (Fig. 1C). He experienced significant relief from back pain soon after the surgery. Pathological examination confirmed the diagnosis of LCH. Notably, the epidural lesion visible in the preoperative MRI had almost completely resolved in the MRI performed 7 days postsurgery (Fig. 1D and E). A subsequent CT scan, taken 6 weeks postoperatively, demonstrated rapid new bone formation at the lesion site (Fig. 2). He was followed up without any adjuvant therapy. Three years postoperatively, a CT scan showed the vertebra appearing almost normal, with the patient remaining asympto-

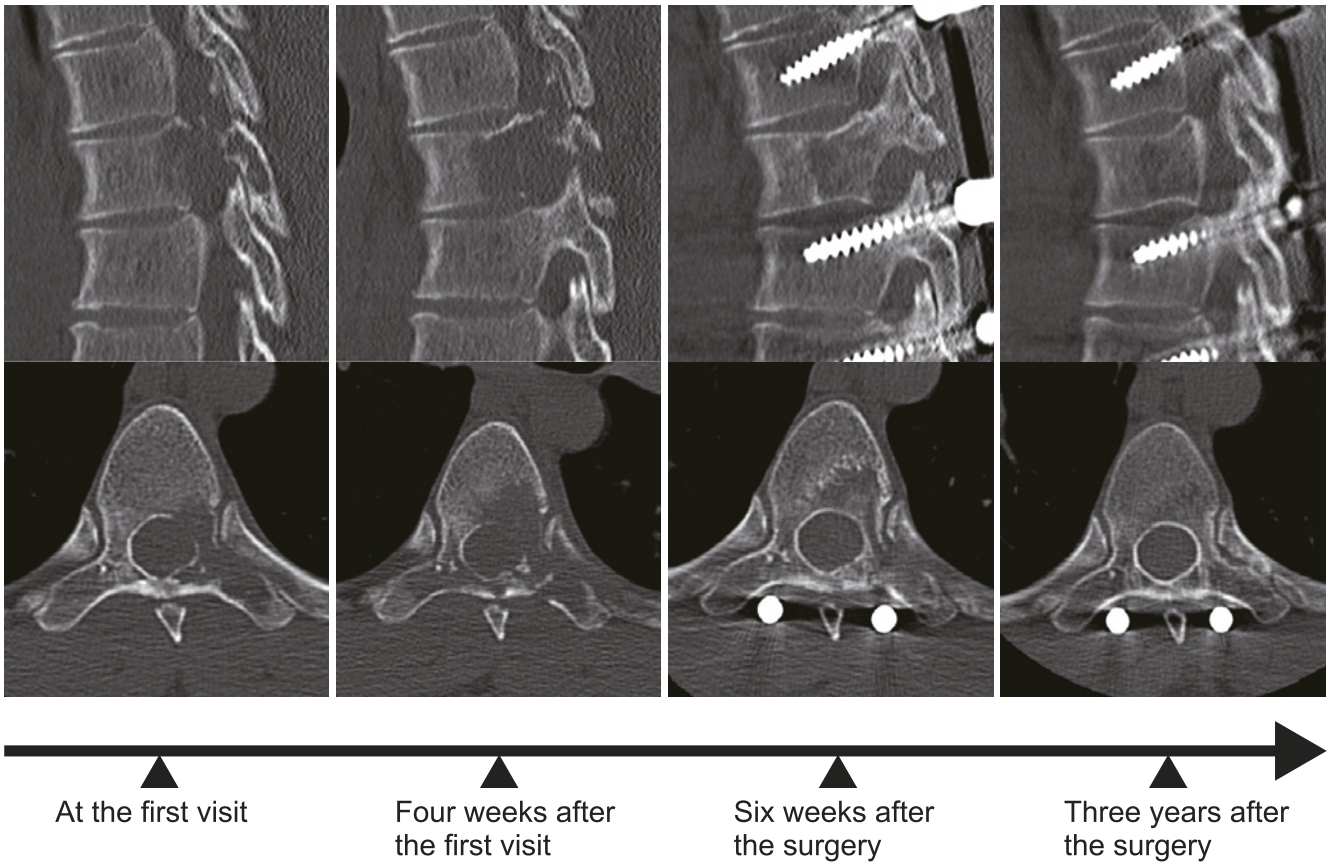

Figure 2. This series shows the progression and treatment response of the bone lysis in Case 1. Following the posterior instrumentation with the percutaneous pedicle screw system, there was a cessation of the rapid bone lysis. New bone formation was observed as early as 6 weeks postsurgery. The final CT scan, taken 3 years postoperatively, reveals a vertebra that appears almost normal.

matic (Fig. 2).

Case 2

A 26-year-old male presented to our hospital with back pain. MRI identified a lesion in the T5 vertebra (Fig. 3A and B), and a CT-guided needle biopsy confirmed the diagnosis of LCH. In addition, a lung CT scan revealed a small abnormal nodule (Fig. 3C). He underwent simple posterior fixation using a PPS system (Fig. 3D) due to rapid progression of bone lysis and accompanying fracture of the distal endplate (Fig. 4). His back pain significantly improved soon after the surgery. A CT scan performed 5 weeks postsurgery demonstrated rapid bone reconstitution (Fig. 4). Further imaging after 7 months revealed the vertebra to be almost normal (Fig. 4). Although the lung lesion also diminished, chemotherapy was recommended and administered at another hospital. Two years after the initial surgery, he underwent successful implant removal. At the last follow-up, he was symptom-free, and the X-ray study revealed no abnormalities (Fig. 3E).

Discussion

This study demonstrated that posterior fixation alone, without bone grafting or curettage, can facilitate rapid reconstitution of spinal LCH lesions in four adult patients. Notably, this bone reconstitution occurred without the need for adjuvant chemotherapy in all but one case where chemotherapy was administered postreconstitution.

The first-line treatment for spinal LCH, in both pediatric and adult patients, typically involves conservative management, with or without chemotherapy[1,13-15]. Bisphosphonates represent another treatment option for spinal LCH and other osseous involvement, with several case series reporting successful outcomes[16,17]. Furthermore, smoking cessation is important for the regression of adult LCH[18]. In pediatric cases, spinal LCH often resolves without surgery, frequently through spontaneous collapse and remodeling of the affected vertebra[2,3]. However, this natural process of remodeling may be inhibited in children over 15 years old or in those undergoing vertebral curettage and may lead to residual spinal deformities[5]. In contrast, adult spinal deformities are less likely to remodel naturally once vertebral collapse occurs, often resulting in persistent vertebral deformities and more common neurological dysfunctions[6,7]. Therefore, surgical intervention may be warranted if bone lysis rapidly progresses, significant bone destruction and potential fractures are anticipated, or partial fractures have already occurred, to prevent vertebral collapse.

Traditional fusion surgery for spinal LCH[8-11] presents several disadvantages, such as extensive tissue damage and the potential for adjacent segment problems[12]. To address these

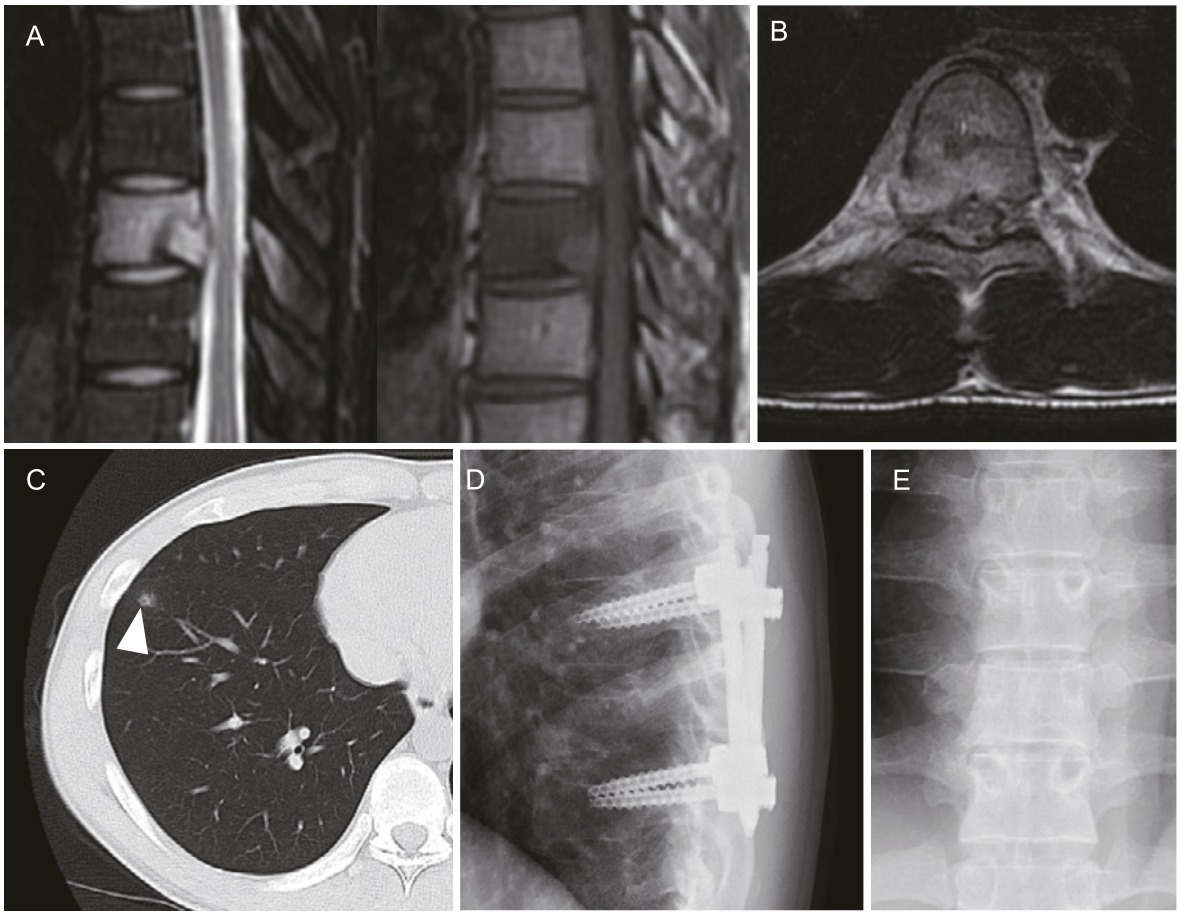

Figure 3. Imaging study of Case 2. A, Fat suppression T2-weighted (left) and T1-weighted (right) magnetic resonance imaging (MRI) showing an abnormal lesion in the T5 vertebra of Case 2. B, Fat suppression T2-weighted axial MRI revealing perivertebral and epidural lesions. C, A small nodule observed in the right lung. D, X-ray taken after the fixation surgery, showing the postoperative status. E, Postoperative X-ray taken 2 years later, after the removal of the instrumentation.

issues, we adopted a customized approach of posterior instrumentation without curettage or bone grafting. This method was based on the hypothesis that preserving the vertebral body's anatomical shape might allow future remodeling and avoiding curettage could help maintain normal structure. Furthermore, this technique, particularly using the PPS system[19], allows for minimally invasive fixation, particularly beneficial in the thoracic and lumbar spine. Furthermore, this method can save mobile segments if instrumentation is removed after recovery.

Our study observed an immediate halt in the progression of bone lysis postsurgery, with rapid bone reconstitution in all patients. The first case notably showed a significant decrease in the epidural lesion just 7 days after surgery (Fig. 1 B). The exact mechanisms behind this rapid remission remain unclear. LCH has two aspects, neoplastic nature[20,21] and a reactive disease[22]. We hypothesize that mechanical loading will promote the disease progression of LCH via upregulation of cytokine signaling[22] by uncertain mechanisms; thus, the posterior instrumentation may cease the progression of LCH lesion.

The rapid bone destruction observed in our adult spinal LCH cases underscores the critical need for timely intervention. Delayed treatment can lead to irreversible deformities and neurological issues. Once a pathological fracture occurs, the resulting deformity may persist even after bone reconstitution, as demonstrated in Case 2. Therefore, early instrumentation surgery emerges as a viable option for severe cases. Although we have not experienced cases of continued disease progression postsurgery, should this occur, additional treatments such as corticosteroid injections[23] or chemotherapy[1] could be considered.

Although the results are promising, the small sample size of our study necessitates cautious interpretation. In addition, we did not have access to health-related quality of life scores or visual analog scales for back or neck pain. The mechanisms driving LCH remission remain unclear. Therefore, further research involving larger cohorts and longer follow-up periods is essential to confirm the efficacy and safety of our treatment strategy.

In conclusion, the use of posterior fixation without curettage or bone grafting in four adult patients with LCH resulted in rapid regression of bone lysis and subsequent bone reconstitution. This less invasive approach, employing the

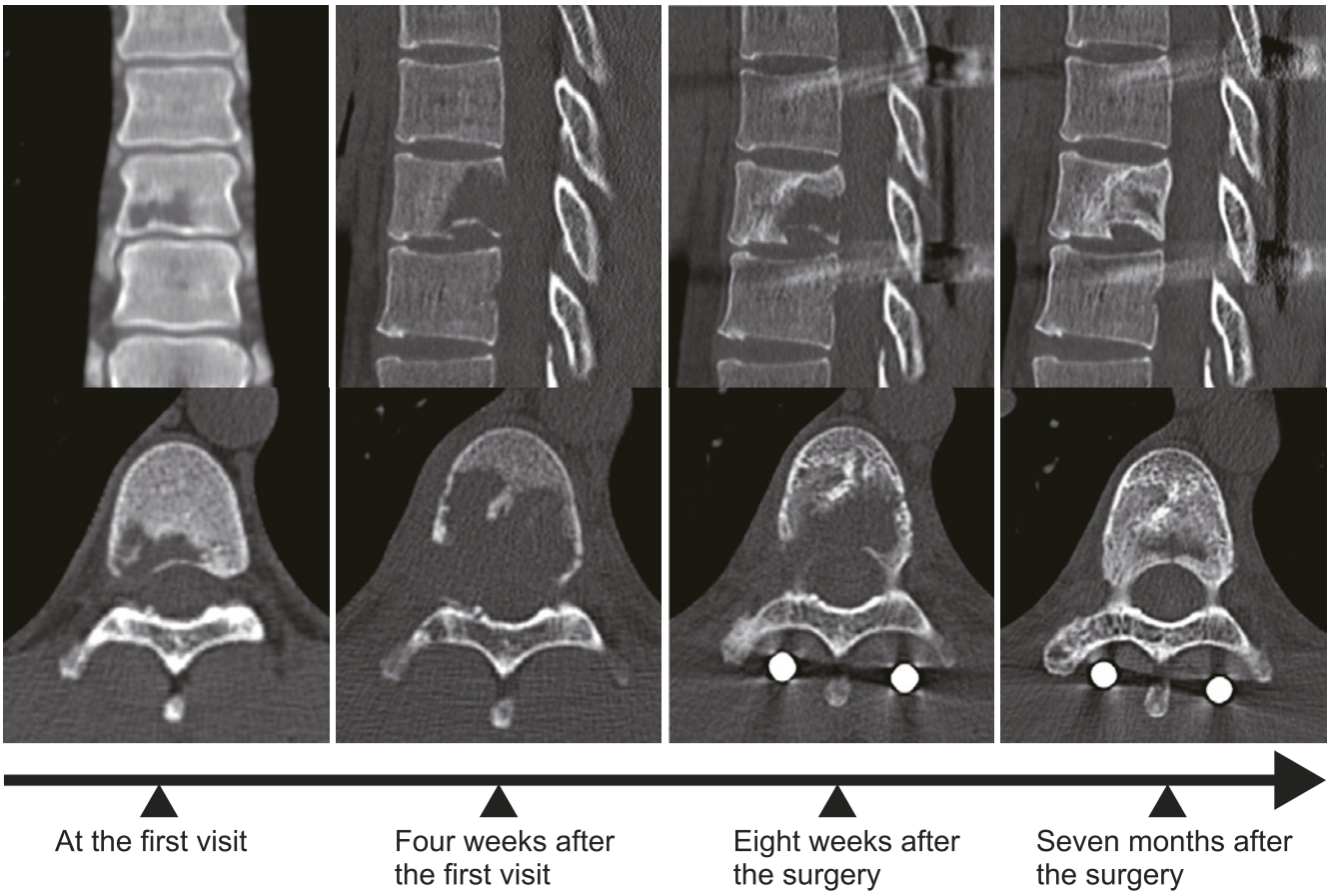

Figure 4. This series shows the progression and subsequent response to treatment of bone lysis in Case 2. Following the posterior instrumentation using the percutaneous pedicle screw system, a halt in the rapid progression of bone lysis was observed. Seven months after operation, significant bone reconstitution is evident, although some deformation of the endplate persists, as was present prior to the operation.

PPS system, is an effective treatment strategy for managing progressive LCH lesions in the adult spine, particularly in cases where conservative treatment alone may not suffice to prevent vertebral fracture.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

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Author Contributions: B.O. (Bungo Otsuki): conceptualization, methodology, data curation, and writing original draft preparation.

H.K. (Hiroaki Kimura), S.F. (Shunsuke Fujibayashi), T.S. (Takayoshi Shimizu), T.S. (Takashi Sono), and K.M. (Koichi Murata): visualization, investigation, and writing-reviewing and editing.

S.M. (Shuichi Matsuda): supervision, reviewing, and editing.

Ethical Approval: This study was approved by the Institutional Review Board of Kyoto University (R2508). Patients were not required to give informed consent to the study because the analysis used anonymous clinical data that were obtained after each patient agreed to treatment by written consent. In addition, we used an opt-out method to obtain consent for this study through our institute's homepage, which was approved by the Institutional Review Board.

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