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CASE REPORT

Recurrent primary osseous hemangiopericytoma in the thoracic spine: a case report and literature review

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Abstract
Purpose Primary osseous hemangiopericytoma (HPC) of the spine is exceedingly rare. HPC has malignant potential and has the capacity for metastasis and local recurrence. We herein present the first case of recurrent primary osseous HPC in the thoracic spine that was successfully treated by total spondylectomy at three vertebral levels and spinal reconstruction.
Methods We performed a two-stage operation for recurrent HPC using anterior and posterior approaches at the T5–T7 vertebrae. The preoperative embolization of the tumor was performed to prevent massive intraoperative bleeding. Then, total spondylectomy was performed (T5–T7) to resect the tumor. Anterior spinal reconstruction and posterior instrumentation were performed, with abundant bone autograft and allograft used to achieve sufficient boney fusion following the removal of the tumor.
Results At 2 years after surgery, the patient had made a sufficient recovery from his symptoms. The bone union was complete without tumor recurrence or implant failure.
Conclusions Total spondylectomy and spinal reconstruction with instrumentation might be useful for performing the safe and adequate excision of recurrent HPC of the spine. However, patients should be closely monitored to detect local recurrence and the malignant degeneration of the tumor after surgery.

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Keywords Hemangiopericytoma · Recurrence · Spine · Myelopathy · Spondylectomy

Introduction

Hemangiopericytoma (HPC) usually occurs in various soft tissues, such as the subcutaneous tissues, the pelvic fossa and the retroperitoneum, and is occasionally found as meningeal tumor in the cranial cavity [1]. On the other hand, primary HPC is rare in the skeleton [2–4]. Among these, primary osseous HPC of the spine is exceedingly rare [1, 5].

Although HPC is slow-growing tumor and the prognosis is generally more favorable than that of sarcomas, HPC in both the soft tissues and the bones has malignant potential and has a capacity for metastases and local recurrence [6, 7]. Thus, wide en bloc excision is the best treatment for HPC [8]. However, in some cases of HPC of the spine, performing an appropriately wide en bloc excision is impossible due to the anatomy of the spine and neural structures and the risk of excessive hemorrhage from the tumor [1, 9]. Furthermore, in cases of recurrent spinal tumors, scar adhesion between the tumor and the neural structure and unclear tumor margins due to previous surgery are a great hindrance to adequate and safe excision [10]. Because of the rarity of primary osseous HPC of the spine and the difficulty of appropriate resection, the optimal treatment remains unknown.

We herein present the first case of recurrent osseous HPC of the thoracic spine to be successfully treated by total spondylectomy at multiple vertebral levels and spinal reconstruction. In addition, we review the literature on primary osseous HPC of the spine. The patient gave his informed consent for the publication of the clinical data.

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Case report

Clinical history

This 25-year-old man having numbness in the body trunk and bilateral lower extremities presented to a regional hospital. Imaging findings revealed spinal tumor in the T6 vertebral body compressing the spinal cord. Then, paraplegia deteriorated acutely and emergency surgery for posterior decompression and tumor resection was performed. However, tumor was not removed completely due to severe hemorrhage at 11,000 ml during the surgery. Although histopathological analysis of the tumor was performed, definite pathological diagnosis was unable to be made. The paraplegia was almost recovered but only numbness remained slightly in the body trunk and bilateral lower extremities after surgery.

During the postoperative follow-up period, imaging findings revealed a gradual increase of the size of the residual tumor and compression of the thoracic spinal cord. His numbness was worse again when he was 30 years old, and he was re-operated at the same hospital. Before the second operation, arterial supplies to the tumor from the segmental arteries at T5 and T6 levels on bilateral sides were confirmed and embolization of these arteries was performed. Then, a two-stage operation though anterior and posterior approaches was performed. First, removal of the tumor in the T6 vertebral body and spinal reconstruction were performed through anterior approach, and then laminectomy with posterior instrumentation was added through posterior approach. The total amount of blood loss was 6000 ml. His symptoms were almost recovered but only numbness remained slightly in the body trunk and bilateral lower extremities following the second surgery.

However, his numbness in the body trunk and the lower extremities was gradually worse and imaging findings revealed recurrent tumor was developed and increased over 13-year period. Then, he was referred to our department for further treatment when he was 43 years old.

Examination

His gait was spastic and slightly unstable, although he was able to walk without any assistance. On physical examination, pinprick and light touch demonstrated hypoesthesia in the body trunk and the lower extremities. His patella and Achilles tendon reflexes were exaggerated. No muscle weakness in the upper and lower extremities was observed. His neurological diagnosis was considered to be recurrence of thoracic myelopathy. The modified JOA scale for thoracic myelopathy was 4 of 11 points [11].

Plain radiographs showed the postoperative thoracic spine with instrumentation (Fig. 1). Magnetic resonance imaging (MRI) revealed that the recurrent tumor was involving vertebrae from T5 to T7 (Fig. 2) and gradually developed surrounding bony structures (Fig. 2). The ventral side of the spinal cord was slightly compressed by the tumor. The recurrent tumor was also observed in the epidural space. Computed tomography (CT) of thoracic spine showed boney destruction from T5 to T7 vertebrae (Fig. 3). In a preoperative angiography, arterial supplies to the tumor from the segmental arteries at T5, T6 and T7 levels on the left side were confirmed and embolization of these arteries were performed.

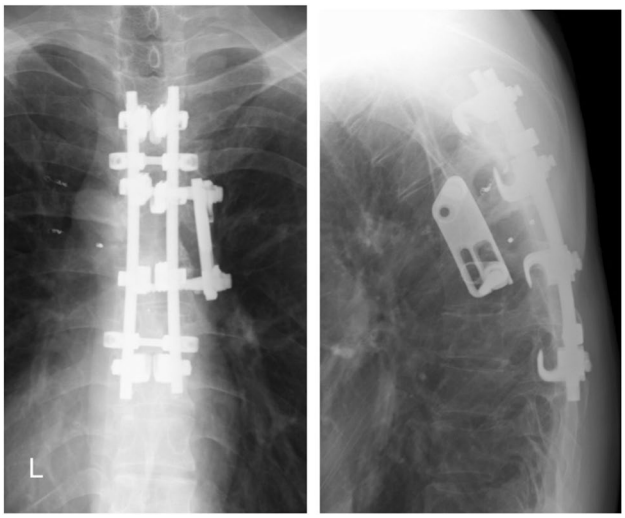

Fig. 1 Radiographs obtained before the third operation

Operation

We performed total spondylectomy from T5 to T7 in a two-stage operation through anterior and posterior approaches. In the first-stage procedure, which was performed using a posterior approach, laminectomy and pediculectomy were performed from T5 to T7 with implant removal. The tumor in the pedicle on the left side and epidural space at T6 level was completely removed. The spine from T2 to T10 was stabilized using a pedicle screw and rod system. Both autograft from the iliac bone and allograft were used abundantly to achieve posterior fusion securely (Fig. 4). The total amount of intraoperative blood loss was 1200 ml.

One week after the first-stage procedure, vertebrectomy was performed through an anterior approach from T5 to T7 to remove the tumor and anterior reconstruction was performed using a titanium mesh cylindrical cage. Both autograft from the iliac bone on the right side and allograft were packed into and around the cage (Figs. 4, 5). The total amount of intraoperative blood loss was 1200 ml.

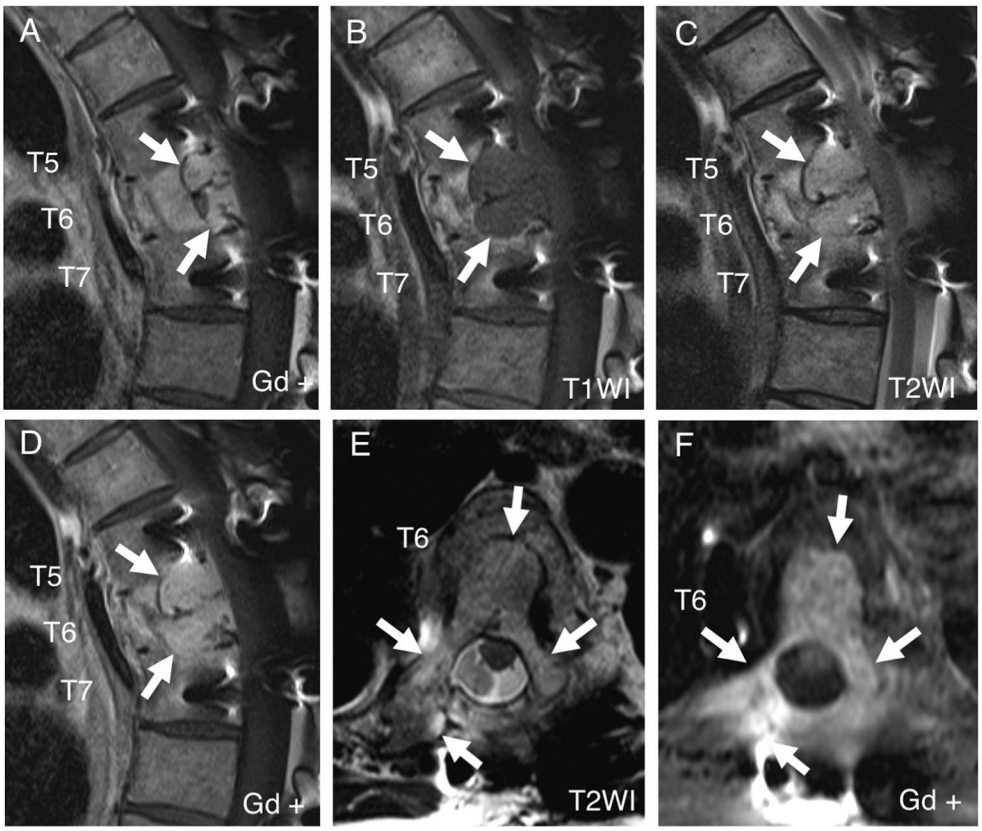

Fig. 2 MR images obtained before the third operation. Sagittal Gd-enhanced T1-weighted (**a**) images at 9 years after the second surgery showed the recurrent tumor involving the vertebral bodies at the T5–7 levels. Sagittal T1-weighted (**b**), T2-weighted (**c**) and Gd-enhanced T1-weighted (**d**) MR images obtained 13 years after the second operation demonstrated that the recurrent tumor extended into the vertebral bodies in comparison to the images that were obtained at 9 years after the second operation. On axial T2-weighted (**e**) and Gd-enhanced T1-weighted (**f**) images obtained at 13 years after the second operation, the recurrent tumor was observed in the vertebral body and the epidural space at the T6 level

Postoperative course

The postoperative course went well and his symptoms were recovered but only slight sensory change remained in the body trunk and bilateral lower extremities. At 6 months after the surgery, CT revealed complete bone union with no implant failures and MRI showed the spinal cord was sufficiently decompressed at T6 level (Fig. 6). Follow-up imaging showed no recurrence at 2 years. JOA score is improved to 7 of 11 points.

Histopathological findings

A histopathological examination showed sheets of spindly cells surrounding numerous ectatic capillaries, so-called "staghorn vessels". Tumor cells had from mildly to deeply stained nuclear chromatins, but no anaplasia with scarce mitotic figures. On immunohistochemistry, the tumor cells were positive for vimentin and partially positive for HHF-35 and αSMA. CD34 expression was positive on the vascular endothelium. The Ki-67 index was less than 20%. Based on these findings, histopathological diagnosis was consistent with HPC.

Discussion

Primary osseous HPC of the spine is exceedingly rare. Only 15 cases of primary osseous HPC of the spine have been reported (Table 1). To our knowledge, there have only been two reported cases of recurrent primary osseous HPC of the spine [2, 12]. HPC is generally associated with a higher risk of local recurrence after surgical treatment unless the gross total resection of the tumor is performed [13]. However, the overall prognosis (i.e, the survival rate) is not necessarily poor if a histological examination reveals no malignancy [14]. In our literature review of studies on primary osseous HPC of the spine (Table 1), patients with malignant tumors tended to show worse overall survival. Both of the two reported cases of recurrent

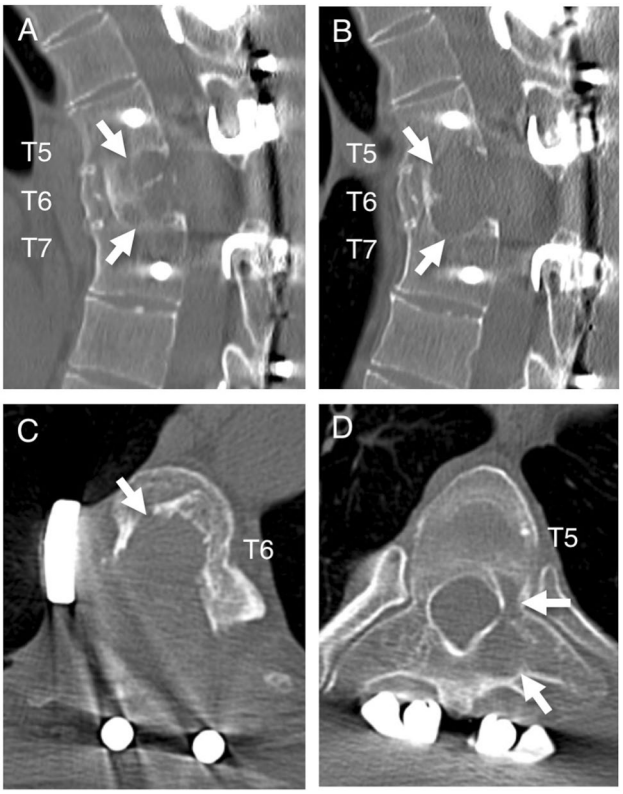

Fig. 3 CT scans obtained before the third operation. Sagittal (**a**) images at 9 years after the second surgery showed the recurrent tumor involving vertebral bodies at the T5–7 levels. Sagittal (**b**) images at 13 years following the second surgery demonstrated the recurrent tumor more extended into the vertebral bodies compared to that at 9 years after the second surgery. On axial (**c**, **d**) images at 13 years following the second surgery, the recurrent tumor was observed in the vertebral body at T6 level (**c**), and the lamina and the pedicle on the left side at T5 level (**d**)

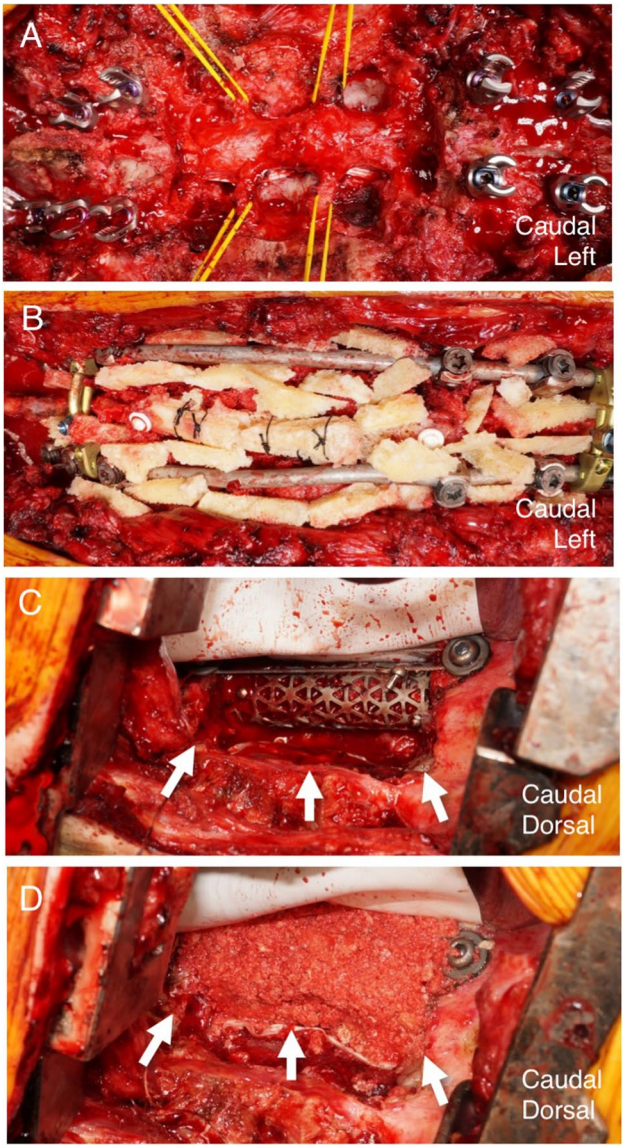

Fig. 4 Intraoperative photographs via the posterior (**a**, **b**) and anterior (**c**, **d**) approaches in the third operation. The posterior part of the tumor, which extended into the laminae, facet joints, transverse processes and the epidural space from T5 to T7 levels was totally removed (**a**). Posterior fusion using instrumentation with both autograft from the iliac bone and allograft was performed from T2 to T10 (**b**). The anterior part of the tumor in the vertebral bodies from T5 to T7 was totally removed and a mesh cylindrical cage was used for anterior reconstruction via an anterior approach (**c**). Both autograft from iliac bone and allograft were used abundantly inside and around the cylindrical cage (**d**)

primary osseous HPC of the spine had malignant histopathology [2, 12]. In the present case, the histopathological examination revealed that the tumor was not malignant. Although the tumor re-recurred following the second surgery, after the third operation to perform total resection without adjuvant therapy, there was no local recurrence or metastasis.

Recent studies have advocated gross total resection for HPC [13, 15]. The extent of resection is increasingly correlated with improved long-term and recurrence-free survival [13, 16]. A meta-analysis demonstrated that the 10-year survival rate of patients undergoing total resection was 69%, while that of patients who underwent subtotal resection was 44% [15]. In the present case, we totally excised the recurrent tumor in a two-stage operation. As a result, there was no recurrence after the third surgery and the patient made a sufficient recovery from his symptoms. Taking these facts into consideration, total resection might be effective for achieving local control and preventing symptomatic recurrence.

HPC is a highly vascular tumor. The adequate and safe resection of HPC of the spine can be difficult because of the risk of excessive bleeding from the tumor during the surgery [1, 7]. Many reports have suggested that preoperative embolization reduces the amount of intraoperative bleeding from hypervascular tumors of the spine [1, 8, 17, 18]. Reducing the intraoperative bleeding can improve

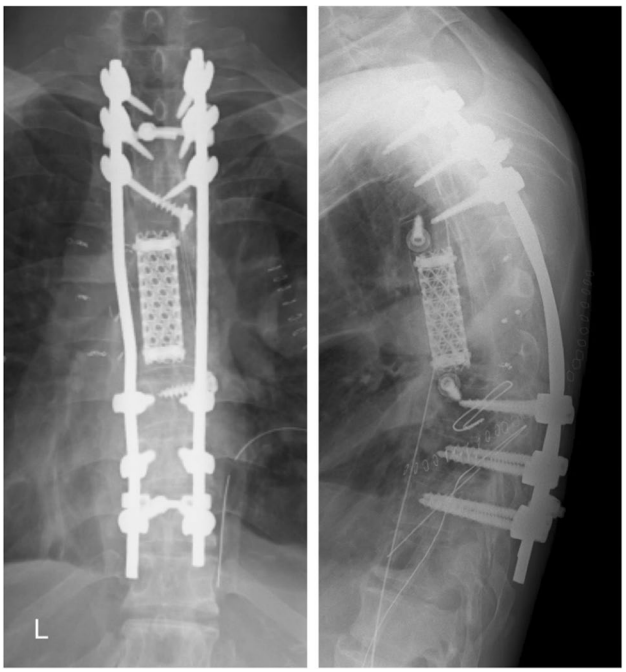

Fig. 5 Radiographs obtained after the third operation

the visibility of the operative field and make it both easier and safer to remove the tumor [19]. Among the previous studies on primary osseous HPC of the spine, there are two reported cases in which preoperative embolization was performed [1, 12]. In the present case, preoperative embolization was performed in the second and third operations and the amounts of intraoperative bleeding were reduced in comparison to the initial operation. Preoperative embolization might be a useful procedure for reducing intraoperative blood loss and for performing the safe and adequate tumor excision of HPC of the spine.

There is a high risk of implant failure (i.e, rod breakage, cage breakage, screw back-out, and cage subsidence) following spinal reconstruction after total spondylectomy at multiple levels [20–22]. Meticulous preparation of the bone graft site and a longer posterior fixation are recommended for the prevention of instrumentation failure [20]. Osseous fusion between the grafted bone in the cage and the vertebral bodies can prevent cage subsidence and consequently reduce the risk of instrumentation failure. In the present case, we transplanted a large amount of bone on both the anterior and posterior sides using autologous and allogenic grafts to achieve secure spinal fusion. Consequently, no instrumentation failure was observed and solid spinal fusion was achieved after the operation. The abundant transplantation of both autologous bone and allogenic bone might be useful for ensuring spinal fusion and reducing the risk of implant failure after total spondylectomy at multiple levels.

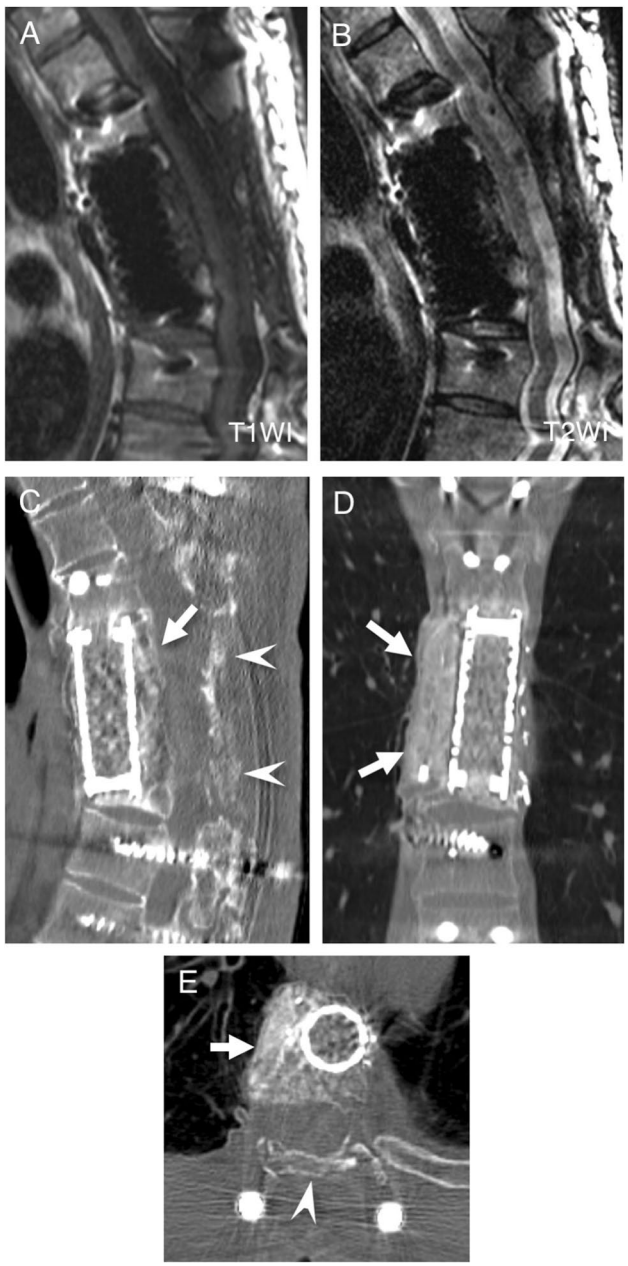

Fig. 6 MR images and CT scans obtained after the third operation. In sagittal T1-weighted (**a**) and T2-weighted (**b**) images, cerebrospinal fluid was detected around the spinal cord from T5 to T7 and the spinal cord was sufficiently decompressed. Sagittal (**c**), coronal (**d**) and axial (**e**) CT scans showed that anterior spinal fusion at the lesion was appropriately achieved (arrows) and longitudinal bone formation was observed on the posterior side of the spine (arrowheads)

Conclusions

To the best of our knowledge, this is the first reported case of recurrent osseous HPC of the thoracic spine that was successfully treated by total spondylectomy at multiple vertebral levels and spinal reconstruction. Total spondylectomy

Table 1 Summary of reported cases of primary osseous HPC in the spine

References	Age (years)	Sex	Level	Initial treatment	Adjuvant therapy	Histology	Recurrence	Follow-up/outcome
Gerner et al. [6]	62	M	L5	Biopsy	Postop. RT	ND	ND	ND
Wold et al. [2]	33	F	ND	Surgery	None	Malignant	26 years	31 years/died
	37	F	Sacrum	Surgery	RT	Malignant	ND	5 years/died
	38	M	ND	Surgery	None	Benign	ND	7 years/alive
	47	F	Sacrum	Surgery	RT	Malignant	ND	1.5 years/died
	62	F	Sacrum	ND	ND	Malignant	ND	ND/died
Tang et al. [12]	19	M	L2	Surgery	Preop. RT + CT	Grade II–III	3 years	4 years/alive with disease
	43	F	Sacrum	Surgery	Preop. RT + CT	Grade II	None	7 months/alive, tumor free
Kozlowski et al. [3]	13	M	Sacrum	Surgery	ND	ND	ND	ND
Lin et al. [5]	16	F	C2	Surgery	None	Benign	None	17 months/alive, tumor free
Boriani et al. [9]	ND	ND	ND	Surgery	None	Malignant	ND	ND
Kumar et al. [8]	16	F	T4–5	Surgery	Postop. RT	Malignant	None	9 months/alive, tumor free
Zentar et al. [4]	42	F	Sacrum	Surgery	Postop. RT	Benign	None	5 months/alive, tumor free
Ren et al. [1]	54	M	T10	Surgery	Postop. RT	Benign	None	18 months/alive with disease
Ramdasi et al. [18]	28	M	C3	Surgery	Postop. RT	Grade II	None	1 years/alive, tumor free
Present report	25	M	T6	Surgery	None	Benign	5 and 18 years	20 years/alive

Age age (years) at initial surgery, *F* female, *M* male, *Level* level of lesion, *Recurrence* time to recurrence after the initial surgery, *RT* radiation therapy, *CT* chemotherapy, *ND* no description

and spinal reconstruction with instrumentation might be useful for performing the safe and adequate excision of recurrent HPC of the spine. However, the patient should be closely monitored to detect local recurrence and the malignant degeneration of the tumor after surgery.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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