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Intradural extramedullary thoracic spinal cord hemangiopericytoma: A case report and review of literature

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Abstract

Hemangiopericytoma of the central nervous system are rare, and seldom occur in the spine. We describe here a case of primary intradural extramedullary hemangiopericytoma in the thoracic spine and review previously reported cases and treatment. A 57-year-old man experienced numbness and a tingling sensation below the umbilicus. He had decreased sensation of pain, temperature, light touch, and tactile sense below the umbilicus. His knee reflexes were hyperactive bilaterally. Magnetic resonance imaging revealed a mass at T5–6. The patient underwent laminectomy with en-bloc tumor removal and follow-up radiation with no local recurrence or other spinal metastasis over the following 2 years. Regardless of the subtype of spinal hemangiopericytoma, complete surgical removal and radiotherapy appear to be the treatment of choice.

INTRODUCTION

Hemangiopericytoma, a condition first reported by Stout, and Murray in 1942, is a rare tumor of uncertain origin.[1] This type of tumor originates in blood vessels and spreads rapidly; it has been reported to be malignant, because it spreads to extracranial organs, such as the liver and bones, if it arises in the central nervous system. Intracranial primary tumors occur at a frequency of 1%.[2] Reports of intradural hemangiopericytoma in the spinal cord and spinal column are rare, and most reports describe cases that develop from epidural organs, including the spinal column.[3-8] Of the 60 previously reported cases of hemangiopericytoma, 11 were located in the intradural extramedullary (IDEM) region.[2-4,9-12] We report our experience of a rare case of spinal intradural extramedullary hemangiopericytoma and review the literature on reported cases and the treatment of this tumor.

CASE REPORT

A 57-year-old man complained of a tingling sensation and numbness in his left lateral thigh, for which he had not received any treatment over the course of 1 year. Because the symptoms persisted, he was admitted to our hospital. Upon admission, neurological examination revealed that he had numbness and a tingling sensation extending from below the umbilicus to both feet. He had decreased sensation of pain, temperature, light touch, and tactile sense. Both patellar tendon reflexes were hyperactive. All other findings were normal. Preoperative magnetic resonance imaging (MRI) revealed a homogeneously well- enhanced mass at T5-6 on T1-weighted gadolinium enhancement imaging. Enhanced lesions accompanied spinal cord edema around the spinal cord in this region (Figure 1).

Initially, a T5-6 total laminectomy was performed under a microscope. After the dura was opened, an irregular tumor was revealed. The T5-6 tumor adhered to the posterior and left lateral aspects of the cord. The tumor showed abundant vascularity. The boundaries between spinal cord and tumor were not clear. The intradural tumor was gently dissected from the spinal cord and was totally excised. Postoperative MRI showed that the well- enhanced mass at T5-6 had been completely removed (Figure 2).

Pathological examination of the resected tumor demonstrated relatively monomorphic oval- to spindle- shaped cells with densely stained nuclei. The blood vessels were abundant and variable in size. Tumor cells stained positive for actin and CD34 and negative for P53 and Ki-67 (Figure 3). These histological findings are characteristic of hemangiopericytoma. Immediately after the operation, the patient complained of surgical site pain and persistent tingling sensation and numbness in the left lateral thigh, but after 12 hours, his symptoms were improved. The patient was followed- up 1 year after surgery, and the tumor had not recurred as of 2 years postoperatively (Figure 2).

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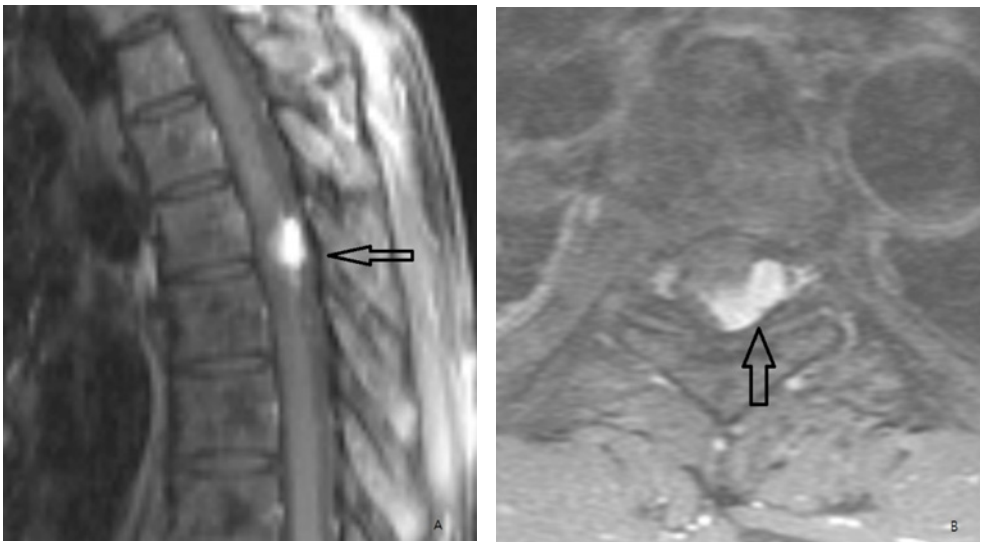

Figure 1. Preoperative sagittal T1-weighted magnetic resonance imaging with contrast enhancement shows a well-enhanced mass displacing the cord interiorly (A). An axial magnetic resonance image shows the homogeneously enhanced mass displacing the cord anteriorly and to the right (B).

DISCUSSION

Hemangiopericytoma of the central nervous system often arises intracranial partly, and often recurs after removal. It is a malignant tumor with remote metastasis.[2] Ecker *et al*. reported that patients with hemangiopericytoma in the central nervous system who were treated before 1990 had a 5-year survival rate of 67% and a 10-year survival rate of 40%, while the 5-year survival rate of such patients treated after 1990 had increased to 93%.[11] Schiariti *et al*. also showed that increased radiation therapy after surgery improved the 5-year survival rate to 93% and the 10-year survival rate to 67%, suggesting the effectiveness of postoperative radiation therapy.[13] However, most of these findings are related to intracranial hemangiopericytoma. Spinal intradural hemangiopericytoma is very rare, and only 19 cases, including the present case, have been reported (Table 1).[5-7,9,10,12,14-19] There were more cases of spinal intradural hemangiopericytoma among men (13 cases) than among women (7 cases), and the average age of these patients was 46 years. As a primary tumor of the spinal cord, hemangiopericytoma are rare and they are difficult to diagnose using only MRI.

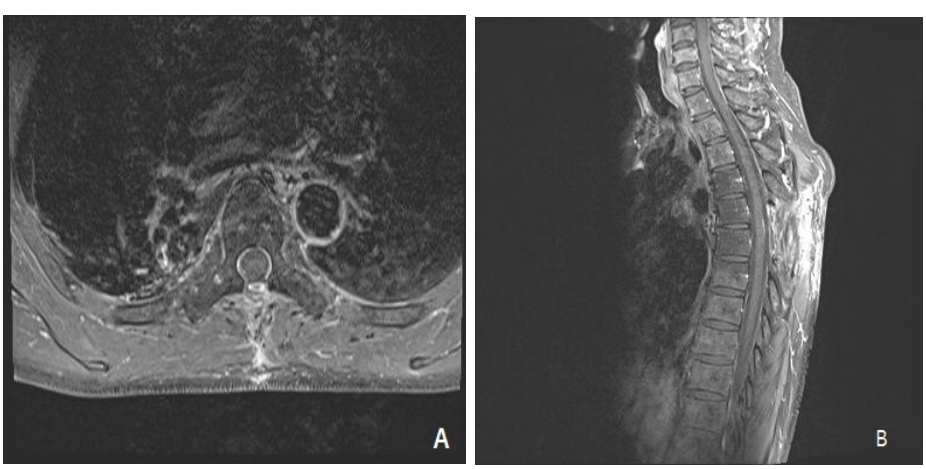

Figure 2. Postoperative axal T1-weighted contrast enhanced magnetic resonance image shows that the mass has been completely removed (A). One year postoperative follow-up magnetic resonance image shows no recurrence of the mass (B).

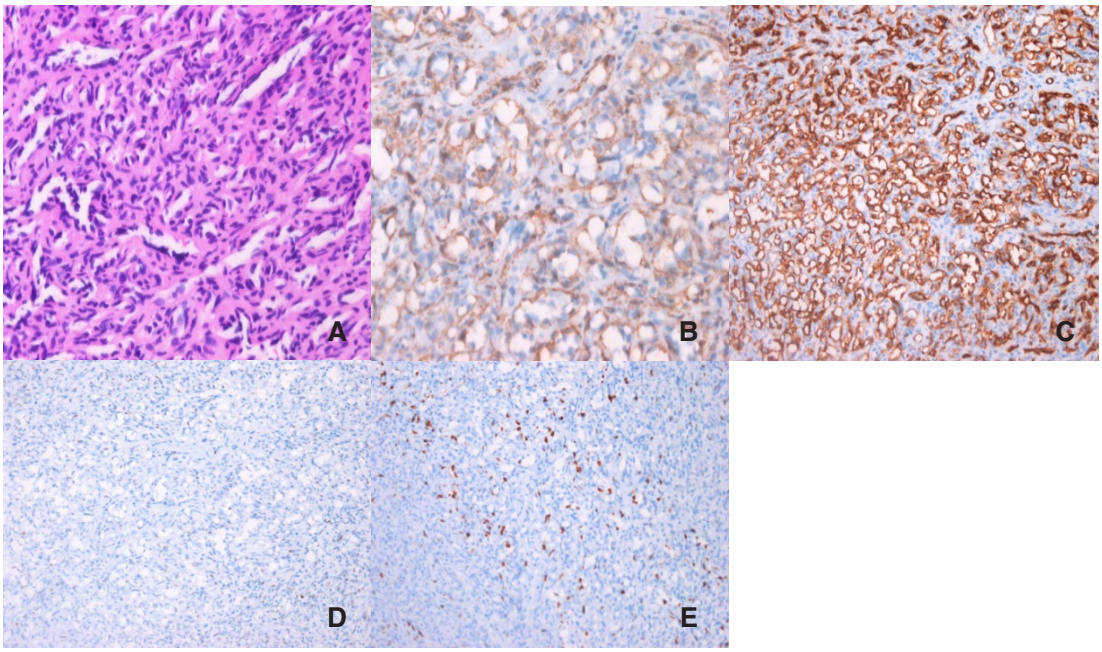

Figure 3. Tumor cells show monomorphic proliferation of spindle-shaped cells (A), which positive for actin (B) and CD34 (C). Tumor cells stained negative for P53 (D) and Ki-67(E). This staining pattern was consistent with hemangiopericytoma.

Table 1: Summary of literature reports on intradural spinal hemangiopericytomas

Author	Age/Sex	Level	Treatment	Postoperative recurrence
Kruse[12]	53/M	C3	GTR + RT	+(at 3 years)
Pitlyk *et al*.[16]	60/M	C4	GTR	Unknown
	49/F	C3	GTR	-(at 10 years)
	39/M	T8	GTR	+(at 9 years)
Dufour *et al*.[6]	45/M	C	GTR	-(at 2 years)
	18/F	T	STR + RT	-(at 4.6 years)
	43/F	T	GTR	-(at 4.1 years)
	38/M	T	GTR	-(at 12.6 years)
Ciappetta *et al*.[10]	48/M	C4	GTR	+(at 6 years)
Betchen *et al*.[3]	31/M	L4	GTR	-(at 6 months)
Kashiwasaki *et al*. [7]	31/F	T6	GTR	Unknown
Fitzpatrick *et al*. [14]	54/M	L4	GTR	Unknown
Moscovici *et al*.[15]	20/F	T9-10	GTR	-(at 2 years)
Ackerman *et al*.[9]	58/M	T9-T11	GTR	No
Torigoe *et al*.[19]	46/F	T7	GTR	-(at 2 years)
Lee *et al*.[18]	21/M	C1-C2	GTR	No
Someya *et al*.[17]	68/F	C	GTR + RT	-(30 months)
Chou *et al*.[5]	80/M	T	GTR	-(at 3 years)
Present study	57/M	T5-6	GTR + RT	-(at 2 years)

+, positive for recurrence; -, negative for recurrence
GTR; gross total resection
STR; subtotal resection
RT; radiation therapy

The presence of flow- void signals on the surface of the spinal cord is regarded as a characteristic feature of theses tumors. Evaluating the blood vessels on the spinal cord surface using MRI can assist in the diagnosis of hemangiopericytoma. In addition, histologically, these tumors are similar to solitary fibrous tumors but prognosis of these two tumor type are significantly different. Importantly, hemangiopericytoma cells have a low positivity for CD34 staining, while high solitary fibrous tumors have a high positivity for patch-type dyeing.[4] As intradural spinal cord hemangiopericytoma is rare, its malignant nature and the effectiveness of radiation therapy are unclear. Anticancer therapy has been attempted for intracranial hemangiopericytoma without success. However, in 1 case in which 6 cycles of doxorubicin was used, the tumor was reduced.[13] To date, no case of chemotherapy for intradural spinal cord hemangiopericytoma has been reported, and thus the effectiveness of this approach remains uncertain. According to some reports, tumors recurred locally within 6 years after surgery, indicating that intradural spinal cord hemangiopericytoma has a high local recurrence similar to intracranial hemangiopericytoma. Therefore, adjuvant therapy after surgery has been considered, while radiation therapy has been recommended only for cases with postoperative recurrence. The reasons for the cautious use of radiation therapy include reducing the exposure dose of the spinal cord, the lack of data on radiation necrosis of the spinal cord, and the uncertain effectiveness of radiation therapy in this condition. Recently, it has become possible to reduce the exposure of normal tissues in the spinal cord to radiation therapy by using approaches such as intensive-modulated-radiation therapy, which is likely to improve therapeutic effect.[20,21] Although further data on treatment outcomes need to be accumulated.

In conclusion, hemangiopericytomas are rare tumors. Imaging diagnosis shows that hemangiopericytomas on the intradural thoracic spinal cord has specific features, but that it is difficult to discern this tumors from other tumors before surgery. Non-surgical interventions have yielded inconclusive outcomes to date. Greater awareness of these tumors may facilitate the diagnosis, decrease local recurrence, and improve patient outcomes.

DISCLOSURE

Conflicts of interest: None

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