Malignant Fibrous Histiocytoma of the Thoracic Spine Manifesting as Rapidly Progressive Compressive Myelopathy

—Case Report—

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Abstract

A 75-year-old woman presented with an extremely rare case of malignant fibrous histiocytoma (MFH) arising in the thoracic spine, causing compression fracture and manifesting as rapidly progressive compressive myelopathy resulting in progressive gait disturbance and weakness of the bilateral legs. Computed tomography and magnetic resonance imaging demonstrated a solid enhanced intravertebral mass destroying the vertebral bodies at the T7 and T8 levels. Maximum resection of the lesion was performed through wide laminectomy from T7 through T9, followed by vertebroplasty using calcium phosphate paste and posterior fixation with a spinal fixation system. Histological examination showed proliferation of pleomorphic spindle-shaped cells having large irregular nuclei with hyperchromasia with storiform pattern and collagenous stroma, consistent with a diagnosis of MFH. This case illustrates the possibility of rapid maximum decompression surgery from the posterolateral side if MFH manifests as progressive transverse myelopathy.

Key words: malignant fibrous histiocytoma, thoracic spine, transverse myelopathy, oncologic emergency

Introduction

Malignant fibrous histiocytoma (MFH) is a rare primary bone tumor that arises from primitive mesenchymal cells capable of multi-directional differentiation,^{6,11,17} probably the histiocytes located in the metaphysis or diaphysis of the long bones.^{2,4,5,9,16} MFH is aggressive with high rates of local recurrence and metastasis,^{2,5,7,22} and the prognosis is poor with 5–20 year survival rates of 28–33%.^{2,4,9} Men are more frequently affected than women and occurrence peaks in the fifth decade.^{4,16,24} The vast majority of MFH cases occur in the long bones, such as femur, tibia, and humerus. MFH with spine involvement is rare, but includes cases originating from the paravertebral tissue with secondary extension into the spinal canal, and metastases to the spine.^{1,4,8–10,12–15,18–23}

Here we describe a case of MFH in a 75-year-old woman referred for treatment of a thoracic spine mass manifesting rapidly progressive weakness of the bilateral legs.

Case Report

A 75-year-old woman without a previous medical history presented to a local hospital with complaints of back pain and rapidly progressive weakness of the bilateral legs. Computed tomography (CT) demonstrated compression

fractures of the T6, T7, and T8 vertebral bodies. Her symptoms were progressive and metastatic vertebral bone tumor was suspected, so the patient was referred to our hospital.

On admission, neurological examination revealed tenderness to percussion over the thoracic spine at the T6, T7, and T8 levels. Motor power was 3/5 (manual muscle test) in the bilateral lower extremities. She could not walk. Sensation was decreased to light touch and pin prick in and below the T8 dermatome level. Vibration sense was also decreased in the bilateral lower extremities. Knee and ankle jerks were increased and the bilateral planter extensor reflexes were positive. Preoperative CT and magnetic resonance (MR) imaging confirmed the presence of a mass destroying the vertebral body and extending into the spinal canal at the T6, T7, and T8 levels, appearing as hyperintense on both T₁- and T₂-weighted MR images (Figs. 1 and 2). The thoracic spine showed kyphotic change and the thoracic cord was remarkably involved. Metastatic vertebral tumor was first considered, but systemic investigations using positron emission tomography, bone scintigraphy, chest CT, and abdominal ultrasound tomography detected no tumor other than that of the thoracic spine.

Emergent decompressive surgery was performed through a posterior approach. Wide laminectomy from T6 through T8 was performed. The tumor appeared as a whitish, elastic hard, and hypovascular mass, tightly adhered

Fig. 1 Axial computed tomography scan demonstrating an osteolytic mass in the T7 vertebral body extending to the spinal canal.

Fig. 3 Photomicrograph showing proliferation of pleomorphic spindle-shaped cells having large irregular nuclei with hyperchromasia with storiform pattern and collagenous stroma. Hematoxylin and eosin stain, original magnification $\times 200$.

Fig. 2 Sagittal T₁-weighted (left), T₂-weighted (center), and T₁-weighted with contrast medium (right) magnetic resonance images demonstrating compression fractures in the T7 and T8 vertebral bodies caused by the intravertebral tumor.

Fig. 4 Sagittal T₁-weighted (left), T₂-weighted (center), and T₁-weighted with contrast medium (right) magnetic resonance images demonstrating resection of the tumor and decompression of the thoracic cord.

to the dura mater, and compressing the thoracic cord from the left ventrolateral side. The mass did not extend into the adjacent paravertebral soft tissue. The tumor did not bleed much, so near total resection was achieved. Vertebroplasty and posterior instrumentation fusion were performed using calcium phosphate and the Legacy[®] spinal fixation system (Medtronic Sofamor Danek, Co., Ltd., Osaka). No blood transfusion was necessary. Histological examination showed proliferation of pleomorphic spindle-shaped cells having large irregular nuclei with hyperchromasia with storiform pattern and collagenous stroma (Fig. 3), consistent with a diagnosis of MFH. Postoperative MR imaging demonstrated resection of the tumor and decompression of the thoracic cord (Fig. 4).

After the surgery, weakness of the lower extremities resolved completely and the patient could walk without a cane. The patient received 50 Gy local radiotherapy to the T6–T8 vertebral bodies and was discharged home.

Discussion

Primary MFH arising within the spine is quite rare, with only reported 15 cases (Table 1).^{1,4,8–10,12–15,18,19,22,23} Most cases occurred in the thoracic spine and manifested as back pain.^{8,10,12,15,18,22,23} Neuroimaging showed purely lytic,

expansive lesions with well-defined margins, usually with cortical destruction and soft tissue invasion, but without periosteal reaction or new bone formation.¹⁾ The prognosis for primary spine MFH is dismal, and about half of the patients were dead within 15 months.^{10,14,15,18,19)}

Differential diagnosis from metastatic lesion is always important for vertebral bone tumor. However, no characteristic neuroradiological features of spinal MFH have been reported, so histological diagnosis and exclusion of other lesions by systemic investigation are necessary. In our case, preoperative imaging and intraoperative findings showed that the tumor was not continuous with the paravertebral soft tissue, and systemic investigation detected no other lesions, so our final diagnosis was MFH of spinal origin.

The present case of spinal MFH manifested as rapidly progressive paraparesis so emergent decompression surgery was essential. The tumor is aggressive with a high rate of local recurrence, so total resection with vertebrectomy is ideal. However, in an urgent situation, maximum tumor resection with vertebrectomy is not always possible for all vertebral tumors. Many of the reported cases of spine MFH underwent wide laminectomy with biopsy as

Table 1 Summary of reported cases of primary malignant fibrous histiocytoma arising in the spine

Author (Year)	Age (yrs)/ Sex	Location	Surgical treatment	Other treatment	Outcome
Newland et al. (1975) ¹⁸⁾	74/M	T8	laminectomy + biopsy	RTx	dead (postop day 1)
Kellett and Dearnaley (1976) ¹³⁾	36/M	lumbar	laminectomy + biopsy	CTx	terminal stage
Dahlin et al. (1977) ⁴⁾	??	sacrum	ND	ND	ND
Teddy and Esiri (1979) ²³⁾	52/M	T10-T11	laminectomy + biopsy	none	dead (6 mos)
	45/F	T11	laminectomy + biopsy	RTx	alive
	46/M	T3-T6	laminectomy + biopsy	RTx	alive
Helle et al. (1983) ⁸⁾	42/M	T10 to sacrum	gross total resection	RTx, CTx	dead (2 mos)
Rechtime et al. (1984) ¹⁹⁾	50/M	C2	none	none	dead (2 mos)
Huvos et al. (1985) ⁹⁾	??	sacrum	ND	ND	ND
Itoyama et al. (1987) ¹⁰⁾	65/F	T1-T2	subtotal resection	RTx	dead (2 mos)
Bidwell et al. (1987) ¹⁾	17/F	L5	biopsy	ND	ND
Kepes (1989) ¹⁴⁾	27/F	L4	partial resection	none	dead (a few mos)
Sturm et al. (1992) ²²⁾	27/M	T8	total resection	RTx	alive (16 mos)
Maillefert et al. (1997) ¹⁵⁾	40/F	T12, L2, L3, L5	biopsy	RTx, CTx	dead (15 mos)
Karantanas et al. (2000) ¹²⁾	46/F	T10-T11	laminectomy + biopsy	CTx	alive (15 mos)
Present case	75/F	T6-T8	gross total resection	RTx	alive (5 mos)

CTx: chemotherapy, ND: not described, postop: postoperative, RTx: radiation therapy.

the initial treatment,^{1,12,14,15,22,23)} with subsequent total vertebrectomy in one case.²¹⁾ In our case, the tumor did not bleed much, so near total resection of the tumor was possible, followed by vertebroplasty using calcium phosphate paste from the posterolateral approach. In an emergent situation such as our case, decompression of the spinal cord and histological diagnosis are essential to achieve a favorable neurological outcome. The present case suggests that maximum tumor resection might be possible from the posterior approach, and the histological diagnosis can be confirmed as soon as the wide laminectomy is completed. If the intraoperative histological diagnosis is MFH, maximum tumor resection should be considered from the posterior approach to enhance the effect of postoperative chemotherapy or radiotherapy and improve the prognosis.

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