Spinal Malignant Fibrous Histiocytoma producing Cord Compression

—Case Report—

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Abstract

Malignant fibrous histiocytoma is a well known neoplasm that probably originates from histiocytes and involves not only soft tissue but also bone. The favorite sites of this tumor are the extremities; the central nervous system is rarely involved. Recently we treated a 65-year-old female who presented with neurological symptoms of spinal cord compression at the level of Th1 and Th2. A computed tomographic scan revealed a huge extradural tumor in that area. The tumor was subtotally removed, and the histological diagnosis was malignant fibrous histiocytoma. Although eight similar cases have been reported, this is the first reported case from Japan, according to our review of the literature.

Key words: spinal tumor, malignant fibrous histiocytoma, computed tomography

Introduction

Malignant fibrous histiocytoma was first described by Kauffman and Stout¹⁰⁾ and O'Brien and Stout.¹⁹⁾ Since their original reports malignant fibrous histiocytoma attracted special interest as a malignant neoplasm of soft tissue. Since 1972, when Feldman and Norman²⁾ reported nine cases of malignant fibrous histiocytoma primarily involving bone, it has also become known as a malignant tumor of bone.

Most malignant fibrous histiocytomas arise in the extremities, particularly the femur and tibia; involvement of the central nervous system is rare. Recently we encountered a 65-year-old female who developed neurological symptoms of spinal cord compression at the level of Th1 and Th2. The histological examination following surgery revealed the tumor to be a malignant fibrous histiocytoma. Only eight similar cases have so far been reported, and ours may be the first such case reported from Japan.

Case Report

A 65-year-old female began complaining of pain in the right shoulder and back in May, 1985. On July 3, she visited a local orthopedic hospital complaining of lumbago. However, neither a lumbar spine x-rays nor a bone scintigram with ^{99m}Tc-MDP showed no abnormalities. On Aug. 3, 3 days prior to visiting our hospital, she suddenly complained of gait disturbance.

The admission neurological examination indicated motor weakness of the right upper and bilateral lower extremities and increased deep tendon reflexes, especially in the lower extremities. Also, there was general hypesthesia below the level of Th1-2. Serum calcium, phosphate, and alkaline-phosphatase were all within the normal range. On the second day of hospitalization her motor weakness worsened in the lower extremities and she developed urinary incontinence. Metrizamide myelography revealed a complete block at the level of Th1-2, where spinal x-rays disclosed an absence of spinous processes (Fig. 1). A computed tomographic (CT) scan revealed a huge epidural tumor at the level of Th1-2 as well as partial destruction of the spinous processes and vertebral

Fig. 1 Metrizamide myelography showing complete block at the level of Th1-2 and absence of spinous processes at Th1-2.

Fig. 2 Metrizamide CT myelography showing a huge epidural tumor at Th1-2. V: vertebral body, R: rib, T: tumor.

bodies. The spinal cord was compressed anteriorly by the tumor (Fig. 2).

Emergency surgery was performed, with the work-

Fig. 3 A, B: Photomicrographs of the tumor specimen showing pleomorphic tumor cells, spindle-shaped cells streaming in a fascicular pattern, multinucleated giant cells, many mitoses, and some tumor cells phagocytosing lymphocytes. HE stain, $\times 100$ (A), and $\times 200$ (B). C: PAP assay with $\alpha 1$ -antichymotrypsin showing a positive response by some tumor cells. PAP method, $\times 200$.

ing diagnosis of metastatic spinal tumor, and a huge epidural tumor about $5 \times 4 \times 4$ cm in size was subtotally removed. The tumor had completely

destroyed the laminae at the Th1-2 level, and further laminectomy was not attempted. The tumor tightly adhered to the dura, but infiltration into the surrounding bone and soft tissue was not obvious, and the tumor was not so hemorrhagic. We suspected that the tumor extended upward and downward along the dural surface of the spinal canal.

Histological examination of the specimen showed pleomorphic tumor cells, spindle-shaped cells, and multinucleated giant cells. The spindle-shaped cells streamed in a fascicular pattern, but did not show the so-called "storiform or cartwheel" configuration peculiar to malignant fibrous histiocytoma. Many mitoses were observed, and some tumor cells exhibited phagocytosis of lymphocytes (Fig. 3A, B). A peroxidase antiperoxidase assay with α 1-antichymotrypsin yielded positive results in some tumor cells (Fig. 3C). The periodic acid-Schiff stain and iron stain (Berlin blue) were both negative. These histological findings are diagnostic for malignant fibrous histiocytoma.

She received postoperative radiation therapy (40 Gy). Her motor weakness improved markedly. However, about 2 months after discharge she died of a respiratory disorder; an autopsy was not permitted.

Discussion

In the 1960s Kauffman and Stout¹⁰⁾ and O'Brien and Stout¹⁹⁾ first described malignant fibrous histiocytoma and characterized it as a malignant soft-tissue tumor probably originating from histiocytes. In the 1970s many papers concerning this neoplasm were published.^{3,11,15,23,25,27)} After 1972, when Feldman and Norman²⁾ reported nine cases of malignant fibrous histiocytoma primarily involving bone, it also excited considerable interest as a malignant bone tumor.^{1,6-8,16-18,24,28)}

The characteristic histological appearance of malignant fibrous histiocytoma includes histiocyte-like cells that perform phagocytosis, multinucleated giant cells, foamy cell transformation, and fibroblast-like cells, which are involved in the production of collagen. While the presence of both histiocyte-like and fibroblast-like cells in these tumors gives them a complex histological appearance,²⁴⁾ it is well known that the histological makeup of a tumor does not always determine its biological behavior.^{23,25)} In 1963 Ozzello *et al.*²⁰⁾ confirmed that histiocytes change into fibroblasts (facultative fibroblasts) *in vitro*, and since then it has generally been believed that fibro-

Table 1 Malignant fibrous histiocytoma primarily involving the spine

Author (Year)	Age/Sex	Location	Treatment	Recurrence	Metastasis	Outcome (Interval after diagnosis)
Newland <i>et al.</i> (1975) ¹⁸⁾	74/M	Th8	radiation, surgery	?	?	died (on post-op day 1)
Kellett <i>et al.</i> (1976) ¹²⁾	36/M	lumbar nerve roots	biopsy, chemotherapy	suspected	to testis	terminal stage
Kepes (1979) ¹³⁾	27/F	L4	partial resection	?	?	died (a few months)
Teddy <i>et al.</i> (1979) ²⁶⁾	52/M	Th10-Th11	biopsy	+	to lungs, mesentery, peritoneum	died (6 months)
"	45/F	Th11	biopsy, radiation	+	?	alive
"	46/M	Th3-Th6	biopsy, radiation	+	?	alive (2 months)
Helle <i>et al.</i> (1983) ⁵⁾	42/M	Th10-sacrum	gross total excision, radiation, chemotherapy	+	to axillary and inguinal region	died (2 months)
Rechtine <i>et al.</i> (1984) ²²⁾	50/M	C1	posterior occipito- cervical fusion	?	multiple systemic lesions	died (2 months)
Present case	65/F	Th1-Th2	gross total excision, radiation	suspected	?	died (2 months after discharge)

Th: thoracic, L: lumbar, C: cervical.

blast-like cells are differentiated from histiocyte-like cells.¹⁰⁾ Recent studies involving ultrastructural techniques have suggested that both histiocyte-like and fibroblast-like cells are differentiated from common primitive stem cells.^{3,15,21,25,27)}

Malignant fibrous histiocytoma usually occurs in individuals in their 50s and 60s, and the incidence in males is twice that in females.^{15,23,25)} Soft tissue tumors commonly involve the extremities and the retroperitoneum, and bone tumors most often arise in the femur and tibia. Malignant fibrous histiocytoma rarely involves the central nervous system. Rare cases in which this tumor arose from the dura mater of the cranial cavity have been reported by Gonzalez-Vitale *et al.*,⁴⁾ Kalyanaraman *et al.*,⁹⁾ and Kepes *et al.*¹⁴⁾ There have been only eight reports describing spinal cord compression to our knowledge (Table 1).^{5,12,13,18,22,26)} The first concerned a tumor of a vertebral body that extended from the retroperitoneum. Rehtine *et al.*²²⁾ ascertained that the tumor they reported had widely metastasized.

With malignant fibrous histiocytoma of soft tissue, recurrence and metastasis to the lungs and/or regional lymph nodes are quite common, and the prognosis is very poor.^{14,27)} On the other hand, malignant fibrous histiocytoma that primarily involves bone seldom spreads to regional lymph nodes.⁶⁾ However, it frequently metastasizes to the lungs *via* the circulatory system and its prognosis is similarly dismal.^{2,8,24)} Most of the reported patients in whom the tumor primarily involved the spine died shortly after the onset of symptoms (Table 1) because of the high rate of recurrence, the difficulty of radical removal, and the apparent ineffectiveness of radiation therapy.

Osteosarcoma, fibrosarcoma, and metastatic carcinoma should be ruled out before making a diagnosis of malignant fibrous histiocytoma. In particular, differentiation from osteosarcoma may be very difficult.²⁴⁾ Because an autopsy was not performed on our patient, we could not be certain whether or not her spinal tumor was primary. However, the preoperative examination, which included radioisotope, produced no evidence that the tumor was metastatic from an original tumor elsewhere.

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