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Storiform-Pleomorphic Type of Multifocal Malignant Fibrous Histiocytoma of the Lumbar Spine

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Male, 67
Final Diagnosis: Malignant fibrous histiocytoma
Symptoms: Back pain • leg weakness • weight loss
Medication: —
Clinical Procedure: —
Specialty: —

Objective: Rare disease
Background: We present the extremely rare case of a 67-year-old male with malignant fibrous histiocytoma, arising in the lumbar spine, demonstrated with radiological and pathological studies.
Case Report: The patient and his relatives refused open surgical approach and we performed transpedicular vertebral corpus biopsy and vertebroplasty under spinal anesthesia. His pathological result was malignant fibrous histiocytoma. The spine is a very uncommon site for malignant fibrous histiocytoma.
Conclusions: The management of malignant fibrous histiocytoma relies on the combination of maximum decompression surgery, chemotherapy and radiotherapy. Total removal is unrealistic and diagnosis is difficult. The prognosis in terms of continuing neurological deficit after surgery appears to be poor.

MeSH Keywords: Histiocytoma, Malignant Fibrous • Lumbar Vertebrae • Prognosis

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Background

Malignant fibrous histiocytoma (MFH) is a soft-tissue sarcoma and a rare primary bone tumor that arises from primitive mesenchymal cells, being capable of multi-directional differentiation. Usually the histiocytes are located in the metaphysis or diaphysis of the long bones. MFH is aggressive with high rates of local recurrence and metastasis and the prognosis is poor, with 5–20 year survival rates of 28–33% [1,2]. Men are more frequently affected than women and occurrence peaks in the fifth decade [3,4]. The vast majority of MFH cases occur in the long bones such as the femur, tibia, and humerus. MFH with spinal involvement is rare, and the spine is a very uncommon site of localization. Here we describe a case of MFH in a 67-year-old man referred for treatment of a lumbar spine mass, manifesting rapidly progressive weakness of the legs and lower back pain.

Case Report

A 67-year-old man had lost 10 kg in weight in a month and presented with complaints of back pain and rapidly progressive weakness of the legs. Neurological examination show that motor power was global 4/5 (manual test) and sensation test scores decreased in the lower extremities. Knee and ankle jerks were increased and bilateral planter reflexes were indifferent. The patient had anemia (Hb. 7.74 g/dl. Hct. 24.4%), hyperglycemia (blood glucose level 282 mg/dl), and he had high levels of alkaline phosphatase and lactate dehydrogenase – ALP: 209 U/L (30–128 mean), LDH 1254 U/L (0–248 mean), respectively. His symptoms were progressive and a metastatic vertebral tumor suspected. Computed tomography (CT) demonstrated compression fractures of L2-3-4 vertebral bodies and L3 paraspinal mass (Figure 1). Magnetic resonance imaging (MRI) of lumbar spine demonstrated a mass in the right paravertebral area from L2-3-4. Axial MRI scan confirmed the extension of the tumor into the adjacent spinal canal through the neural foramina, forming an extradural mass which resulted in the compression of right nerve roots (Figure 2A, 2B).

The lumbar spine showed multifocal osteolytic change. A metastatic vertebral tumor was first considered but systemic investigating using CT, MRI, and scintigraphy detected no extra tumor.

The patient underwent a resection of the paravertebral tumor by transpedicular vertebral corpus biopsy and vertebroplasty under spinal anaesthesia. The tumor appeared as a whitish elastic, hard and hypovascular mass, tightly adhered to the transverse process and compressing the foramen from the right ventrolateral side. The tumor did not bleed much. Near total resection was achieved. Vertebroplasty was performed using calcium phosphate to palliate pain.

Figure 1. Computed tomography showing compression fractures of L2-3-4 vertebral bodies.

Histological examination showed proliferation of pleomorphic spindle-shaped cells with large irregular nuclei and hyperchromasia with storiform pattern and collagenous stroma consistent with a diagnosis of the storiform-pleomorphic type of MFH (Figure 3A–3C)

After surgery, weakness of the lower extremities did not resolve completely and the patient could not walk without support. He and his family refused radiotherapy and chemotherapy. He died 6 weeks later.

Discussion

Primary lumbar spine MFH is quite rare [1,5]. Most cases occurred in the thoracic spine and manifested as back pain [6,7]. 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. The prognosis for primary spine MFH is dismal, and about half of the patients were dead within 15 months [8,9].

Figure 2. (A, B) Magnetic resonance imaging showing the mass in the right paravertebral area from L2-3-4 and extension of the tumor into the adjacent spinal canal through the neural foramina, forming an extradural mass.

MFH has been categorized into five types. The histopathologic subtypes are storiform-pleomorphic, myxoid, inflammatory, giant cell, and angiomatoid variant. The MFH of the spine tends to extensively invade paraspinal structures at multiple spinal levels, with aggressive osteolytic destruction in the vertebrae resulting to locally huge-mass radiculopathy and myelopathy. Regardless of recent advancements in the diagnosis, treatment, and adjuvant therapies, due to its biologically aggressive nature it frequently recurs at the primary site and metastasizes, which leads to a worse prognosis.

Differential diagnosis from metastatic lesion is always important for the vertebral tumors. 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 continuous with the paravertebral soft tissue and systemic investigations for metastasis work-up detected no other lesions. Thus, our final diagnosis remained MFH of spinal origin.

This tumor has a high propensity for local recurrence, so total resection with the vertebrectomy is the ideal. However, in an emergency, maximum tumor resection with vertebrectomy is not always possible and decompression of the spinal cord, wide laminectomy, stabilization, and vertebroplasty are essential to achieve a favorable neurological outcome [1,5]. Maximum tumor resection is likely to improve the prognosis [5]. The effect of either chemotherapy or radiotherapy is presently unclear. Stabilization procedures including vertebroplasty and kyphoplasty have further allowed palliation of pain and symptom relief from compression fractures [1].

Prognostic factors known to correlate with survival in patients with MFH include tumor grade, depth, size, metastatic status, patient age, and histological subtype [10,11]. Favorable

Figure 3. (A–C) H&E, vimentin, and CD-68 staining showing storiform-pleomorphic type of MFH which had proliferation of pleomorphic spindle-shaped cells having large irregular nuclei with hyper chromopsia with storiform pattern and collagenous stroma.

prognostic factors include age less than 60 years, tumor size less than 5 cm, superficial location, low grade, the absence of metastasis, and myxoid subtype in histology [10,11].

Wide local excision of the skin for therapy is possible in most cases [12], but the present case illustrates that total removal

in the spine is unrealistic and diagnosis is difficult. The prognosis in terms of continuing neurological deficit after surgery appears to be poor. Our patient presented to us in a very late stage and his general condition was very poor. The patient refused radiotherapy and chemotherapy.

Conclusions

Prognostic factors of MFH are important, and early diagnosis and treatment is important for MFH, as in other malignant tumors. We diagnosed MFH with transpedicular biopsy from the vertebral body. We think that transpedicular biopsy may help establish the diagnosis in patients with poor general condition.

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