

Primary Malignant Fibrous Histiocytoma in Lumbar Vertebra: Case Report

Bel Omurunda Primer Malign Fibröz Histiositoma

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ABSTRACT A 71-year-old man with a clinical picture of a low back pain radiating to right leg caused by primer malignant fibrous histiocytoma of L2 vertebra was reported in this paper. Malignant fibrous histiocytoma is classified as soft tissue tumor for pleomorphic soft-tissue sarcoma showing no specific line of differentiation. Malignant fibrous histiocytoma most commonly involves the deep soft tissues of the extremities, mainly the thigh, with a predilection for men. However it is rarely seen in vertebrae. In our case, lung and multiple brain metastases were seen at the same time. The patient was operated for neural decompression and radiotherapy and chemotherapy were performed after diagnosis. The survival was only three months.

Key Words: Histiocytoma, malignant fibrous; low back pain; spine

ÖZET Bu makalede 71 yaşındaki hastada bel ve sağ bacağı yayılan ağrı kliniği ile ortaya çıkan L2 vertebrada olan primer malign fibröz histiositoma rapor edilmiştir. Malign fibröz histiositoma, yumuşak doku tümörleri içinde pleomorfik yumuşak doku sarkomu olarak sınıflandırılır ve herhangi bir spesifik diferansiyasyon göstermez. Primer malign fibröz histiositoma sıklıkla ekstremitelerin derin yumuşak doku kısmında ve özellikle uylukta görülmektedir. Malign fibröz histiositoma erkeklerde daha sık gözlenmektedir. Birincil vertebra tutulumu nadiren görülmektedir ve literatürde rapor edilen vertebra tutulumu çok azdır. Bu olguda aynı zamanda akciğer ve çoklu beyin metastazları tespit edilmiştir. Hasta nöral doku dekompresyonu için ameliyat edildi. Doku tanısından sonra radyoterapi ve kemoterapi tedavisi gören hastanın surveyi sadece üç ay sürmüştür.

Anahtar Kelimeler: Malign fibröz histiositoma; bel ağrısı; omurga

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Malignant fibrous histiocytoma (MFH) is a well recognized malignancy. It is classified by World Health Organization as soft tissue tumor for pleomorphic soft-tissue sarcoma showing no specific line of differentiation.^{1,2} MFH most commonly involves the deep soft tissues of the extremities, mainly the thigh, with a predilection for men.¹⁻³

Primary spinal bony involvement is exceedingly rare.²⁻⁴ Metastases occur predominantly in the lung (82%), lymph nodes (32%), liver and bone (15% each). We discussed clinical features, radiologic findings and mode of treatment in our MFH patient.

CASE REPORT

A 71-year-old man presented with complaint of radicular low back pain on his right leg for 15 days. Neurological examination revealed tenderness to percussion over the upper lumbar spine. Sensation was decreased to pinprick on the right leg beginning from L1 dermatome.

Motor strength was 4/5 in the right iliopsoas. Magnetic resonance imaging (MRI) of lumbar spine revealed an extradural tumor with widespread extension in right part of L2 corpus, pedicle, facet joint, paravertebral area, widening neural foramina (Figure 1, 2). Thorax radiography and computed tomography (CT) showed mediastinal and hilar lymphadenopathies in addition to 43 x 31 cm tu-



FIGURE 1: Sagittal T2-weighted turbo spin echo image of the lumbar spine showing a low signal intensity lesion in the L2 vertebra.

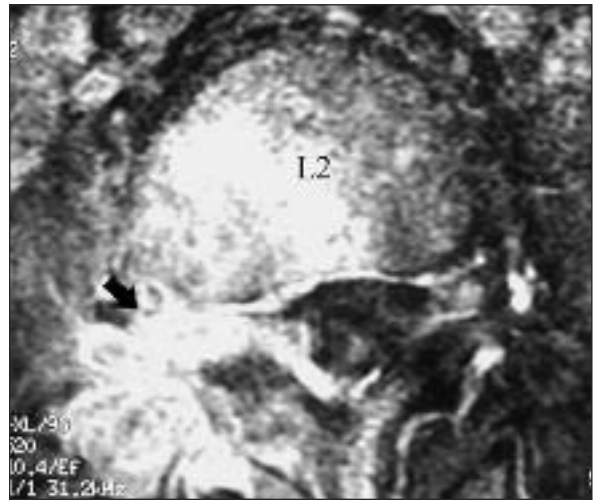


FIGURE 2: Neural foramina widening due to malignant fibrous histiocytoma (T1-weighted spin echo axial image with contrast).



FIGURE 3: Thorax CT showing lung metastases.

moral lesion in pleural baseline of right lung's upper lobe (Figure 3). In addition, whole body MRI for metastases revealed tumors in L2 vertebra, lung and brain (Figure 4). The brain MRI showed multiple brain metastases (Figure 5). As the patient had neural compression and intensive pain related to it, he underwent L2 hemilaminectomy and foraminotomy for decompression. Dural sac and nerve root were strictly covered with tumor tissue. Tumor was removed from epidural space and dural sac, and the nerve root was freed.

Histopathological examination revealed that there were cellular areas with giant cell formations and extensive necrosis areas in the tumoral lesion (Figure 6). Cellular areas of the tumor consisted of

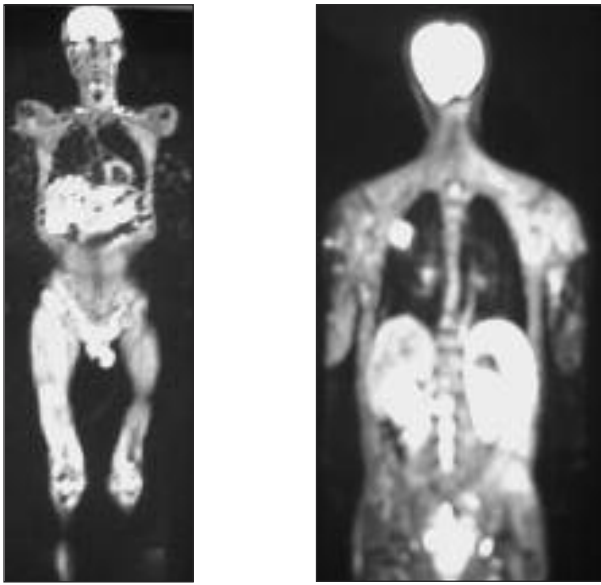
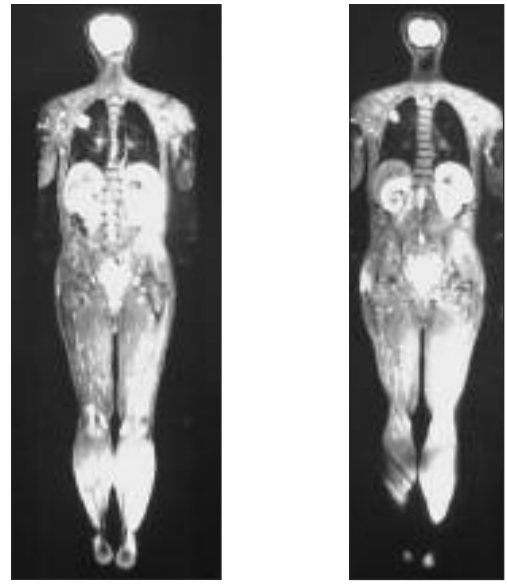


FIGURE 4: Whole body MRI showed only lung and brain metastases.



spindle shaped atypical cells with focally bizarre figures. Atypical mitosis was frequently noted. Immunohistochemical analysis showed diffuse and strongly cytoplasmic positivity with vimentin and low molecular weight cytokeratin (LMWCK) in cellular tumor areas while tumor cells did not show any positivity with carcinoembryonic antigen (CEA), desmin and high molecular weight cytokeratin (HMWCK). The MFH diagnosis was suggested on the basis of these findings. Radiotherapy and chemotherapy were performed after surgery; but prognosis was poorly despite treatment.¹ The patient died three months after the diagnosis.

DISCUSSION

Widening of the intervertebral foramen at the level of a tumor is suggestive of a benign lesion such as schwannoma and neurofibroma; rare malignant nerve sheath tumors have the same effect.³ MFH of bone was first described as a separate entity in 1972.⁵ It is an uncommon tumor seen in older age groups.⁵ The vast majority of osseous MFH occur in long bones, with the femur, tibia, and humerus accounting for approximately 75% of all cases, but the spine is a very uncommon site of localization.³ Only 12 primary spinal MFH cases have been described.¹⁻⁴ Pain relief is seen after laminectomy, but may recur with further tumor growth. Metastases

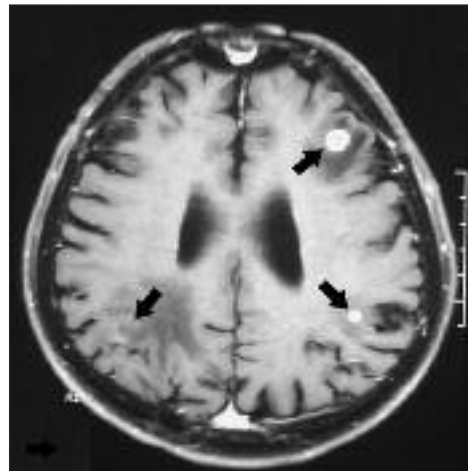


FIGURE 5: Brain metastases in T1-weighted MRI axial image with contrast.

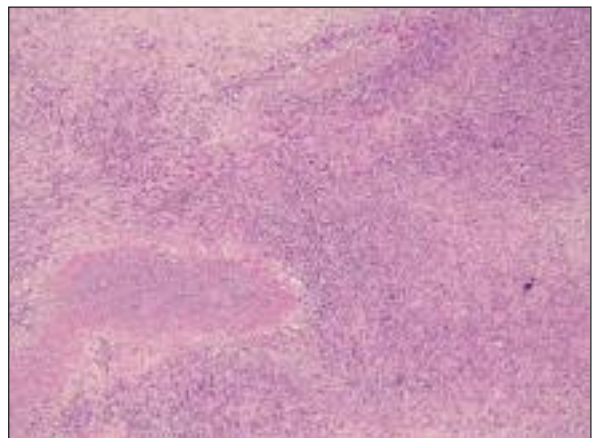


FIGURE 6: Panoramic imaging of mesenchymal tumour, highly cellular, including geographic necrotic areas (Haematoxylin-eosin, x20).

occur usually to lung or other bones, brain metastases occur rarely. Brain metastases are usually detected after surgical procedure or chemotherapy. In our case, metastases to brain and lung occurred before surgery and indicated a short survival time.

As MFH is primarily a bone tumor and we could not detect any tumor in the long bones we considered this case as a primary MFH of spinal. Total tumor removal is difficult because of invasion, local recurrences are frequent and survival time is short.

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