

Case Report

Primary Paravertebral Low-Grade Fibromyxoid Sarcoma

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The authors report a 58-year-old woman with low-grade fibromyxoid sarcoma primarily located in the right paravertebral area with extension to L4 neural foramen. The patient complained lower back pain with radiating pain along the posterolateral aspect of the right lower leg. She underwent subtotal surgical removal and Cyber Knife therapy. Diagnosis was made by strikingly characteristic microscopic appearance of a bland spindle cell sarcoma which contained numerous giant collagen rosettes and was also supported by immunohistological findings. The diagnostic image findings and literatures are reviewed and discussed.

KEY WORDS : Low grade fibromyxoid sarcoma · MRI · Cyber Knife therapy.

INTRODUCTION

Low-grade fibromyxoid sarcoma (LGFMS), first described by Evans²⁾ in 1987, is a rare sarcoma exhibiting bland histological feature but paradoxically aggressive behavior by showing a high rate of local recurrence (33%)⁴⁾ and metastasis (58%)⁴⁾.

The incidence of the tumor is unclear but only three patients were seen in 56 years in a major general hospital with special interest in soft-tissue tumors³⁾. Over the last 2 decades, roughly 150 cases have been documented with local recurrence, metastasis, and death¹²⁾. There was a predominance in male and young adult group between the age of 25 and 46 reported by Goodlad et al.³⁾.

The tumor most commonly arises in the deep soft tissue of the lower extremities, but rare cases were seen in paravertebral region^{3,6,7)}. There are few reports of the magnetic resonance imaging (MRI) feature of LGFMS.

We present a case of primary paravertebral LGFMS, emphasizing its MRI, clinical, and histological features.

CASE REPORT

A 58-year-old female visited our hospital with 2-3 months

history of progressive back pain and radiating pain along the posterior aspect of right lower extremity. Lumbar spinal magnetic resonance image (MRI) taken at the local clinic showed a right-sided large mass encroaching nerve root at L4-5 intervertebral foramen. She was referred to our institution for further work up and management. There was a palpable paraspinal mass at low back, and SLRT was 50°/80°. There was also hypesthesia along right L5 dermatome, but there was no motor weakness. Erosion of bone at the transverse process of L5 was suspected by plain radiograph of lumbar spine (Fig. 1) The MRI (Fig. 2, 3) and CT (Fig. 4) revealed a

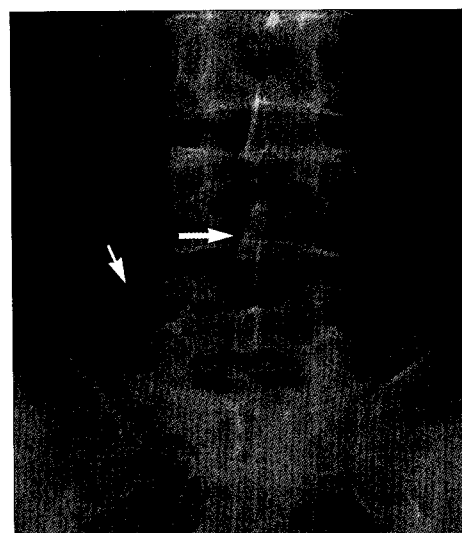


Fig. 1. Anterior-posterior view of plain X-ray of the lumbar spine. Bone erosion is suspected at right transverse process (arrow) of L5 and right side of spinous process of L4 (arrow head).

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Fig. 2. Preoperative lumbar sagittal MR image T1WI (T2WI, T1WI after contrast injection) shows large multilobulated soft mass with low to slightly high signal intensity (SI) on T1WI and low to slightly high SI on T2WI, and heterogenous enhancement on T1WI after contrast injection.

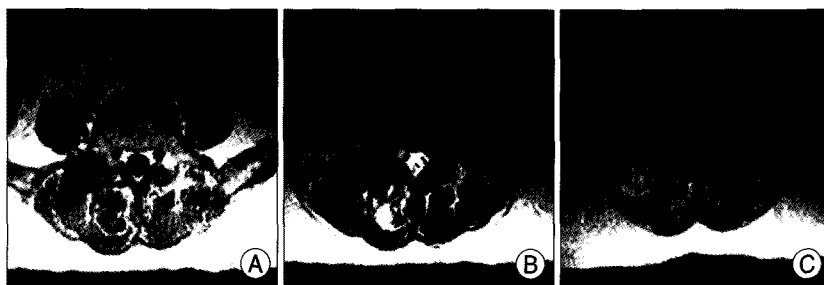


Fig. 3. Preoperative axial lumbar MR image (A : T1WI, B : T2WI, C : T1WI after contrast injection) shows a large multilobulated soft tissue mass in right lower back erector muscles and extension to right L4 neural foramen with compression of L4 nerve root with low to slightly high signal intensity (SI) on T1WI and low to slightly high SI on T2WI, and heterogenous enhancement on T1WI after contrast injection.



Fig. 4. Preoperative lumbar axial CT without contrast shows a large multilobulated soft mass with bone erosion in right lower back erector spinal muscles and extension to right L4 neural foramen with compression of L4 nerve root.

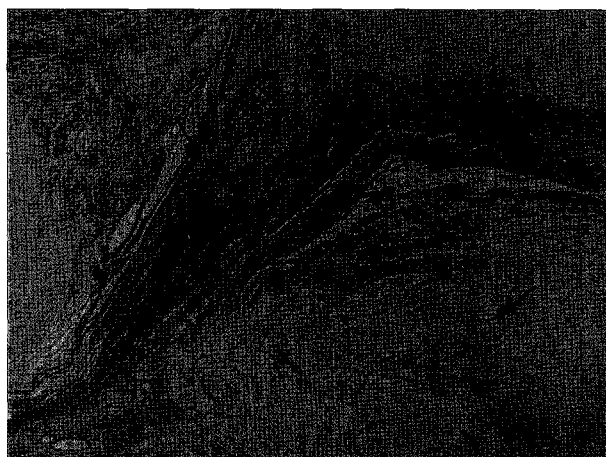


Fig. 5. Junction between fibrous (short arrow) and myxoid zones (long arrow) in low-grade fibromyxoid sarcoma. Alternating fibrous and myxoid zones in a hyalinizing spindle cell tumor with rosettes (HE stain $\times 40$).

large multilobulating soft tissue mass, sized $102 \times 51 \times 74$ mm, with bony erosion at the level of right lower back erector spinal muscles and extension of mass to right L4 intervertebral foramen resulting encroachment of L4 nerve root and spinal stenosis at L4-5 intervertebral foramen. The mass showed low to slightly high signal intensity on T1 weighted image and heterogeneously low to high signal intensity on T2 weighted image, and heterogenous enhancement on T1 weighted image after intravenous injection of gadolinium. There was no relevant family history and no past history of significant disease. Blood chemistry and blood count values were within normal limits and the chest X-ray revealed no abnormality. The patient underwent the operation. Well-circumscribed mass, which was attached firmly to the nerve root sheath and dura mater, was subtotally removed. Grossly, the tumor was an irregular tan-gray mass. The cut surface of the tumor was grayish-white, trabecular fibrotic pattern and focal cystic with mucoid material.

Microscopic examination revealed that tumor cells were embedded in a fibrous or myxoid stroma which tended to be alternate in different areas of the tumor (Fig. 5). The tumor was composed of bland spindle-shaped cells

with pale eosinophilic cytoplasm and small oval nuclei and showed an admixture of hypocellular myxoid cells and hypercellular spindle cells placed in a collagenous stroma. There were no mitotic figures, necrosis nor hemorrhage. A prominent network of capillary-sized blood vessels was seen in the myxoid zones (Fig. 6). Numerous large hyalinized collagen rosettes were surrounded by a cuff of tumor cells with round nuclei (Fig. 7). Immunohistochemical analysis of the tumor cells revealed a diffuse expression of Ki-67 (approximately 8%), but the cells did not express EMA CD34, SMA, or S-100 protein. The histopathological and immunohistochemical findings were similar with that of low-grade fibromyxoid sarcoma.

Postoperative MRI demonstrated a small tumor remnant in the L4-5 intervertebral foremen area. Cyber knife radiosurgery was started right after the operation (total of 2,100

cGy for 3 days). Six months later, the patient complained right side radiating pain with back pain. The repeated MRI examination revealed that the remnant mass in size was slightly increased (about 1.3 times) compared to the previous MRI (Fig. 8). Patient did not want to go through the second operation for total removal of the tumor, so we planned the second cyber knife radiosurgery.

DISCUSSION

Low-grade fibromyxoid sarcoma was first described by Evans²⁾ as a slow-growing, asymptomatic soft tissue tumor with apparently benign histological characteristics belying a high metastatic potential. In recent years several new entities of low-grade fibrosarcomas have been described^{1,4,11)}; 1) low-grade myxofibrosarcoma (LGMFS), 2) low-grade fibromyxoid sarcoma (LGFMS), 3) hyalinizing spindle cell tumor with giant collagen rosettes (HST), and 4) sclerosing epithelial fibrosarcoma (SEF). Myxofibrosarcoma may show a progression to high grade sarcoma, but the remaining three types almost always show low-grade fibrosarcoma.

The usual presentation of the tumor is a slowly growing, painless, deep soft tissue mass that ranges from 1 to 18 cm in greatest diameter, although most are about 8-10 cm. The tumor most commonly arises in the deep soft tissue of the lower extremities, particularly the thigh and following regions can be affected in decreasing order of frequency: the chest wall/axilla, shoulder region, inguinal region, buttock, and neck^{10,11)}. Rare cases have also been described in unusual sites including the retroperitoneum, small bowel mesentery, mediastinum, and paravertebral region^{3,6,7)}.

The imaging characteristics of LGFMS described a heterogeneous MR imaging appearance with low to slightly high SI on T1, heterogeneously low to high SI on T2 and heterogeneous postcontrast enhancement⁸⁾.

In our patient, the tumor was a firm nodular mass in paravertebral region, measuring 102×51×74 mm encroached to the right L4 nerve root on MRI with heterogeneous enhancement. Grossly, the tumor was an irregular tan-gray mass. On the cut surface, focal areas of glistening grayish-white substance were admixed with poorly demarcated firm area. Neither necrosis nor hemorrhage was present.

The diagnosis of "fibrosarcoma" for these tumors resulted from the following observation. Characteristically, the tumor cells showed fibroblastic characteristic: they were spindle-shaped with fusiform nuclei and with faintly eosinophilic cytoplasm lacking perinuclear vesicles. By means of immunohistochemistry, the cells did not show any lineage specificity [negativity for α -smooth muscle actin (SMA), desmin, S100, and epithelial membrane antigen (EMA)]. CD34 was posi-

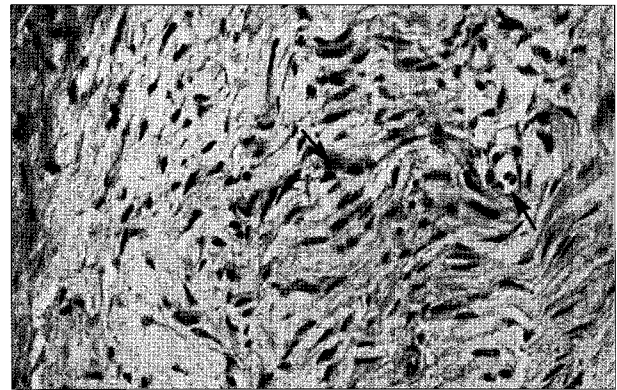


Fig. 6. Medium power view of low-grade fibromyxoid sarcoma (LGFMS), showing bland fibroblasts arranged in a swirling pattern, with myxoid stroma and a prominent net work of branching capillary-sized blood vessels (arrow), but no nuclear pleomorphism or mitotic figures reminiscent of LGFMS (HE stain x 400).

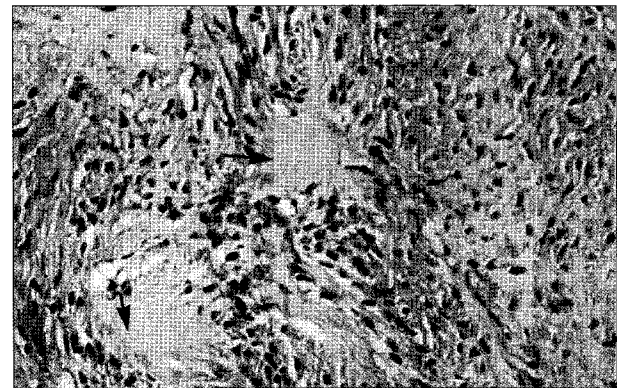


Fig. 7. Giant rosettes (arrow) with hyalinized central collagen surrounded by plump to oval cells (HE stain x 400).

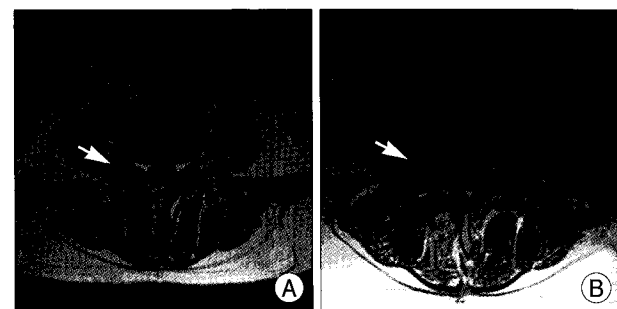


Fig. 8. Postoperative MRI with contrast. A: Pre-Cyber knife therapy MRI shows a small non-enhanced mass at L4-5 intervertebral foramen (white arrow). B: On MRI taken 6 months after Cyber knife therapy, a mass (white arrow) shows slight enlargement compared with the mass shown on previous MRI (A).

tive in some cases^{3,10)}. In our case, tumor cells did not express any lineage specificity (CD34, S-100, SMA, EMA), but Ki-67 was expressed in approximately 8% at most active area, which indicate that the tumor had a benign characteristics.

The differential diagnosis of low-grade fibromyxoid sarcoma includes numerous benign and malignant soft tissue lesions characterized by a fibrous and myxoid stroma. Myxoid neurofibroma is composed of cells with more slender

and wavy nuclei that consistently express S-100 protein. Perineurinomas have been suggested as a possible source of confusion as they can present a deep seated spindle cell tumor with fibrous and myxoid areas. However, they usually show neither the typically alternating fibrous and myxoid pattern, nor the arcades of small vessels. Meanwhile, they have a long, slender cytoplasmic processes which are not seen in LGFMS. And immunohistochemical detection of EMA allows its distinction from LGFMS. Nodular fasciitis is characterized by a cleft-like space, extravasation of erythrocytes, and presence of multinucleated giant cells which are not found in LGFMS. Malignant peripheral nerve sheath tumors may contain myxoid foci, but the cells are more elongated or wavy, and S-100 protein is stained. Myxoid liposarcoma always contains an atypical lipomatous component that includes the presence of lipoblasts with a well-developed plexiform vascular pattern^{5,12}.

Some authors have reported the use of adjuvant radiotherapy following local excision^{6,9}. We considered radiotherapy (cyber knife) after the operation due to the report indicating that it could benefit the patient, but chemotherapy was not considered. However, the patient was admitted to our department for radiating pain on the posterolateral aspect of the right leg a year after the operation. At that time, tumor grew about 1.3 times bigger compared to the postoperative remnant tumor, but there were no evidence of metastasis to other sites. Rando et al.⁶ and Tun et al.⁹ suggested that adjuvant radiotherapy might lengthen the period between recurrence of tumor. Although radiotherapy does not appear to change the course of the disease with regard to subsequent recurrences and metastasis, we suggest that radiotherapy might be an option in the management of LGFMS for life expectancy. Even though LGFMS showed resistance to radiotherapy in our case, we planned to start the second cyber knife.

CONCLUSION

In conclusion, accurate histological assessment of primary

paravertebral LGFMS requires clinical and radiographical correlation. In diagnosis of primary paravertebral LGFMS, the differential diagnosis of other primary sarcomas and neurofibromas should be kept in mind.

Although surgery is the mainstay of therapy for this tumor, adjuvant radiotherapy may compensate for the difficulty in removing a safety margin of normal tissue.

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