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Intramuscular Low-Grade Fibromyxoid Sarcoma: An Efficacy of Cytoplasmic Mucin 4 Immunoreexpression

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Dear Editor:

A 37-year-old Japanese male presented with a movable, subcutaneous nodule measuring 75 mm×70 mm on the right side of his back (Fig. 1A). T2-weighted magnetic resonance imaging (MRI) revealed a hyperintense mass in the right latissimus dorsi muscle (Fig. 1B). A hematoxylin and eosin (H&E)-stained specimen demonstrated well-demarcated, admixed heavily collagenous and myxoid zones in the muscle bundles (Fig. 2A). In portions of the specimen, arcades of small blood vessels were easily seen (Fig. 2B).

Collagen rosettes surrounded by epithelioid fibroblasts were identified (Fig. 2C). Proliferating spindle cells were deceptively bland (Fig. 2D). Neoplastic cells were negative for CD34, CD68, α -smooth muscle actin, S-100 protein expression, and anaplastic lymphoma kinase. Mucin 4 (MUC4) was diffusely and strongly expressed in the cytoplasm of neoplastic cells (Fig. 2E). The Ki-67 proliferation index was 2% to 3% (Fig. 2F). A diagnosis of intramuscular low-grade fibromyxoid sarcoma (LGFMS) was made. We performed wide local excision with a 30-mm

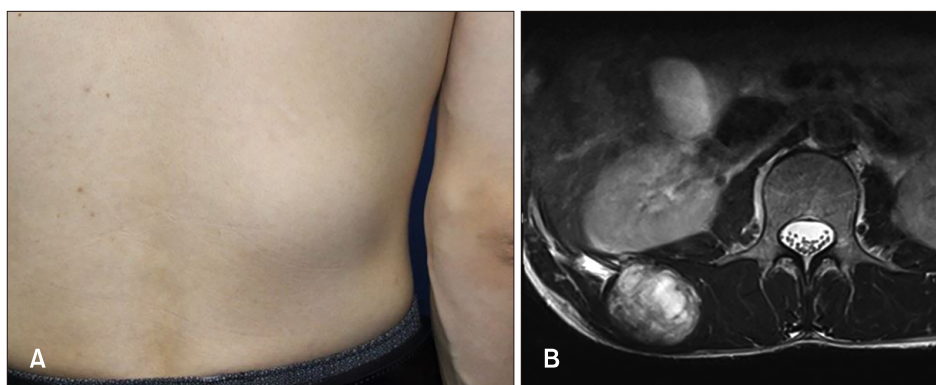


Fig. 1. (A) A 75-mm×70-mm, movable, subcutaneous nodule on the right side of the lower back (clinical view of a 37-year-old Japanese male). (B) A high-density area in the right latissimus dorsi muscle (T2-weighted magnetic resonance imaging).

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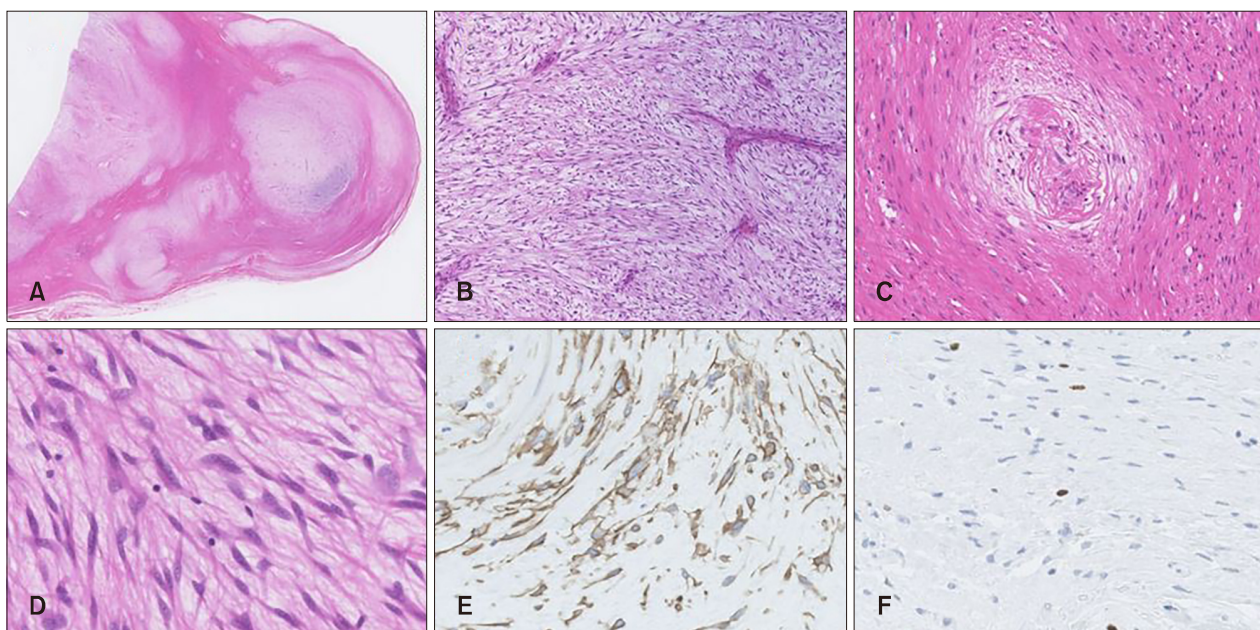


Fig. 2. (A) Admixed heavily collagenous and myxoid zones in the muscle bundles (H&E, $\times 4$). (B) Arcades of small blood vessels (H&E, $\times 100$). (C) Collagen rosettes (H&E, $\times 200$). (D) Deceptively bland spindle cells (H&E, $\times 400$). (E) Cytoplasmic expression of mucin 4 (immunohistochemistry, $\times 400$). (F) The Ki-67 proliferation index was 2%~3% (immunohistochemistry, $\times 400$).

margin. The histological margin was clear, and neither local recurrence nor distant metastases have appeared during 16 months of follow-up.

LGFMS was first described by Evans in 1987¹, and is also called Evans tumor. LGFMS is a fibroblastic/myofibroblastic sarcoma with the potential for late recurrence and metastasis¹. LGFMS is composed of deceptively bland spindle cells in admixed heavily collagenous and myxoid zones, accompanied by a whorling growth pattern and arcades of blood vessels¹. Collagen rosettes are frequently identified. Many benign and malignant fibroblastic/myofibroblastic histological mimics reveal spindle cell proliferation and fibromyxoid changes in soft tissue: nodular fasciitis, dermatofibrosarcoma protuberans, solitary fibrous tumor, inflammatory myofibroblastic tumor, low-grade myofibroblastic sarcoma, myxoinflammatory fibroblastic sarcoma, fibrosarcoma, myxofibrosarcoma, and LGFMS. Among the above, dermatopathologists rarely consider LGFMS in the differential diagnosis, because it is uncommon. LGFMS usually arises in the deep soft tissue of the lower extremity, particularly the thigh, but may occur in other location. Furthermore, intramuscular LGFMS has rarely been reported²⁻⁴. Misdiagnosis of intramuscular LGFMS could be as a benign soft tissue tumor because of its bland-looking histologic features. Intramuscular myxoma should be kept in mind as a histological mimic. Detection of *FUS* gene rearrangement by fluorescence *in situ* hybridization (FISH) and *FUS-CREB3L2/FUS-CREB3L1* chimeric fusion genes

by RT-PCR has been reported to be reliable for diagnosis in LGFMS⁴. Recently, Doyle et al.⁵ found that cytoplasmic expression of MUC4 was highly sensitive and specific for LGFMS. Other fibroblastic/myofibroblastic mimics have barely reported to be positive for MUC4. Needless to say, the immunostaining method is a more useful tool than FISH for clinicians. However, immunoexpression of MUC4 has rarely been reported in intramuscular LGFMS to the best of our knowledge². Similar to conventional LGFMS, MUC4 may be a useful marker for intramuscular LGFMS.

In summary, we have described a rare case of intramuscular LGFMS of the right latissimus dorsi muscle demonstrating cytoplasmic MUC4 immunoexpression.

We received the patient's consent form about publishing all photographic materials.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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