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Case Report

A rare case of low-grade fibromyxoid sarcoma with ossification which was radiologically detected as apparent calcification and histopathologically proven

Yasuaki Tsuchida[a], Yoshitane Tsukamoto[a,b,*], Hiroyuki Futani[c], Shunsuke Kumanishi[c], Takahiro Watanabe[a], Noriko Kajimoto[d], Shohei Matsuo[b], Shinichi Yoshiya[c], Seiichi Hirota[a]

[a] Department of Surgical Pathology, 1-1 Mukogawa-cho, Nishinomiya, Hyogo 553-8501, Japan
[b] Department of Pathology and Laboratory Medicine, Takarazuka City Hospital, 4-5-1 Kohama, Takarazuka, Hyogo 665-0827, Japan
[c] Department of Orthopaedic Surgery, Hyogo College of Medicine, 1-1 Mukogawa-cho, Nishinomiya, Hyogo 553-8501, Japan
[d] Department of Pathology, Meiwa Hospital, 4-31 Age-Naruo-cho, Nishinomiya, Hyogo 663-8186, Japan

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ABSTRACT

Low-grade fibromyxoid sarcoma (LGFMS) is a rare soft-tissue malignant neoplasm with a deceptively benign histological appearance and a potential for late recurrence and metastasis. LGFMS cases with significant ossification are extremely rare. To our knowledge, only three cases of LGFMS with bone formation which is detected as apparent calcification by radiological examinations and proven by histopathology have been reported. Here, we report the fourth such case. A 39-year-old female presented with a 10-year history of a painless tumor in her right buttock. Computed tomography images showed multiple foci of intratumoral calcification. A needle biopsy specimen of the tumor revealed a spindle cell neoplasm with hyalinizing/collagenous stroma and ossification. The tumor cells were immunohistochemically positive for MUC4 and the molecular analysis of the tumor detected *FUS-CREB3L2* fusion. Pathological diagnosis of LGFMS was made. Total resection of the tumor with wide margins was performed, and histology of the resected sample showed multiple foci of intratumoral ossification.

1. Introduction

Low-grade fibromyxoid sarcoma (LGFMS) was first described by Evans in 1987 [1]. LGFMS is characterized by benign-looking histology but malignant behavior after the long-term follow-up. Fourteen out of 33 cases of LGFMSs (42%) died of tumor during 3- to 42-year follow-up period. Twenty-one of 33 (64%) had recurrence during up to 15-year follow-up period, and 15 of 33 (45%) had metastases during up to 45-year follow-up period. Dedifferentiation is reported to be the only histological hallmark of aggressive clinical behavior [2].

Recurrent fusion-genes are identified, and *FUS-CREB3L2* is in approximately 95% of the cases and *FUS-CREB3L1* in < 5% of them [3–8]. Currently, MUC4 is considered to be a highly sensitive and specific marker for LGFMS [9,10]. LGFMSs show morphological and molecular overlap with sclerosing epithelioid fibrosarcomas (SEFs) [7], but FUS rearrangements have been reported to be rare in pure SEFs [11].

Radiological examinations such as roentgenography or computed tomography (CT) can detect calcification, but it is difficult to distinguish whether it is simple calcification or true ossification. Only pathological examination can prove the nature of the calcification. Small foci of osseous metaplasia or mature bone formation in LGFMSs are reported in some cases [2,7]. However, LGFMSs with ossification which is detected as apparent calcification by radiological examinations and proven by histopathology are extremely rare. To our knowledge, only three such cases of LGFMS have been reported.

Here, we report the fourth LGFMS case with ossification which was radiologically detected as apparent calcification and histopathologically proven. MUC4 expression by immunohistochemistry and presence of *FUS-CREB3L2* fusion by RT-PCR confirmed that this is really a case of LGFMS.

1.1. Case report

A 39-year-old female patient with Crohn's disease had been followed at a local gastroenterology clinic after resection of the small intestine and colon. Abdominal CT taken for investigation of abnormal genital bleeding unexpectedly revealed a tumor within the gluteal muscles. She reported a 10-year history of a painless tumor in her right buttock. The patient was referred to the Department of Orthopaedic

* Corresponding author at: Department of Pathology and Laboratory Medicine, Takarazuka City Hospital, 4-5-1 Kohama, Takarazuka, Hyogo 665-0827, Japan.
E-mail address: tsuka-y@hyo-med.ac.jp (Y. Tsukamoto).

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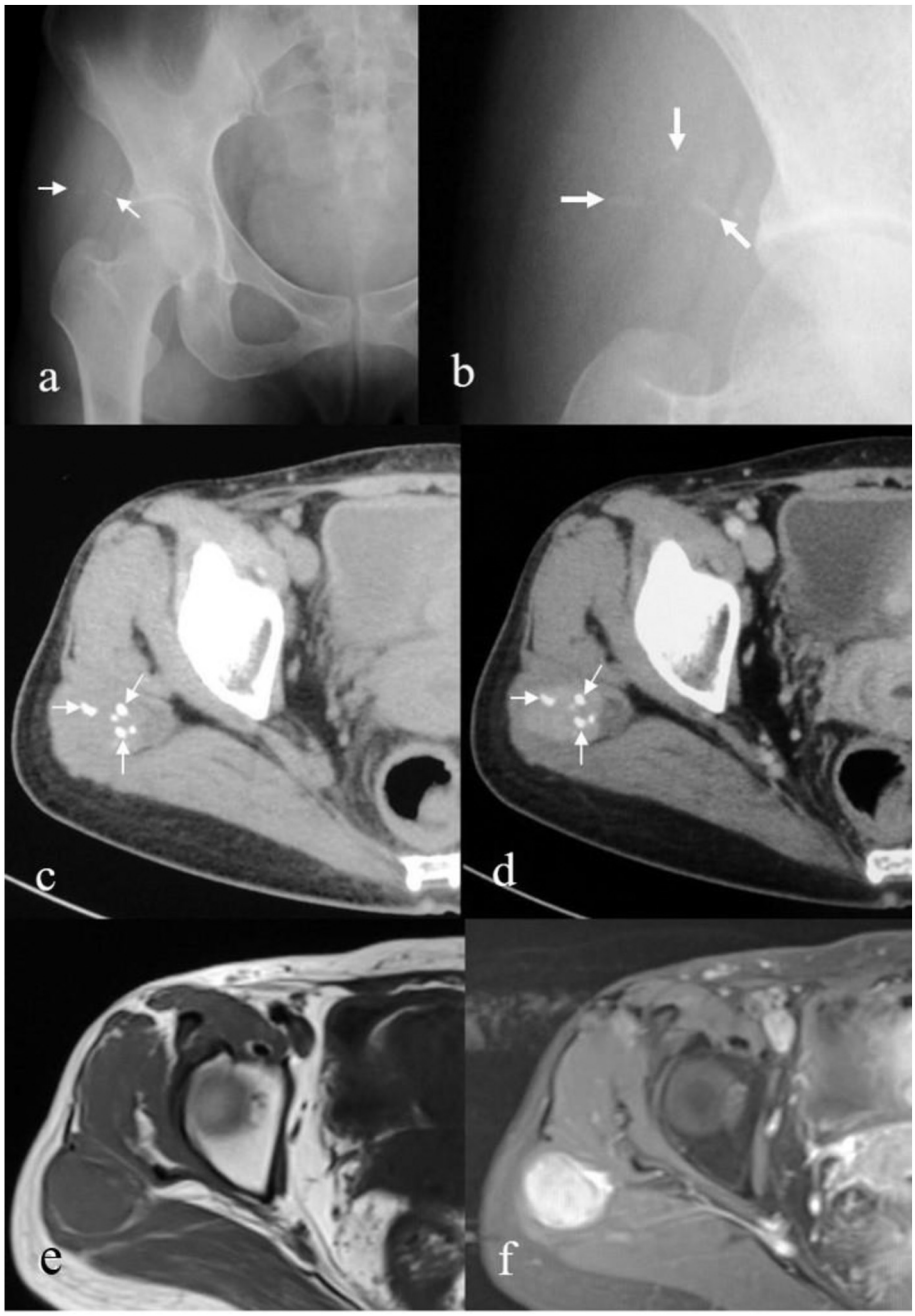

Fig. 1. Radiological findings show calcification. Roentgenogram (Fig. 1a & b) and abdominal CT image (Fig. 1c & d) show the mass with multiple foci of calcification. Fig. 1b is the image of high magnification of Fig. 1a. Fig. 1c shows plain CT image, and Fig. 1d contrast-enhanced one. White arrows in Fig. 1a–d indicate calcification. MRI (Fig. 1e & f) shows the gadolinium enhanced mass in the right buttock (Fig. 1e & f show the image without or with gadolinium enhancement, respectively).

Surgery, Hyogo College of Medicine Hospital for further investigation. Physical examination showed that the tumor was elastic hard. Roentgenogram revealed calcification in the posterior aspect of the right buttock (Fig. 1a & b), and CT images clearly demonstrated the tumor with multiple foci of apparent intratumoral calcification (Fig. 1c & d). Magnetic resonance imaging (MRI) revealed a 60 × 45 × 30 mm well-defined tumor. T1-weighted images showed an area of low signal intensity (Fig. 1e), and T2-weighted images a high signal intensity area including a low signal intensity area within the tumor (data not shown). The gadolinium-enhanced T1 image demonstrated good enhancement but contained non-enhanced foci (Fig. 1f). These findings suggested that the soft tissue tumor had focal but apparent calcification. For the definite diagnosis, a needle biopsy was performed. After the pathological diagnosis of the biopsied material, total resection of the tumor with wide margins was done. The patient is alive without disease recurrence 27 months after the first visit to our hospital.

2. Materials and methods

2.1. Immunohistochemistry (IHC)

Biopsied and resected tissues were fixed in 10% buffered formalin and embedded in paraffin. Three-micrometer-thick sections were cut and stained with hematoxylin and eosin (H&E). IHC for alpha-SMA (1A4, DAKO, Glostrup, Denmark, 1:500), Ki-67 (MIB-1, DAKO, 1:300), Desmin (DE-R-II, Leica, Wetzlar, Germany, 1:50), CD34 (QBend/10, Leica, 1:2000), S100p (polyclonal, Leica, 1:5000), STAT6 (s-20, Santa Cruz Biotechnology, Santa Cruz, CA, 1:1000), MUC4 (8G7, Santa Cruz, 1:1000), beta-catenin (14/Beta-catenin, Becton Dickinson, Franklin Lakes, NJ, 1:400) was performed using Bond Polymer Refine Detection (Leica). IHCs without primary antibodies were performed as negative controls.