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Case Report

Intramuscular dendritic fibromyxolipoma of the right thigh region: a case report and review of the literature

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Abstract: We report a case of rare benign soft tissue tumor of the right thigh region which was diagnosed as a dendritic fibromyxolipoma (DFML). The mass was 15.0×9.5×6.0 cm in size, with well-circumscribed margins and rubbery texture. The resected mass was characterized microscopically by small spindle or stellate cells, admixed with mature adipose tissue and prominent, abundant myxoid stroma with ropey collagen bundles. An immunohistochemical study showed the adipocytes expressed S-100 protein, and the spindle and stellate cells were strongly positive for vimentin and CD34. The cells were not positive for Bcl-2, SMA, CD99, Ckpan, or Desmin. A clinical follow-up one year after surgery revealed no signs of recurrence or metastasis of the mass. The present study reports the first case of an intramuscular DFML occurring between the vastus lateralis muscle and quadriceps in the thigh region. The clinicopathological features of the mass and differential diagnosis are briefly discussed, followed by a review of literature.

Keywords: Dendritic fibromyxolipoma, soft tissue, spindle cell lipoma, thigh region, intramuscular

Introduction

Dendritic fibromyxolipoma (DFML) is an uncommon, benign soft tissue tumor that shares many clinicopathological features with the rare, myxoid variant of spindle cell lipoma and solitary fibrous tumor with myxoid stroma [1]. Histologically, a mixture of spindle and stellate cells, mature adipose tissue, and abundant myxoid stroma with prominent collagenization characterizes a DFML. A DFML is usually located in the subcutis of the head and neck, chest wall, shoulders, forearm, back or groin [1-10]. These neoplasms typically show positive immunoreactivity for vimentin, CD-34 and Bcl-2. The tumor's adipocytes may also show positive staining for the S-100 protein. The present study reported a rare case of a DFML of the right thigh region in a 58-year-old patient. To the best of our knowledge, no other relevant case of intramuscular DFML of the thigh has been previously reported.

Clinical summary

A 58-year-old male presented to the Taizhou People's Hospital, Taizhou, Jiangsu province, China with a history of a mass in his right thigh for two months duration. Reportedly, the mass had grown slowly and was painless. The patient had no previous medical history of disease. Physical examination revealed a 16×7-cm subcutaneous, soft, non-tender mass over the right thigh region. Laboratory data and tumor markers, such as CEA and CA19-9, were normal. Magnetic resonance imaging (MRI) revealed that the mass was located in outer front soft tissue of the right thigh region. MRI demonstrated increased or normal signal intensity on the T1W1 imaging sequence and hybrid signal intensity on T2W1 imaging sequence (**Figure 1**). Surgical exploration located the tumor between the vastus lateralis muscle and quadriceps in the thigh region. The patient underwent complete, local excision of the mass.

The excised specimen measured 15×9.5×6.0 cm and was a well-encapsulated soft-tissue mass, with a small amount of striated muscle within its surface. The cut surface was yellow-gray and white in color with a myxoid and soft texture. Microscopically, the tumor was mainly composed of a proliferation of small spindle or stellate cells, prominent abundant myxoid stro-

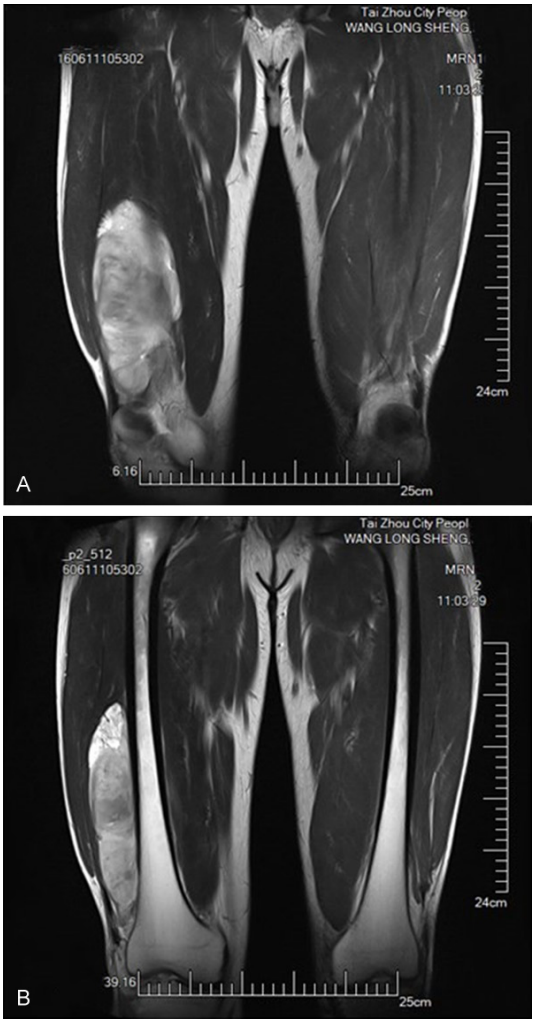

Figure 1. Magnetic resonance imaging of the lesion. The mass was located in the outer front of the right thigh region, and it demonstrated increased or normal signal intensity on T1W1 imaging sequence and hybrid signal intensity on the T2W1 imaging sequence.

ma with ropey collagen bundles, and an admixture of mature adipose tissues (**Figure 2A**). The spindle and stellate cells strongly expressed CD34 and vimentin (**Figure 2B**, **2C**). The cells were negative for Bcl-2, smooth muscle actin, and desmin. The adipocytes and a limited number of spindle and stellate cells expressed S-100 protein (**Figure 2D**). Ki-67 showed low proliferative activity. Recurrence of the mass or metastases was not reported at the one-year post-operative clinical follow-up.

Discussion

Dendritic fibromyxolipoma (DFML) is an extremely rare, benign soft tissue lesion that was originally described in 1998 by Suster et al. [1]. To date, only 21 cases of DFML tumors from 10 reports have been described in the English literature and cited in PUBMED (**Table 1**). A review of these cases shows that the age of the patients ranged between 24 and 81-years-old (median age, 59.4-years-old) at the time of diagnosis. Of the reported cases, there was a higher prevalence of males, with only five cases being females. The tumors were most frequently located in the subcutaneous tissue or muscular fascia of the head and neck, shoulders, chest wall, or back. Infrequently, masses were reported to be located in the intramuscular tissue, forearm, lower lip region, inguinal region, or perineum regions. The reported tumor size varied from 1 to 24 cm (average size, 7.4 cm), and all tumors were benign. Local mass excision is considered to be the most effective treatment for a DFML. To date, a DFML lesion located in the thigh region has not been reported.

The most predominantly histologic feature of a DFML is the admixture of abundant myxoid stroma with ropey collagen bundles, spindle or stellate cells, and mature adipose tissues. Additionally, as described by Karim et al. [2], chondroid metaplasia is an uncommon feature of a DFML. Immunohistochemically, vimentin and CD34 immunohistochemical stains accentuated the cell's dendritic nature by revealing slender, complex cytoplasmic prolongations, and these finding contributed to the DFML name [1].

Histology should be differentiated from a DFML from certain benign and malignant lesions, such as the: spindle cell lipoma (SCL), solitary fibrous tumor (SFT), myxoid liposarcoma (MLS), lipoblastoma, lipoblastomatosis and nodular fascitis [6]. DFML shares many characteristics with SCLs, including the more frequent occurrence in males and elderly, gross features and tumor location. Both types of tumors occur predominantly in the head and neck and shoulder region. They are considered to be benign lesions, and local recurrence is exceptionally rare even with incomplete surgical excision [7]. Suster et al. [1] emphasized that a DFML can be readily distinguished from an SCL by three distinctive histological features: (i) Dendritic nature of the spindle cells; (ii) The abundance of keloidal-type collagen; and (iii) The occasional plexiform proliferation of capillary-sized vessels.

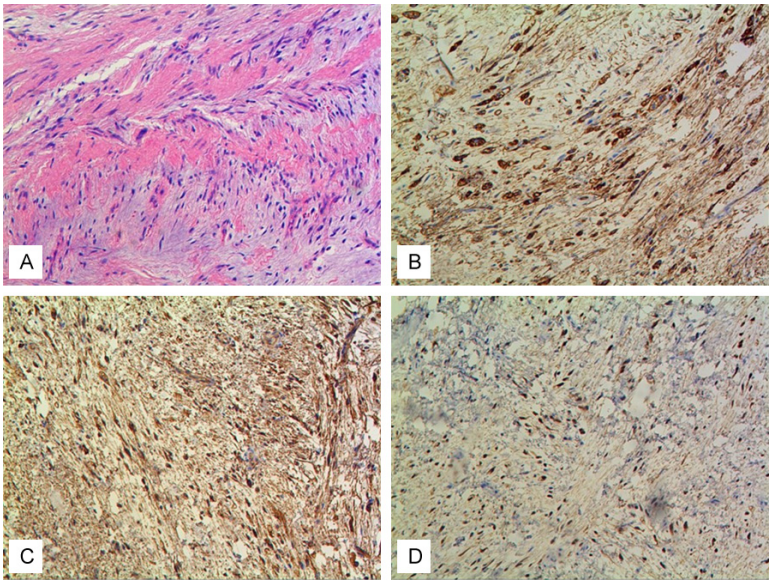

Figure 2. Microscopic imaging of the tumor. The mass was mainly composed of a small spindle or stellate cells, prominent abundant myxoid stroma with ropey collagen bundles, and an admixture of mature adipose tissues (A). On immunohistochemistry studies, the spindle and stellate cells strongly expressed CD34 (B) and vimentin (C). The adipocytes expressed S-100 protein (D).

Other benign spindle cell tumors that should be considered in the differential diagnosis of DFML include SFTs. Having a predilection for the thoracic cavity, SFTs are rare, fibrous neoplasms. SFTs are classically composed of short spindle cells that are singly separated by strands of rope-like collagen. Additionally, SFTs have a "hemangiopericytoma-like" vascual pattern where the lesional cells are densest around the small and mediately ectatic and branching vessels [5]. The "hemangiopericytoma-like" vascular pattern and the lack of an adipose tissue component are two histological features that distinguish an SFT from a DFML [2].

Due to the tumor's relatively large size, plentiful myxoid matrix, and proliferation of capillaries, it is more subtle and difficult to distinguish a DFML from an MLS. Sharing many features with DFML, the MLS is the most common subtype of liposarcoma with a low-grade malignancy rate. Macroscopically, an MLS is indiscernible from a DFML with both tumor types being usually well-circumscribed masses with tan, gelatinous cut surfaces. Primitive, non-lipogenetic mesenchymal cells and the absence of signet-ring lipoblasts are classically present in MLS tumors, and this observation may help differentiate a DFML from an MLS. Furthermore, molecular studies have previously sh-own that MLS tumors are characterized by recurrent translocations t (12; 16) (q13; p11), and rarely, t (12; 22) (q13; q12), which fuse FUS/EWSR1, respectively, to DDIT3 on chromosome 12 gene [11]. Zhang et al. [12] revealed that molecular testing was useful for all deep-seated and large (>15 cm), lipoma-like tumors. Narendra et al. [13] confirmed that FISH with-DDIT3 break-apart probe was a valuable adjunct for excluding masses that morphologically mimics MLS tumors. Additionally, information obtained from clinical and radiological diagnostics showed that a frequent feature of malignancy is a deep-seated soft tissue mass with infiltrative borders. Although routine immunohistochemical studies are not usually performed for MLS tumors, the neoplastic MLS cells are diffusely positive for S-100 protein and are CD34 negative, which differs from DFML cases [7].

In summary, to our knowledge, this is the first reported case of a rare, intramuscular DFML of the right thigh region. Although the location and size of this tumor was unusual, the diagnosis was based on the typical morphological and immunohistochemical features of DFMLs. Although rare, it is important to properly identify a DFML, as it can be confused with other more aggressive neoplasms such as a myxoid liposarcoma, myxofibrosarcoma or low-grade fibromyxoid sarcoma. Complete local excision is the treatment option for dendritic fibromixolipoma.

Disclosure of conflict of interest

None.

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Table 1. Clinical summary of reported cases of dendritic fibromyxolipoma in the English literature

References	Gender/Age	Tumor size (cm)	Location	Immunohistochemical	Follow up
Suster et al. (1998) [1]	M/33	11	Left posterior shoulder, acromium region	CD34+, Bcl-2+	NA
	M/54	5.0×5.0×4.0	Right posterior neck	CD34+, Bcl-2+	NA
	M/58	7.5×5.5×3.0	Right shoulder	CD34+, Bcl-2+	NED, 7 years
	M/63	6.0×5.5×2.0	Upper back	CD34+, Bcl-2+	NA
	M/66	8.0×3.5×2.5	Back of the neck	CD34+, Bcl-2+	NA
	M/66	9.0×7.0×6.5	Back, posterior axilla	CD34+, Bcl-2+	NA
	M/70	2.0×2.0×2.0	Face, right nasal area	CD34+, Bcl-2+	NED, 11 years
	M/73	7.0×5.5×2.5	Right posterior neck	CD34+, Bcl-2+	NED, 13 years
	M/77	3.0×2.0×1.5	Back of neck	CD34+, Bcl-2+	NED, 5 years
	M/79	3.5×3.0×2.5	Right chest wall	CD34+, Bcl-2+	Died of unrelated disease
	M/81	3.5×3.0×3.0	Left chest wall	CD34+, Bcl-2+	NED, 5 years
	F/50	6.0×5.5×5.0	Right upper back	CD34+, Bcl-2+	Lost to follow-up
Karim et al. (2003) [2]	M/73	13×8.0×5.5	Between infraspinatus and deltoid muscles	CD34+, Bcl-2+	NED, 8 months
Al-Maskery et al. (2011) [3]	F/36	2.0×2.0×2.0	Lower lip	CD34+, CD-99+, Bcl-2+	NED, 2 years
Dahlin et al. (2012) [4]	F/65	2.0×3.2×1.0	Adherent to median nerve of left forearm	Vimentin+, CD34+, Bcl-2+	NA
Zhang et al. (2013) [5]	F/32	24×10.5×5.0	Right inguinal and perineum regions	Vimentin+, CD34+, Bcl-2+	NED, 9 months
Han et al. (2014) [6]	M/69	1.0×1.0	Nasal tip	Vimentin+, CD34+, Bcl-2+	NA
Wong et al. (2014) [7]	M/67	7	Left shoulder region	CD-34+, Bcl-2+	NED, 4 months
Xu et al. (2015) [9]	M/24	14×8.5×8.0	Left shoulder region	Vimentin+, CD34+, Bcl-2+	NED, 4 years
Liu et al. (2015) [8]	M/53	2.0×1.5×1.5	Latissimus dorsi of the right back	Vimentin+, CD34+, Bcl-2+	NED, 1 year
Ciloglu et al. (2016) [10]	F/59	1713×10	Left inguinal region	Vimentin+, CD34+, Bcl-2+	NED, 3 years
AlAbdulsalam et al. (2016) [14]	M/38	7.5×6.5×0.5	Hypopharynx	Vimentin+, CD34+, Bcl-2+	NA
Present case	M/58	15×9.5×6.0	Right thigh	Vimentin+, CD34+, S-100+	NED, 1 year

M, male; F, female; +, positive; NED, no evidence of recurrence; NA, not available.

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