CASE REPORT

Haemorrhagic low-grade fibromyxoid sarcoma: MR findings in two young women

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ABSTRACT. Low-grade fibromyxoid sarcoma (LGFMS) is a rare soft-tissue sarcoma characterised by a deceptively bland histological appearance and a paradoxically aggressive behaviour. LGFMS usually presents in young-to-middle-aged adults as a painless, slow-growing mass with the potential for local recurrence and metastasis despite low-grade histology. Several case reports have described variable MR findings of LGFMS without haemorrhage or necrosis. We report here on the MR findings in two young women with haemorrhagic LGFMS in the thigh.

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Low-grade fibromyxoid sarcoma (LGFMS) is a rare soft-tissue sarcoma, typically affecting young-to-middle-aged adults (median age at presentation 34 years), with an equal incidence in men and women. LGFMS can occur at various anatomical locations, including the superficial and deep soft-tissues of the proximal extremities, trunk, small bowel mesentery and intracranially, and frequently presents as a slow-growing, non-tender, firm tumour [1–4]. Recurrence and metastasis, usually to the lung, have been reported at variable but significant rates [1, 5, 6].

Typically, LGFMS appears as a well-circumscribed encapsulated mass, without haemorrhage or necrosis, and is usually indicative of a benign lesion. Histologically, the tumour consists of bland, deceptively benign-appearing spindle cells in alternating fibrous and myxoid stroma [5]. Less is known about the radiological appearance of LGFMS compared with the histological signs. We describe here haemorrhagic LGFMS occurring in the thigh in two young women.

Case report

Case 1

A 34-year-old woman presented with a 6 month history of a palpable and painless mass on her right inner-medial thigh area with increasing swelling. She had no history of injury or inflammation in that area and was otherwise healthy. Laboratory evaluation was unremarkable. A physical examination revealed a large

firm, non-tender and non-movable mass extending from the right inguinal area to the right mid-thigh. There were no functional or nerve deficits.

Radiographs obtained outside our clinic were not available. MRI, performed using a 3T scanner (Magnetom Verio instrument, Siemens, Erlangen, Germany), showed a huge, well-defined ovoid mass in the deep soft tissue of her right proximal medial thigh, obliterating the adductor brevis muscle. On T_1 weighted images, the mass was hypointense with a large internal, relatively hyperintense area suggesting haemorrhage or proteinaceous fluid (Figure 1a). T_2 weighted images showed a large hypointense outer portion and central hyperintensity, which corresponded to the relatively hyperintense region on T_1 weighted images. The T_2 weighted images also demonstrated multiple fluid–fluid levels of haemorrhage (Figure 1b). On fat-suppressed contrast-enhanced T_1 weighted images, the peripheral hypointense area showed heterogeneous contrast enhancement with some nodular appearance, whereas the central area revealed little contrast enhancement (Figure 1c).

In surgery, the mass was located in the medial aspect of the proximal thigh through the inguinal area. A wide surgical excision was performed. The mass appeared encapsulated with a fibrotic tissue component in the adductor brevis muscle. On gross examination, the resected specimen showed a large, well-circumscribed intramuscular mass measuring 11.6 × 9.0 × 5.9 cm. The cut surface of the tumour was pale yellow-white and glistening in appearance with central cystic degeneration and haemorrhage (Figure 1d). On microscopic examination, the tumour contained alternating fibrous and myxoid components, with the myxoid area showing tumour cells in a whirling growth pattern with capillary-sized blood vessels (Figure 1e). Sections were immunohistochemically stained with bcl-2, vimentin, CD34,

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Figure 1. A 34-year-old woman with a palpable mass on the right inner-medial thigh. (a) The axial T_1 weighted image (time of repetition (TR)/time of echo (TE) 730/11) shows a large ovoid mass (arrows) obliterating the adductor brevis muscle. The mass is hypointense with a large internal, relatively hyperintense area (arrowheads). (b) The axial T_2 weighted image (TR/TE 4370/71) reveals a large outer hypointense mass (arrows) with a central hyperintense area corresponding to the relatively hyperintense area on T_1 weighted images. Multiple different fluid–fluid levels (arrowheads) are observed in the hyperintense area, suggesting intralesional haemorrhage. (c) On the axial fat-suppressed contrast-enhanced T_1 weighted image (TR/TE 804/16), the outer portion demonstrates heterogeneous enhancement with some nodular appearance (arrowheads), whereas the central portion shows little contrast enhancement. (d) The gross specimen shows a well-circumscribed intramuscular mass showing a yellow-white cut surface with central cystic degeneration (arrows). (e) Alternating myxoid (arrows) and fibrous stroma (asterisks) are noted. The tumour cells in the myxoid area show a whirling growth pattern (haematoxylin and eosin stain, $\times 40$) (left). The tumour cells reveal diffuse strong positive for vimentin ($\times 200$) (right).

actin, desmin and cytokeratin AE1/AE3 antibodies. The tumour cells were positive for vimentin (Figure 1e) and bcl-2 and focally positive for actin. The final diagnosis was LGFMS.

Case 2

A 22-year-old woman presented with a slow-growing, palpable painless mass on the medial aspect of her left thigh that had been present for 12 months with increasing swelling. She had no history of trauma in that area and was otherwise healthy. On physical examination, a large, firm, non-tender and non-movable mass could be palpated on her left mid-thigh. Laboratory evaluation was unremarkable.

Radiography showed soft-tissue swelling with no mineralisation of her mid-thigh. MRI, performed using a 1.5T scanner (Twin Speed instrument, GE Medical Systems, Milwaukee, WI), showed a large, well-defined

and ovoid mass in the adductor magnus muscle of her left medial thigh. On T_1 weighted and T_2 weighted images, the mass was hypointense with an internal area of high-signal intensity indicative of haemorrhage within the mass (Figure 2a,b). On contrast-enhanced T_1 weighted images, the outer hypointense area on T_1 weighted and T_2 weighted images showed intense contrast enhancement (Figure 2c). The differential diagnosis included any aggressive lesions with haemorrhage such as a necrotic (likely high grade) sarcoma, possibly fibrous in origin in view of the signal characteristics, and lesions containing a low signal such as fibromatosis.

In surgery, the mass was solid, firm, encapsulated and with a fibrotic tissue component but without definite invasion of neurovascular structures. Surgical resection revealed that the mass was pale brown in colour, elastic, well-circumscribed and measured $7.5 \times 5.5 \times 5.5$ cm. On gross examination, the tumour showed a yellow-white solid cut surface with a glistening appearance and cystic degeneration with haemorrhage (Figure 2d). Histological

Figure 2. A 22-year-old woman with a 12 month history of a palpable mass in the left medial thigh area. (a) The axial T_1 weighted image (time of repetition (TR)/time of echo (TE) 650/13) shows a well-defined ovoid mass (arrows) in the adductor magnus muscle of the left medial thigh. It is hypointense with an internal hyperintense portion. (b) The coronal T_2 weighted image (TR/TE 4950/82) reveals a large outer hypointense mass (arrows) with internal hyperintensity. (c) The coronal contrast-enhanced T_1 weighted image (TR/TE 650/13) demonstrates intense contrast enhancement (arrows). (d) The gross specimen shows a well-circumscribed mass showing yellow-white solid cut surface and internal cystic degeneration with haemorrhage (arrows). (e) Alternating myxoid (arrows) and fibrous stroma (asterisks) are present (haematoxylin and eosin stain, $\times 40$) (left). The tumour cells reveal diffuse strong positive for vimentin ($\times 200$) (right).

examination revealed alternating areas of fibrous and myxoid stroma with the myxoid area containing a prominent network of branching capillary-sized blood vessels (Figure 2e). Sections were immunohistochemically stained with S-100, chromogranin, neurofilament, bcl-2, CK MNF 116, EGFR, vimentin, S-100, CD 99, CD 117, CD34, actin and desmin antibodies. The tumour was positive for vimentin (Figure 2e) and bcl-2. The final diagnosis was LGFMS.

Discussion

LGFMS is a rare spindle cell sarcoma characterised by a deceptively bland histology and a paradoxically aggressive clinical course, including relatively frequent recurrence and metastasis [1–3]. LGFMS was first described in 1987 [5]. LGFMS has been recognised in the latest World Health Organization classification of soft-tissue sarcomas as a distinctive variant of fibrosarcoma. The most common clinical presentation is a firm, slow-growing painless mass. Most tumours occur in the subcutaneous or deep soft tissues of the lower limbs,

shoulders or thoracic wall with others in the axilla, groin, buttocks, neck, mesentery and retroperitoneum [1–4]. LGFMS is not accompanied by typical macroscopic evidence of malignancy, such as necrosis, haemorrhage, an infiltrative border or nodularity [2, 5]. Recent studies [1, 6] have found that local recurrence (9%) and distant metastases (6%) are less frequent after aggressive surgery. The most common metastatic site is the lungs. A long follow-up period has been recommended because of the potential for late metastasis. The treatment of choice is wide local excision to reduce recurrence [1, 2].

Macroscopically, LGFMS is a well-circumscribed and encapsulated mass, indicative of a benign lesion. The cut surfaces of such tumours have a firm, fibrous yellow-white appearance with glistening areas secondary to the accumulation of myxoid ground substance. Histologically, these tumours are characterised by bland, benign-appearing spindle cells arranged in a whirling growth pattern with intermingled fibrous and myxoid stroma and without necrosis or haemorrhage. The degree of cellularity is low to moderate, mitotic figures are uncommon and nuclear pleomorphism is usually absent or slight. A prominent and rich capillary vascular

Table 1. Summary of reported cases of low-grade fibromyxoid sarcoma with MRI

Age/sex (reference number)	Location	T ₁ weighted images	T ₂ weighted images	Contrast-enhanced T ₁ weighted images
37/M (2)	Pleura	Hypointense	Hypointense	Heterogeneous enhancement
10/M (3)	Shoulder	Hypointense	Heterogeneously hypointense	Heterogeneous enhancement
64/M (4)	Small bowel mesentery	Hypointense to intermediate	Heterogeneously low-to-high signal intensity, multiple intralesional nodules	Heterogeneous enhancement
34/M (7)	Thigh	Hypointense to intermediate	Heterogeneously low-to-high signal intensity, multiple intralesional nodules	Heterogeneous enhancement
47/M (7)	Chest wall	Hypointense to intermediate	Heterogeneously low-to- high signal intensity	Heterogeneous enhancement
29/F (9)	Lower leg	Heterogeneously intermediate	Heterogeneously low-to-high signal intensity, multiple intralesional nodules	Heterogeneous enhancement
74/M (10)	Chest wall	Heterogeneously intermediate	Heterogeneously low-to- high signal intensity	Heterogeneous enhancement

M, male; F, female.

network is commonly present in myxoid areas [4, 5, 7]. Immunohistochemically, the cells of LGFMS stain strongly and diffusely for vimentin and bcl-2 but do not usually express smooth muscle actin, desmin, S-100 protein, neuron-specific enolase or cytokeratin [8, 9]. Recently, cytogenetic and molecular genetic analyses using reverse transcription polymerase chain reaction (RT-PCR) have been used as ancillary diagnostic tools for LGFMS [9].

Several case reports have described the variable MR findings observed in patients with LGFMS (Table 1) [2–4, 7, 9, 10]. The thigh masses in our two patients showed similar MR findings; the outer portions of the masses showed large hypointense signals on T₁ weighted and T₂ weighted images, a characteristic of such tumours as most soft-tissue tumours display intermediate- (occasionally high-) signal intensity on T₂ weighted images. The inner portions of the masses of our patients yielded large hyperintense areas on T₂ weighted images. On T₁ weighted images, these inner portions were slightly hyperintense in patient 1 and distinctly hyperintense in patient 2, suggesting haemorrhage with cystic degeneration. These findings were in disagreement with those of previous reports [2, 5, 7], which found that LGFMS does not contain areas of haemorrhage. In our patients, areas of haemorrhagic signal were seen in sites of cystic degeneration in these low-grade tumours, with no histological evidence of necrosis to suggest a high-grade sarcoma.

In case 1, the outer portion on fat-suppressed contrast-enhanced T₁ weighted images appeared as multiple nodules, consistent with findings of multiple intralesional nodules in several previous reports [4, 7, 9, 10]. Such nodules may be related to the alternating fibrous and myxoid areas in LGFMS. However, in contrast with previous findings [7], we did not observe a target-like appearance on T₂ weighted images.

Our MR findings were in good agreement with the pathological features of LGFMS, which consists of tumour cells with fibrous or myxoid stroma. In general, fibrous tissue components appear hypointense on T₁ weighted and T₂ weighted images and show slight enhancement. Myxoid tissue components are hypointense on T₁ weighted images, hyperintense on T₂ weighted images

and show variable enhancement on contrast-enhanced T₁ weighted images [2–4, 7].

Based on imaging characteristics, the differential diagnosis of LGFMS can include many types of benign and malignant fibrous- and myxoid-containing tumours, including myxoid malignant fibrous histiocytoma (myxofibrosarcoma), myxoid liposarcoma, fibromatosis, myxoid neurofibroma and solitary fibrous tumour [2–4, 7, 10]. In contrast to LGFMS, myxoid malignant fibrous histiocytomas are prone to necrosis and haemorrhage. However, haemorrhage was observed in both of our patients. Myxoid liposarcomas are hyperintense on T₂ weighted images in contrast to the large externally hypointense signals seen on T₂ weighted images from our patients. Fibromatosis is usually hypointense on T₂ weighted images but may also show heterogeneous hyperintensity, reflecting marked cellularity or myxoid tissue. Neurogenic tumours are characterised by the entry and exit of nerves [11].

In summary, we have reported the MR findings in two young women with haemorrhagic LGFMS presenting in the thigh.

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