CASE REPORT

Mammary-type myofibroblastoma of the psoas

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SUMMARY

Mammary myofibroblastoma is a rare, benign, mesenchymal neoplasm that was first described in the breast in 1987. However, extra-mammary-type myofibroblastoma (MTMF) was not described until 2001 as a distinct entity. We report a case of extra-MTMF arising from the retroperitoneum, which was encountered in our clinic. We also completed a review of the literature using PubMed in patients with extra-MTMF.

BACKGROUND

Mammary myofibroblastoma is a rare, benign, mesenchymal neoplasm that was first described in the breast by Wargotz *et al*¹ in 1987.² However, mammary-type myofibroblastoma (MTMF) was first described as a distinct entity in 2001 by McMenamin *et al*.³ The two are histologically and immune-phenotypically identical.⁴

A review of the literature revealed that there have been ~146 cases of extra-MTMFs published to date. These have occurred in various and unexpected locations; however, none have been described to arise from the psoas muscle. We present the case of a 69-year-old man who was incidentally found to have an extra-MTMF arising in the retroperitoneum from the psoas muscle. In addition, a comprehensive review of the collective properties of extra-MTMF is provided.

CASE PRESENTATION

A 69-year-old man presented to our clinic for the follow-up of an incidentally detected retroperitoneal mass on CT scan. He was diagnosed with prostate cancer 1 month ago and was undergoing the subsequent workup which included imaging studies. He was asymptomatic during this period. There was a past history of lumbar disc displacement, and his only surgery included an inguinal hernia repair. He had a history of tobacco use but quit 25 years ago. Family history included a paternal uncle with pancreatic adenocarcinoma. On examination, his vital signs were within normal limits. His abdomen was soft, non-tender and non-distended. Furthermore, there was an absence of organomegaly, and no masses were felt on palpation of the abdomen. Laboratory studies were within normal limits. An MRI of the abdomen with and without intravenous contrast revealed a 7.2×5.4×5.8 well-circumscribed, encapsulated mass arising from the lower aspect of the left psoas muscle (figures 1 and 2). The mass was homogeneously T2 hyper-intense and contained a 2.5 cm focus of fat within the inferior aspect of the psoas muscle (figure 3). There was no retroperitoneal

adenopathy. Of note, an MRI conducted 6 months prior for lumbar disc herniation did not reveal a retroperitoneal mass.

On suspicion for a retroperitoneal sarcoma (particularly liposarcoma), the mass was biopsied. CT-guided needle biopsy revealed findings of a low-grade myxoid neoplasm. Because of the possibility of sampling error and the inability to rule out a low-grade malignancy, the patient chose to undergo surgery.

Radical resection of the left retroperitoneal mass with partial resection of the psoas muscle was performed without complications. A frozen section revealed a mesenchymal tumour, and pathologic evaluation of the tumour revealed an MTMF (figures 4 and 5). On immunohistochemistry staining, the mass was found to co-express CD34 and desmin (in the absence of myogenin). Additionally, FISH for MDM2 amplification to rule out liposarcoma was negative as were several immunostains (SOX10, S-100 protein, smooth muscle actin, pan muscle actin and STAT6) to exclude common benign tumours (benign nerve sheath tumour, solitary fibrous tumour and leiomyoma).

OUTCOME AND FOLLOW-UP

The patient's postoperative course was uneventful, and he was subsequently discharged. The patient has been followed for 7 months without recurrence.

DISCUSSION

On reviewing the literature in detail, we found 146 previously published cases (from 2001 to 2016) of extra-MTMF (table 1).^{2–21}

Of note, a series by Howitt *et al*¹⁸ comprised a large portion of the published cases and is the largest series of MTMF to date. Of the patients reported in our review, 63% were men (M:F ratio of 1.8). The mean, range and SD of the age of the patients reported were 53, 4–96,¹⁸ 18.2, respectively. The mean tumour size in the series of Howitt *et al*¹⁸ was 6.6 cm (range of 1–22 cm). The anatomic distribution of these tumours was widespread, with the most common, general site of involvement being the groin/inguinal region.^{5 6 9 12 18 21} Therefore, it has been theorised that extra-MTMFs occur along the embryonic milk-line from the axilla to the medial groin; however, these tumours have frequently been found in other locations as well. Extra-MTMFs have been described to occur in the paratesticular region,⁶ scrotal sac,²¹ seminal vesicle,¹³ prostate,⁴ vulva,^{9 12} perianal region,¹⁰ buttock region,¹¹ periumbilical region,²⁰ internal oblique muscle,¹⁹ liver,² mandibular angle,⁸ eye,¹⁵ popliteal fossa,⁷

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Figure 1 MRI (coronal) of the abdomen and pelvis with and without contrast: a 7.2×5.4×5.8 well-circumscribed, encapsulated mass (yellow arrows) arising from the lower aspect of the left psoas muscle (asterisk).

Figure 2 MRI (coronal) of the abdomen and pelvis with and without contrast: a homogenously T2 hyper-intense, 7.2×5.4×5.8, well-circumscribed, encapsulated mass (yellow arrows) arising from the lower aspect of the left psoas muscle (asterisk).

axilla,²¹ thigh¹⁷ and big toe.¹⁶ Furthermore, MTMF arising in the retroperitoneum as in this patient was described in one case series.¹⁸ To the best of our knowledge, this patient's case is the

first primary, extra-MTMF arising from the psoas muscle. Of note, there was one case in the literature of secondary mammary myofibroblastoma of the psoas muscle due to

Figure 3 MRI (axial) of the abdomen with and without contrast. The mass (yellow arrows) was homogenously T2 hyper-intense and contained a 2.5 cm focus of fat within the inferior aspect of the psoas muscle (asterisk).

Figure 4 100× (A) and 400× (B) photomicrographs: bland spindle cells with adipocytes as an integral part of the process.

Figure 5 400× photomicrographs (A–D): (A) well-margined lesion (bottom-right) with adjacent adipocytes and skeletal muscle (top-left). (B and C) Short bands of collagen are noted between bland spindle cells. (D) Occasional small vessels display perivascular hyalinisation.

metastasis from the breast.²² Despite this diffuse anatomic variation, most patients are asymptomatic and detected incidentally as in this patient's case. If symptomatic, the most common presentation is a painless, non-tender, enlarging mass over the course of days-years.¹⁸

Extra-MTMF frequently has a fatty component that can be visualised on CT scan and MRI. Hypodense regions with negative Hounsfield units (–10 to –100 HU) on CT scan are indicative of fat.² In addition, MRI with T1/T2 imaging can demonstrate adipose tissue. The value of CT scan and MRI in the diagnosis of myofibroblastoma has been described by many authors.^{19–23} The most appropriate diagnostic test for a clinically or radiologically evolving soft tissue mass is a biopsy, due to suspicion of sarcoma. In this patient's case, suspicion for a retroperitoneal liposarcoma prompted a CT-guided needle biopsy.

The differential diagnosis for extra-MTMF includes neoplasms of a benign (benign nerve sheath tumour, solitary fibrous tumour, leiomyoma, spindle cell lipoma (SCL), cellular angiofibroma, angiofibroblastoma, angiomyxoma and soft tissue perineurioma) and malignant (liposarcoma, dermatosarcoma protuberans) nature.^{14–17} However, extra-MTMF is most often confused with SCL since they are both composed of CD34 positive spindle cells interspersed with adipose tissue.^{5–17} Additionally, both of them may demonstrate a loss of the RB1 and FOXO1 gene on chromosome 13q14.^{5–17–21–24–25} The differentiating factor lies in SCL's lack of desmin staining in contrast to extra-MTMF which generally stains for desmin.^{5–17} Furthermore, the adipose tissue component of extra-MTMF is less than that in SCL.¹⁷

Extra-MTMFs are characteristically white to grey, well-circumscribed, non-encapsulated tumours that lack haemorrhage

Table 1 MTMF: literature review

Year	Case number	Sex/age	Site	Desmin	CD34	SMA	Recurrence
2001 ³	*	M/52; F/58; M/52; M/67; M/62; M/35; M/60; M/53; F/43	Anterior abdominal wall, posterior vaginal wall, left inguinal region, mid-back, right groin, left inguinal region, left paratesticular area, right inguinal region, left buttock	100%+	89% +	33% +	—
2006 ⁵	1	M/37	Groin	+	+	—	—
2007 ⁶	1	M/85	Left paratestis	+	+	NA	—
2008 ⁷	1	M/36	Left popliteal fossa	+	+	—	—
2008 ⁸	1	F/45	Head/neck	+	+	+	—
2009 ⁹	1	F/56	Vulva	NA	NA	NA	—
2010 ¹⁰	1	F/40	Perianal	+	+	—	—
2011 ¹¹	1	F/40	Right buttock	+	+	—	—
2011 ¹²	1	F/80	Vulva	+	+	NA	—
2011 ¹³	1	M/79	Right seminal vesicle	+	+	+	—
2012 ¹⁴	*	F/9	Left groin	+	+	NA	—
2014 ²	1	F/46	Liver	+	+	—	—
2014 ¹⁵	1	F/66	Left eye	+	+	NA	—
2014 ¹⁶	1	F/83	Left big toe	—	+	NA	—
2015 ¹⁷	1	M/50	Right thigh	+	+	—	—
2016 ¹⁸	128†	94 (66%) M and 49 (44%) F/Mean age 54; range 4–96 years	Groin/inguinal region (65), chest wall/axilla (7), trunk (17), thigh (11), lower leg (3), foot/toe (4), intra-abdominal/retroperitoneal (14), vaginal (2), head/neck (3), upper extremity (2)	91%+	89% +	37% +	1 (<1%)
2016 ¹⁹	1	F/51	Internal oblique muscle	NA	—	+	—
2016 ²⁰	1	M/23	Periumbilical	NA	NA	NA	—
2016 ⁴	1	M/51	Prostate	+	+	NA	—
2016 ²¹	2	M/30, M/58	Right scrotal sac, right axilla	+/+	+/+	—/—	—

*Included in series of Howitt *et al.*¹⁸

†The series included 143 patients; however, 15 occurred in the breast and were excluded from this table.

MTMF, mammary-type myofibroblastoma.

or necrosis.^{2 19} However, in this patient's case, the mass was relatively encapsulated. They are typically composed of spindle-shaped cells arranged in fascicles, interspersed within a collagenous stroma.^{2 4 14 17–19} There is also a variable adipocytic component.^{2 18} These findings were seen in this patient's case on histological examination (figures 4 and 5). Furthermore, intralesional fat lobules and intranuclear grooves have been described.¹⁷

On immunohistochemistry staining, extra-MTMFs are typically positive for desmin (>91% of cases) and CD34 (89% of cases).¹⁸ Of note, the markers with the highest sensitivities to detect extra-MTMF are CD34 (89%) and desmin (91%).⁴ Additional tests that can narrow the differential diagnosis include smooth muscle actin (SMA), epithelial membrane antigen (EMA), S100 and SOX10 (to rule out melanoma), MDM2 (to rule out liposarcoma), STAT 6 (to rule out solitary fibrous tumour) and CDK4. The loss of the RB and FOXO1 genes on chromosome 13q14 can be seen using fluorescence in situ hybridisation.^{5 21 24}

Since extra-MTMFs are frequently asymptomatic and detected incidentally, the treatment is most often elective resection. This patient underwent surgical resection since the possibility of a low-grade malignancy could not be ruled out. Extra-MTMFs have an excellent prognosis after resection. These lesions do not metastasise, and recurrence has only been documented once after 20 years, even with positive excision margins.¹⁸ Therefore, when faced with an incidentally detected or slow-growing soft tissue mass, it is necessary to obtain a biopsy to rule out malignancy. If malignancy cannot be ruled out or the patient is symptomatic, surgical resection is appropriate.

Learning points

- ▶ Mammary-type myofibroblastoma (MTMF) is a benign, mesenchymal neoplasm that was first described in the breast; however, extra-MTMF is now being documented more frequently in the literature.
- ▶ In the presence of an incidentally detected or slow-growing soft tissue mass, it is necessary to obtain a biopsy to rule out malignancy. If malignancy cannot be ruled out or the patient is symptomatic, surgical resection is appropriate.
- ▶ Extra-MTMF is a rare tumour; therefore, pathological examination should include immunohistochemistry staining (CD34, desmin, SMA, EMA, MDM2) to rule out differential diagnosis.
- ▶ Despite the numerous classifying factors and anatomic locations, extra-MTMF essentially has no risk of metastasis or local recurrence, even in the presence of positive excision margins.¹⁸

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