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Ossifying fibromyxoid tumor in the lower extremity mimicking a sebaceous cyst. Case report and literature review

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ABSTRACT

INTRODUCTION: Ossifying fibromyxoid tumor (OFMT) is an uncommon soft tissue neoplasm, with malignant potential and unclear histogenesis. OFMT exhibits a spectrum of histopathologic features including benign (typical), atypical and malignant subtypes. To the best of our knowledge, about 300 cases have been reported worldwide. We present the first reported case from Qatar.

PRESENTATION OF CASE: A 36-year old Egyptian male, with no comorbidities was admitted electively as a day case for excision of left thigh suspected sebaceous cyst under local anesthesia. History, physical examination and soft tissue ultrasound imaging were unremarkable. Intraoperatively, the patient was found to have a hard-calcified mass adhering to the surrounding fascia which was excised en bloc. The histopathology result was of ossifying fibromyxoid tumor. The post-operative course along with 40 months follow-up were uneventful in terms of surgical complications and recurrence.

DISCUSSION: OFMT has marked features in terms of cytology. Though it is difficult to diagnose preoperatively, it should be considered in tumors involving soft tissue that demonstrate prominent ossification and calcification.

CONCLUSION: OFMT is a rare soft tissue neoplasm, and should be considered as a differential diagnosis in any subcutaneous swelling with a bony component. All OFMT patients should undergo a long course of follow-up to rule out and assess any recurrence or metastasis in the malignant variants.

1. Background

Ossifying fibromyxoid tumor (OFMT) is a rare soft tissue neoplasm that can arise anywhere but mostly affects the subcutaneous layer presenting as a superficial swelling/mass [1]. OFMT is seen in adults of all ages, mostly in middle-aged individuals, with a slight male preference (1.5 times) [1]. Clinically, OFMT presents as a single well-circumscribed, slow growing painless mass ranging from 1 to 14 cm (average 4–5 cm) with normal overlying skin, mostly attached to the underlying structures such as tendons, fascia or skeletal muscles [2]. OFMT runs a chronic clinical course ranging from 1–20 years or longer [3]. Histologically, OFMT is yet of unclear differentiation but a typical tumor demonstrates a lobular and encapsulated pattern with surrounding lamellar bony component; and is composed of oval or spindle-shaped uniform cells with vesicular nuclei [4]. The malignant variant of OFMT has a 20–27% tendency of local recurrence and though extremely rare, may exhibit metastasis to soft tissues, including the opposite extremity/thigh, lungs and adrenals [5].

In 1989 Enzinger et al. reported 59 OFMT cases, most of which demonstrated histological benign appearance which he categorized as typical [1]. In 2003, Folpes et al. reviewed 70 cases where he proposed that OFMT, due to its variation, should be classified into typical, atypical and malignant variants [2]. To the best of our knowledge, globally, only about 300 cases OFMT were reported to date, of which about 90 cases were in the proximal lower limb (thigh). We report the first OFMT in the thigh to be diagnosed in Qatar and the second reported from the Arabian Gulf region and middle eastern countries. We report this case in line with the updated consensus-based surgical case report (SCARE) guidelines [6].

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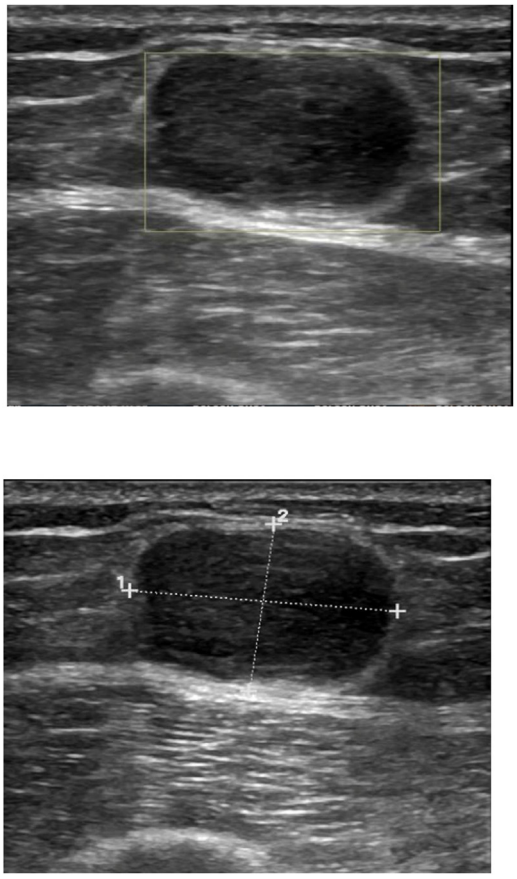

Fig. 1. Ultrasound (US) of the left lateral thigh showing a well-demarcated hypoechoic spherical cystic lesion, suggestive of epidermal inclusion cyst B 1: 2.5 cm, 2: 2 cm.

2. Case presentation

A 36 year old Egyptian male presented to our surgical outpatient clinic, at Hamad General Hospital (largest tertiary care facility in Doha, Qatar) with a 14-month history of a left thigh painless swelling. The swelling was gradually increasing in size, and then became static with no further growth. He had no comorbidities, and systemic review was unremarkable. The patient is a non-smoker, was not on any medications, and with no relevant past medical history or family history.

Clinical examination revealed a well build, vitally stable male with a swelling in the mid lateral aspect of the left thigh, about 3 cm in diameter. The swelling was firm, non-tender, with normal overlying skin and no regional lymphadenopathy. Prior to this index visit, he had undertaken a soft tissue ultrasound scan (US) of the lower limb that revealed a well-defined soft tissue mass within the subcutaneous fat abutting the tensor fascia lata, approximately 2.4 × 1.9 cm with minimal vascularity, suggestive of epidermal inclusion cyst (Fig. 1). At our facility, he was booked by the surgical team as a day case for surgical excision under local anesthesia, after which he underwent standard preparation for the procedure which was undertaken by an experienced general surgeon. Under aseptic measures, local anesthesia was infiltrated around and above the swelling, and a vertical incision about 4 cm was made in the mid part of the lateral aspect of the left thigh. After separation of the subcutaneous layer, the impression was a hard-calcified mass of about 4 × 3 × 1.5 cm, adhering to the surrounding fascia. It was excised en block and sent for histopathological assessment in a formalin filled container (Fig. 2).

Histopathology revealed a neoplasm with hypercellular and hypocellular areas, with bundles of interspersed collagen fibers and a well-vascularized stroma. The cellular component comprised lobules of uniform rounded nuclei and small distinct nucleoli with pale cytoplasm and indistinct cell boundaries. The tumor cells were strongly positive with Vimentin, S100, CD56, Calponin, IN1, CD99 and patchy CD57 (Fig. 3).

The post-operative period was uneventful. In the initial period after the surgery, the wound showed optimal healing with no complications, and there were no complications or signs of local recurrence or metastasis up to 40 months follow up (Fig. 4).

3. Discussion

OFMT is a relatively rare soft tissue neoplasm of uncertain histogenesis. We report a case of a middle-aged Egyptian male with a thigh OFMT that mimicked the US appearance of a sebaceous cyst.

In terms of age and gender, OFMT affects adults of all ages, mostly the middle-aged, with slight male preference (1.5 times) [7]. Our patient's age is in agreement, and the literature review we undertook (Table 1) showed an age range of 24–76 years, with most patients being middle-aged. Table 1 did not depict a gender preference, as the 8 patients we identified had an equal gender distribution.

In terms of site, OFMT is common in the extremities [1], although it has also been reported in the soft parts of the head and neck [8,9].

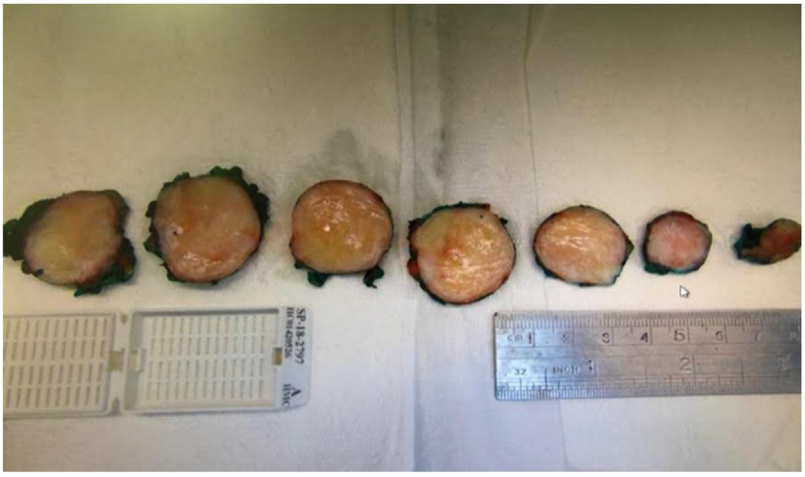

Fig. 2. Sections of the excised specimen showing rubbery fibroadipose tissue in the centre surrounded by bony periphery.

Table 1
Reported cases of myxoma in lower limb in last 20 years.

Case	Ag	G	LO	D	Dur	Clinical pic	Pre-op investigation	Intra-op[a]	NG	C	MC	PC	F/R
Current case	36	M	Thigh	3	1.1	Firm non-tender swelling, normal overlying skin	US: well demarcated subcutaneous spherical hypoechoic cystic lesion suggestive of epidermal inclusion cyst	White/yellow, hard lobulated rounded mass, thick fibrous capsule, WC, adhered to subcutaneous layer	L	L	<1/50 HPF	T	40, no R
Kumari 2020 India	55	F	Thigh	7	20	Non-tender well defined swelling, firm consistency, healthy overlying skin, not attached to surrounding	US: well-defined heteroechoic mass in subcutaneous plane FNA: moderately cellular, singly scattered, monomorphic few large-sized epithelioid/ovoid cells in variably myxoid and fine fibrillary pink matrix, round/oval nuclei with finely dispersed chromatin, inconspicuous nucleoli, moderate/ abundant amount of eosinophilic to amphophilic cytoplasm	Spherical, WC, encapsulated tumor in subcutis, incomplete layer of peripheral lamellar bone	–	–	<1/50 HPF	T	–
Umer 2019 Pakistan	32	M	Thigh	6 × 5 × 4	1	No significant findings	X-ray: soft tissue swelling, medial focal calcification MRI: rounded well-defined lesion, lobulated margins, subcutaneous location T1-weighted sequence: isointense to muscles. T2-weighted sequence: heterogeneous high signals, low signals foci of calcifications corresponding with plain x-ray	Focally attached skin, cut surface shows multi nodular tumour, firm, focally gritty areas representing bony part	–	L	>2/50 HPF (4)	A	24, no R
Ahmed 2015 USA	56	M	Groin	3.5 × 2.9	6	–	CT: peripherally calcified, round lesion, anterior to adductor musculature	WC, dense solid cut surface, areas of calcifications	L	L	>2/50 HPF	T	–

Table 1 (*Continued*)

Case	Ag	G	LO	D	Dur	Clinical pic	Pre-op investigation	Intra-op[a]	NG	C	MC	PC	F/R
							FNA: pauci-cellular myxoid material CB: cords of cytologically bland cells in fibromyxoid stroma Immunohistochemical stains: neoplastic cells focally positive for S-100 protein, negative for Desmin, smooth muscle actin, CD34 and cytokeratin AE1/AE3						
Chen 2011 Taiwan	30	F	Thigh	0.6 × 0.5 × 0.5	3	WC, firm, asymptomatic subcutaneous mass	—	—	L	L	No mitosis	T	—
Al-Brahim 2008 Kuwait	60	F	B	7.5 × 5.5 × 4	—	Hard mobile mass, no skin changes	None	Hard, difficult to cut, encapsulated lobulated, smooth external surface	L	L	<2/50 HPF	T	—
Choi 2007 Korea	24	M	B	5 × 3 × 3	0.5	Hard movable WC mass	US: well-demarcated oval hypoechoic mass, marginal calcifications with posterior shadowing at both lateral aspects, avascular lesion with posterior enhancement	Solid WC, lobulated spherical mass, thick fibrous pseudo capsule	L	L	<2/50 HPF	T	—
Cibull 2007 USA	76	F	B	3	—	WC mass	—	—	H	H	>2/50 HPF (17)	M	—

For space considerations, only the first author is cited. A: Atypical; Ag: Age; B: Buttock; CB: Core Biopsy; C: cellularity; D: dimension in cm; Dur: duration in years; F/R: Follow-up/ Recurrence in months; F: female; FNA: Fine needle aspiration; G: Gender; H: high; HPF: high power field; L: Low; LO: Location; M: Male; M: Malignant; m: month MC: Mitotic count/ 50 HPF ; NG: Nuclear grade; No: not reported; PC: Prognostic classification; R: recurrence; T: Typical; US: ultrasound; WC: well-circumscribed.

[a] Intra-op gross appearance.

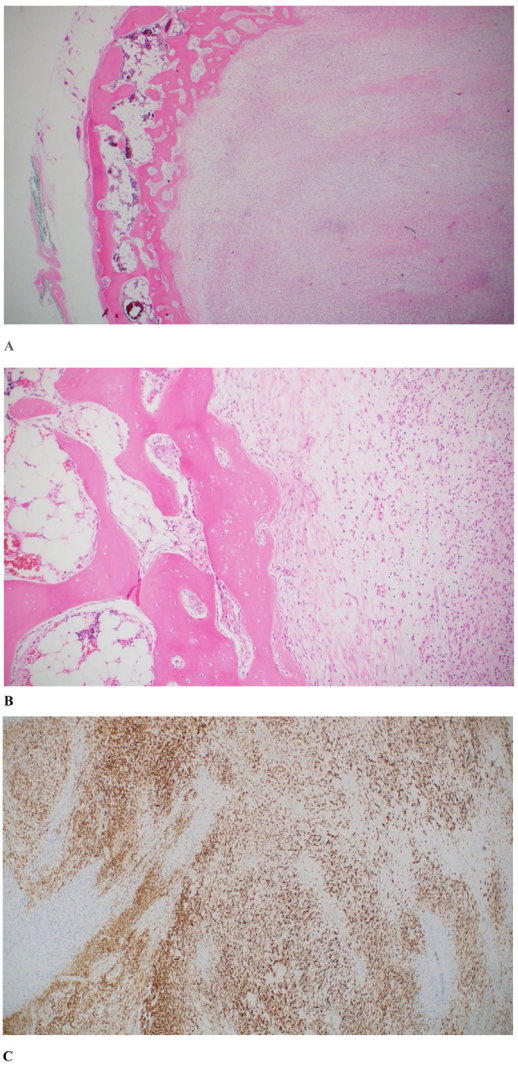

Fig. 3. A. Myxoid spindle cell soft tissue neoplasm surrounded by well-defined fibrous capsule containing trabecular bone and intertrabecular marrow elements; **B**. High power shows bone and intertrabecular marrow spaces with adjacent hypercellular and hypocellular areas of uniform rounded nuclei and small distinct nucleoli with pale cytoplasm and indistinct cell boundaries; **C**. Spindle cells are positive with S100 immunostain.

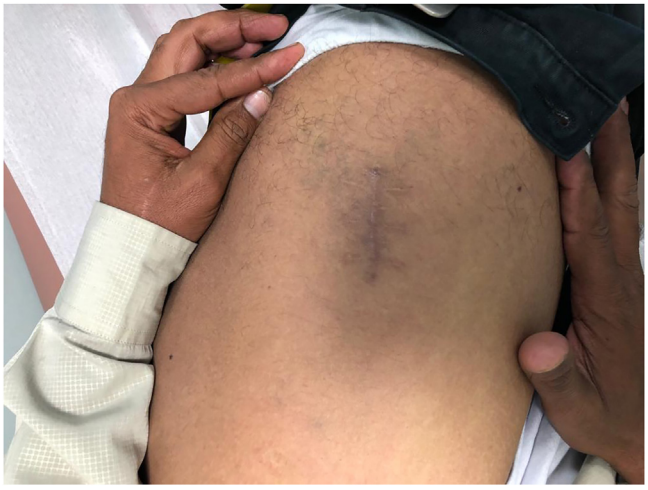

Fig. 4. Post excision follow-up.

As for presentation, OFMT presents as an asymptomatic slow growing deeply-seated mass attached to the surrounding structures such as fascia or skeletal muscles [7]. Our case is in support, presenting with a mid-lateral part of the left thigh, painless firm to hard mass of short duration attached to surrounding structure with no overlying skin changes (no central punctum as seen in sebaceous cyst). Table 1 reveals that in most of the cases, the main presentations agree with such description. However, one patient had no significant clinical findings [10].

As for the clinical course, OFMT follows a chronic course [2,7], in agreement with our patient where the mass gradually increased in size for 8 months, followed by a 4 month 'static' period of stable size. As for size, OFMT ranges between 1–14 cm (average 4–5 cm) [2,7], and our case had a 3×2 cm mass which is relatively small, although Table 1 shows that a similar size swelling was observed in a 76 year old female [11].

In connection with imaging, US features of OFMT appear as an avascular well-demarcated mass with an incomplete ring of peripheral and scattered calcifications in the substance of the neoplasm [17]. This was not the exact picture we observed, where the US revealed a well demarcated subcutaneous spherical hypoechoic cystic lesion with neither peripheral bony/lamellar rim enhancement, nor posterior shadowing, which suggested epidermal inclusion cyst (sebaceous cyst) (Fig. 2). Our patient was booked for excision without further imaging. Interestingly, Table 1 reveals that US is not regularly used in the diagnosis of OFMT, where only two of the reported cases underwent preoperative US [12,13].

Further radiological imaging as computed tomography (CT) is recommended where, in 60–70% of cases, OFMT appears as a nodular soft tissue mass with an incomplete peripheral rim of ossification [18]. Our review (Table 1) observed that only one of the reported cases underwent CT scanning which revealed a peripherally calcified, round lesion, anterior to the adductor musculature, which is agreement with the suggested description [14]. When CT is not conclusive, MRI can be utilized, where OFMT usually appears isointense to muscles on T1-weighted images and shows intermediate to high signal intensity on T2-weighted images due to high degree of vascularity in the atypical variant, which may suggest hemorrhage, leading to a differential diagnosis of myositis ossificans or ossifying hematoma [7]. Table 1 depicts that the only reported case of thigh OFMT during the last 2 decades had preoperative MRI assessment, and showed findings in agreement with the above-mentioned MRI description [10].

As for preoperative biopsy and fine needle aspiration (FNA), only two reported cases underwent preoperative FNA (Table 1), of which OFMT diagnosis was narrowed in the differentials after correlating both the FNA and the imaging findings in one case. For these patients, FNA demonstrated singly scattered, monomorphic medium- to few large-sized epithelioid to ovoid tumor cells set in a variably myxoid and fine fibrillary pink matrix in the first patient; and pauci-cellular myxoid material along with cords of cytologically bland cells within a fibromyxoid stroma in the other patient [13,14]. Other differential diagnosis fitting such descriptions are sclerosing epithelioid fibrosarcoma and low-grade fibromyxoid sarcoma [13,14].

As regards to the surgical approach, evidence recommends complete excision of OFMT [1,2,7]. In our case, initial intraoperative assessment revealed a subcutaneous white to yellow, hard, lobulated rounded mass with thick fibrous capsule, well circumscribed and adhered to the surrounding subcutaneous layer. It was cautiously separated and excised en block keeping it intact with no residue. There were no complications and the postoperative period was uneventful. None of the reported cases had complications in

term of surgery (Table 1), and almost all patients underwent complete surgical excision in keeping with the recommended surgical approach of OFMT.

Gross examination of excised tumour revealed a white to yellow in colour and firm to hard in consistency (4 × 3 × 1.5 cm). The cut surface was tan white, rubbery fibroadipose tissue in the centre surrounded by bony periphery (Fig. 2). Microscopically (Fig. 3A and B), it revealed a myxoid soft tissue neoplasm surrounded by well-defined fibrous capsule containing trabecular bone and intertrabecular marrow elements. The neoplasm had hypercellular and hypocellular areas with bundles of interspersed collagen fibres and a well-vascularized stroma. The cellular component comprised lobules of uniform rounded nuclei and small distinct nucleoli with pale cytoplasm and indistinct cell boundaries. Rare mitoses were seen (2 mitotic figures/50 high power fields). There were scattered inflammatory cells (mainly CD3 + and CD20 + small lymphoid cells) with scattered CD68 positive cells. The tumour cells were strongly positive with Vimentin, S100 (Fig. 3C), CD56, Calponin, IN1, CD99 and patchy CD57. The following markers were negative: CD68, CD34, ALK1, SMA, Desmin, Caldesmon, MNF116, Ae1/Ae3, p63, MelanA, HMB45, GFAP, NSE, NF, SOX10 and EMA. The proliferation fraction (Ki67) was less than 1%. Our case resulted to be of the typical variant as most of the reported OFMT cases in Table 1 [12–16], with the exception of two patients that were atypical and malignant variants respectively [10,11].

In terms of recurrence and follow-up, OFMT was categorized into three variants: benign (typical) with low nuclear grade, low cellularity and a mitotic rate < 2/50 HPF; malignant OFMT with a high nuclear grade, high cellularity and a mitotic rate > 2/50 HPF; and intermediate (Atypical) [2,7]. Our patient's specimen was of the typical variant and though it is only recommended that only the atypical and malignant variants should have long follow-up, our patient was followed for 40 months during which he underwent MRI and US with no signs of recurrence. Most of the reported cases (Table 1) were typical with the exception of two cases: an atypical variant followed up for 24 months with no recurrence [10]; and a malignant variant where neither the duration nor the follow-up outcomes were reported [11].

4. Conclusion

Though OFMT is a rare soft tissue neoplasm it should be considered as a differential diagnosis of subcutaneous swelling with bony component seen as peripherally calcified rim in imaging along with mineralized stroma and bland cytology within myxoid background seen in FNA. All OFMT patients (typical-malignant variants) should undergo prolonged follow-up course.

Declaration of Competing Interest

Nothing to declare.

Funding

Nothing to declare.

Ethical approval

Approved by Medical Research Center, Hamad Medical Corporation reference number (MRC-04-20-1104).

Consent

Written informed consent was not obtained from the patient. The head of our medical team has taken responsibility that exhaustive attempts have been made to contact the family and that the paper has been sufficiently anonymized not to cause harm to the patients or their families. A copy of a signed document stating this is available by the Editor-in-Chief of this journal on request.

Author's contribution

Sugad Mohamed: data collection, data interpretation, writing the paper. Walid El Ansari: study concept, data interpretation, writing the paper. Mohamed Ben-Gashir: labortaory and histopathology, data interpretation, editing the paper. Abdelrahman Abusabeib: study concept, data interpretation, editing the paper. All authors read and approved the final version.

Registration of research studies

Not applicable.

Guarantor

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Provenance and peer review

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