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Case Report

Mammary-Type Myofibroblastoma of the Vulva: A Case Report and Review of the Literature

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Abstract

Background: Mammary-type myofibroblastomas (MTMF) have been reported to occur at extramammary sites throughout the body of men and women of all ages, including the vulvovaginal and perianal area where they can pose diagnostic and therapeutic challenge due to indistinguishable clinical features from malignant tumors.

Case: A 35-year-old woman, para 1, gravida 3, presented with an asymptomatic 5 cm by 3 cm tumor located along the anterior half of the left labia majora near the clitoris. Following excisional biopsy under general anesthesia, histopathological and immunohistochemical studies showed features consistent with a diagnosis of MTMF.

Conclusion: Vulvar MTMF is a rare benign soft tissue neoplasm but it warrants appropriate marginal excisional diagnosis since it is indistinguishable from benign and/or malignant lesions of the vulva.

INTRODUCTION

Mammary myofibroblastoma was first described in a case series including 11 men and 5 women as a distinct, benign, mesenchymal tumor of the breast by Wargotz et al in 1987 [1]. Consequently, extramammary mammary-type myofibroblastomas (MTMFs) were originally thought to occur only along the embryonic milk line in ectopic breast tissue, similar to accessory breasts [2]. However, MTMF with morphological and immunophenotypic features similar to the breast counterpart have been reported at diverse sites of the body including superficial and deep soft tissue sites and viscera with the inguinal area and lower extremities being the most common [3].

Interestingly, in addition to their diverse localization, these tumors also affect a diverse patient population, including pediatric patients, with men being more commonly affected than women in, approximately, three to one ratio [3,4]. Kuyumcu et al., 2017) of the cases affecting women, very few have been described in the vulvovaginal [5-7] and perianal area [8]. (Zhang et al, 2010); Only two cases of MTMF have been reported in the vulva, one of which was a true collision tumor with intermingling components of eccrine adenocarcinoma of the vulva [6]. (Wei & Zhu, 2011) Herein, we report our experience with an additional case of MTMF found in the vulva and a review of the available literature on such lesions found in the vulvovaginal and perianal area.

IRB approval was not required because the case entailed provision of clinical care and collection of existing data, and patient confidentiality was maintained at all times. The patient has provided consent to publish her case.

CASE

A 35-year-old woman, para 1, gravida 3, BMI 25, presented with a left vulvar lesion. The patient stated that during the last two months, she noticed a lump/swelling in her left upper part of her vulva. There was no history of injury and no pain, discomfort or drainage was experienced at any time. Twelve years prior, the patient had been diagnosed with Cushing syndrome which required laparoscopic left adrenalectomy followed by daily cortisol treatment for adrenal insufficiency. One month following the adrenalectomy, the patient was also diagnosed with hyperparathyroidism treated by excision of a parathyroid adenoma and a right thyrothymic tract. Subsequently, she was also diagnosed with idiopathic seizures requiring anti-epileptic medications for the last 4 years.

Under general anesthesia, examination revealed a soft subcutaneous lesion, approximately 5 cm long and 3 cm wide, running along the anterior half of the left labia near the clitoris.

There was no evidence of trauma, induration or inflammation. The lesion was quite mobile and after the skin was incised, it was easily dissected and excised completely intact and the skin and subcuticular tissue were sutured.

The gross pathology was reported as a rubbery tan fibrous homogenous tissue measuring 5.0 x 2.6 x 2.1 cm. "Sections show a bland spindle cell lesion composed of widely spaced, blunt spindle cells with indistinct cytoplasm and abundant, intercellular ropey collagen. There is no evident mitotic activity, nuclear atypia or necrosis. There are numerous admixed thick and thin-walled blood vessels, and patchy areas of adipose tissue. The lesion extends to the margins of excision.

Immunohistochemical studies show lesional positivity for CD34 (patchy, moderate to strong), ER (diffuse, moderate to strong) and PR (patchy, moderate to strong). There is extensive loss of Rb in lesional spindle cells. Desmin, smooth muscle actin and STAT6 are negative".

Diagnoses considered were mammary-type myofibroblastoma, spindle cell lipoma, cellular angiofibroma, angiomyolipoma, angiomyofibroblastoma and aggressive angiomyxoma. After inter-departmental review, the features were interpreted as being consistent with MTMF.

At 3 months of follow-up, the wound had healed normally with no evidence of recurrent disease.

DISCUSSION

Histopathology

MTMF has been described as a lesion well-circumscribed, composed of haphazardly arranged fascicles of spindle-shaped cells admixed with interrupted adipocytes in a collagenous and myxoid background containing mast cells. The most common immunohistochemical profile seen in these tumors is diffuse positivity for CD34 and desmin as shown in our case [2,3,7] (McMenamin & Fletcher 2001; Howitt & Fletcher 2016; Magro et al. 2012).

Review of literature

We found very few cases of vulvovaginal and/or perineal MTMFs described in the literature with only two MTMFs described in the vulva. In 2009, Diwadkar and Barber reported the first vulvar MTMF in a 56-year-old woman who presented with complaints of a left vulvar mass, pelvic organ prolapse and urinary incontinence. Examination revealed a 3-cm, firm vulvar mass, located in close proximity to the Bartholin's gland. Following local excision, there was no evidence of recurrence over a period of 12 months [5].

In 2011, Wei and Zhu reported the first case of a true collision tumor containing a MTMF with intermingling components of eccrine adenocarcinoma occurring in a vulvar sweat gland. The patient was an 80-year-old Chinese woman who presented with an ulcerated mass involving the left labium majus that had been gradually enlarging for a year. She had no history of breast malignancy. After a complete vulvectomy, she was free of disease but died of heart attack 3 years later [6].

Sweat gland carcinomas of the vulva are rare and collision tumors composed of MTMF intermixed with eccrine adenocarcinoma of the vulva skin is extremely rare. However, this case underscores the importance of marginally excising all tumors of the vulva for histopathologic evaluation.

In 2012, Magro et al, reported on the clinicopathologic features of 10 cases of vulvovaginal myofibroblastoma (MF). Among these, 3 cases composed of spindle-shaped cells arranged in short fascicles with intervening thick collagen bands, closely reminiscent of MTMF were located in the vagina but no case of MTMF was located in the vulva. Two cases of myofibroblastoma were located in the vulva [7].

Finally, Zhang et al, described a unique case of a MTMF located in the perianal region. The patient was a 40-year-old female with no significant past history, who presented with a right side painless perianal mass, approximately 5 cm [8].

Histogenesis

The definitive cell origin and the molecular basis of MTFM remain unknown. McMenamin and Fletcher speculated that fibroblastic/myofibroblastic cells along the embryonic mammary line may be predisposed to forming MTMF [2]. This assertion however, does not account for the widely diverse location of these tumors, most of which are found in extramammary sites. Others have speculated that these lesions probably arise from uncommitted CD34+ stem cells capable of differentiating along several mesenchymal lines giving rise to spindle cell lipoma, solitary fibrous tumor, lipomatous hemangiopericytoma and angiomyofibroblastoma, all of which are probably related entities [2,9].

It is of interest to note that uterine fibroids which are composed of various degrees of cellular elements (myofibroblasts) and matrix (mainly collagen) are thought to originate from *myo-fibroblasts* which phenotypically appear like a chimera of myocyte and fibroblast. This, of course, raises the hypothesis that MTMF may have a similar origin to fibroids originating in myocytes being subjected to some kind of hypoxic insult or physical trauma leading to conversion of a myocyte to a myofibroblast and an exaggerated response of tissue repair. It appears that all elements of normal tissue repair are also present in these myofibroblastic tumors.

Significance

When these tumors are found in the vulva, the differential diagnosis includes all benign or malignant lesions of the vulva, some of which are epidermal cysts, follicular cysts, Bartholin's cysts, lipoma, adenoma phyllodes, syringe angiomas, extramammary Paget's disease, and adenocarcinomas [2].

Generally, MTMF are benign and no malignant counterpart or transformation to malignancy has been reported. Howitt and Fletcher reported on clinicopathologic characterization in a series of 143 cases (94 male, 49 female) of MTMF found throughout the body. The age of these patients varied widely, ranging from 4 to 96 years, with an overall predominance in the fifth and sixth decades. At a median follow up of 15.8 months (range, 0.2 to 174 months), available information in 54 (38%) cases indicated that no case had evidence of MTMF tumor recurrence or metastasis;

however, reportedly, 1 of the cases in this series was a tumor recurrence (slides from the original tumor were not available for review). No cases had recurrence of MTMF, even in the setting of positive excision margins (n=8) [3].

Although these tumors have been reported to be benign, at least one MTMF containing elements of eccrine adenocarcinoma of the vulva has been reported. In such, so called, collision tumors, the eccrine adenocarcinoma was determined to have a greater adverse effect on prognosis than the MTMF containing elements of eccrine adenocarcinoma of the vulva underscoring the importance of appropriate management of all vulvar tumors [6]. Wei & Zhu 2011 The major concern regarding these tumors found in the vulva is the uncertainty of being benign or malignant since they are indistinguishable from all other common variety benign or malignant tumors of the vulva. Therefore, like all other vulvar lesions, they require histological diagnosis and a complete marginal excisional biopsy has a good prognosis since no malignant behavior has been reported.

CONCLUSION

MTMFs have been reported at multiple extramammary sites in males and females of all ages, including the vulva. MTMF is a benign soft tissue neoplasm, and no malignant behavior has been reported after excision. Because it is extremely, rare, its presence in the vulva, warrants a complete marginal excisional biopsy since it is indistinguishable from most, if not all, other benign and malignant lesions of the vulva.

ETHICS AND INSTITUTIONAL REVIEW

Institutional review board (IRB) approval was not required because the case entailed provision of clinical care and collection of existing data, and patient confidentiality was maintained at all times. The patient has provided consent to publish her case.

PRECISE

Mammary-type myofibroblastoma in the vulva may present diagnostic challenge and marginal excisional biopsy is warranted.

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