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Case report

Primary myoepithelial carcinoma of the Vulva: Case of a rare tumor with malignant potential and review of literature

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1. Introduction

Myoepithelial cells are a specialized type of cell located in the salivary glands, breast, respiratory tract, skin and other organs usually located between the epithelioid cells and the basement membrane. They are thin, spindle shaped cells with cytoplasmic processes that extend to acinar and ductal-lining cells with both smooth muscle and epithelial features. Their major role in glandular tissue is the expulsion of saliva and the regulation of electrolyte exchange (Balachander et al., 2015). Myoepithelial carcinoma is a rare neoplasm of soft tissue that may arise from myoepithelial cells in these and other locations (Dimitrijevic et al., 2015). The most common location for myoepithelial carcinoma is the salivary glands, namely the parotid gland, but tumors have also been found in the skin, breast, lung and other soft tissues (Frost et al., 2014). To our knowledge, only seven cases of primary myoepithelial carcinoma of the vulva have been reported (Miyata et al., 2011; Fukunaga, 2003; Hinze et al., 1999; Khazeni et al., 2018; Meenakshi and McCluggage, 2009; Noronha et al., 2006). These tumors often present with lobulated, firm, yellow/white or tan masses that are most often painless (Hornick and Fletcher, 2003). It is a relatively aggressive tumor with the potential for distant metastasis, although it does possess unpredictable biologic behavior with few true prognostic indicators (Kong et al., 2015). Because of their rarity, myoepithelial carcinomas are often mistaken for other tumors of the soft tissue or epidermis, and there is limited information regarding disease causes and treatment options. Because of this, there is no accepted standard of treatment for myoepithelial carcinoma of the vulva. The current recommended definitive therapy for cutaneous myoepithelial carcinoma seems to be complete surgical resection with clear margins with some evidence suggesting systemic therapy such as doxorubicin, cyclophosphamide, and/or carboplatin may have some activity in soft tissue myoepithelial carcinomas (Chamberlain et al., 2020). More information regarding this malignancy is essential to broaden our understanding of disease progression and response to treatment as well as improve patient outcomes. Here we report a case of primary myoepithelial carcinoma of the vulva as well as patient and tumor characteristics of each of the existing cases (Table 1).

2. Case presentation

We present the case of a 40-year-old Caucasian female who reported to her primary gynecologist for evaluation of an enlarging mass in her right groin. The patient provided informed consent for her case to be reported in the scientific literature. Health maintenance records showed that she had a recent Pap smear the previous year that showed no abnormalities. She noted that she first felt a growth five months prior to evaluation and assumed it was an enlarged lymph node but became concerned when it tripled in size and became tender to touch. On initial exam, her primary gynecologist noted a large, mobile lesion in the right labia and pubic area. It was noted to be mildly tender without any overlying skin changes or associated inguinal lymphadenopathy. Initial

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differential diagnosis included possible lymph node enlargement versus a labial cystic fluid collection. Computed tomography demonstrated a round soft tissue mass in the right labia majora measuring 3.4 × 3.0 × 3.8 cm. She was referred to a Gynecologic Oncologist who evaluated her the following month. Examination by the gynecologic oncologist again noted a large, mobile mass in in the right labia measuring approximately 4 cm without overlying skin changes or palpable lymphadenopathy. Gynecologic Oncology recommended excision of the mass. Preoperative PET imaging was considered but ultimately forgone due to the presumed benign nature of the mass. She subsequently underwent an uncomplicated radical anterior vulvectomy with 2 cm margins; gross examination in the operating room was consistent with a necrotic vulvar mass. There was no lymphadenopathy noted at the time of the procedure. Pathology was reviewed at the Mayo Clinic and showed a labial spindle cell IN1 deficient malignant neoplasm favoring myoepithelial carcinoma. The tumor extended to the inked circumferential margins; depth of invasion was approximately 1.5 cm and the deep margin was negative. The patient's pathology was discussed at an interdisciplinary tumor board and re-excision with right groin node dissection was recommended. Chemotherapy was also considered but was not recommended at this time. Twenty days post-resection, a CT of the chest, abdomen and pelvis showed several pulmonary nodules all measuring less than 6 mm read as possible inflammatory changes. PET/CT was obtained 1 month following her initial resection and showed metabolic activity of the right labia majora suggestive of post-surgical inflammation versus residual disease as well as thyroiditis and indeterminate bilateral small pulmonary nodules. Small bilateral inguinal nodes were also reactive but with no other evidence of distant disease. The patient's pathology was discussed at an interdisciplinary tumor board and re-excision with right groin node dissection was recommended due to positive pathologic margins and depth of invasion greater than 1 mm. Unilateral lymph node dissection was recommended over bilateral dissection due to the overall reassuring appearance of her inguinal lymph nodes on PET/CT imaging and distance greater than 2 cm from the midline on gross examination. Chemotherapy was also considered but was not recommended at this time. The patient underwent a right hemivulvectomy with right inguinal lymph node resection three months after initial presentation. Pathology showed no residual disease at the primary site. Ten right inguinal lymph nodes were removed showing reactive changes but were all negative for malignancy. Her case was again discussed at an interdisciplinary tumor board, who recommended whole pelvic radiation and cisplatin sensitizing chemotherapy given her aggressive histology and limited literature showing successful disease-free intervals with surgery, sensitizing chemotherapy, and radiation. This was completed two months after her second surgery. Following chemotherapy and radiation, her surveillance plan included CT scans of the chest, abdomen, and pelvis every 3 months for 1 year followed by yearly CT scans. She has been followed regularly since completion of her chemoradiation; her only residual symptoms have been arthralgias and dyspareunia, which improved with vaginal dilators and pelvic floor physical therapy. Her most recent imaging showed no evidence of recurrent disease and stable small bilateral pulmonary nodules.

3. Discussion

Cutaneous myoepithelial carcinoma is a rare neoplasm arising from myoepithelial cells located in the salivary glands, breast, respiratory tract, skin and other tissues. There have been very few reported cases of primary myoepithelial carcinoma arising from the vulva. The diagnosis of this pathology is often difficult to achieve due to their painless nature and visual heterogeneity, making it easy to confuse with other cutaneous tumors. Myoepithelial carcinoma at most sites often presents with slow growth over several months with an average size of 2.2 cm but do possess metastatic potential and display unpredictable biologic behavior (Frost et al., 2014). One review found a 1:2 male to female ratio of myoepithelial carcinoma, limited by a small sample size (Frost et al., 2014). The same study found a mean age of 43 years with a bimodal distribution of tumors occurring either before the age of 21 or after the age of 60. The peak incidence of these tumors was in the third to fifth decade of life (Hornick and Fletcher, 2003). Myoepithelial carcinomas

Table 1
Cases of Primary Vulvar Myoepithelial Carcinoma Including Characteristics and Treatment Outcomes.

Authors	Age (years)	Size (mm)	FIGO stage (pTNM)	Histology	Immunohistochemistry	Treatment/Outcome
Heinz P *et al*(5)	81	40	NA	Solid, microcystic with plasmacytoid appearance	+ for Cytokeratin, Vimentin, S-100, SMA − for EMA, Desmin, CD34, myoglobin	Bilateral Vulvectomy; Recurrence on abdominal wall at 18 months
Fukunaga M.(4)	52	30 × 25 × 20	II (pT2N0M0	Epithelioid, Trabecular, cord, solid or reticular arrangement with hyalinized stroma	+ for EMA, Vimentin, SMA, GFAP, Keratin, S-100 − for CAM5.2, Calponin	Wide local excision + TAH/BSO + PLND + BILND; Disease not evident at 6 months
Noronha V *et al* (8)	37	35 × 31 × 45	IIIb (pT2N1M0)	Sheeted arrangement of undifferentiated rhabdoid cells with myxoid stroma	+ for CAM5.2, CD68, CEA, CD34, AE1/3, Calponin, S-100 − for Desmin, Myogenin, SMA, Vimentin	Right majora vulvectomy + BILND with adjuvant concurrent chemoradiation with cisplatin followed by chemotherapy; Disease not evident at 42 months
Meenakshi and McCluggage WG(7)	40	30	NA	Sheeted arrangement of tumor cells with moderately pleomorphic spindle-shaped nuclei	+ for AE1/3, CAM5.2, EMA, Desmin, Calponin, SMA − for p63, S-100, GFAP	Wide local excision; Disease not evident at 36 months
Meenakshi and McCluggage WG(7)	44	25	NA	Sheeted, nested arrangement of epithelioid tumor cells with severe nuclear pleomorphism	+ for EMA, SMA, AE1/3, Calponin − for p63, S-100, GFAP, CD34	Wide local excision; NA
Miyata M. *et al* (2)	49	20 × 30	IVa (pT2N2M0)	Solid, nested and trabecular arrangement	+ for CAM5.2, CEA, EMA, p63, S-100, Vimentin − for SMA	Wide local excision + BILND with adjuvant radiotherapy; Disease not evident at 56 months
Khazeni K. *et al* (6)	33	260 × 75 × 105	IIIa (pT1N1M0)	Intermediate-sized polyhedral cells with myxoid to hyalinized stroma	+ for EMA, SMA, − for S-100, CD34, SOX10, p63, GFAP	Radical vulvectomy with adjuvant chemoradiation; Deceased at 9 months
Castelow C. *et al.*	40	55 × 45 × 35	Ib (pT1N0M0)	Fascicular, nested, and lobulated growth. Predominantly spindled cells with ample eosinophilic cytoplasm. Scattered multinucleate cells.	+ for EMA, Calponin, DOG1 − for INI1, CD34, keratin, SOX10, HMB45, S-100, MITF, desmin, myogenin, p63, ERG, CK 8/18, CD117, CD99, TLE1, OSCAR, AE1/3, 34BE12	Right radical Vulvectomy with right lymph node dissection, adjuvant chemoradiation. Disease not evidence at 12 months.

+, positive; −, negative; AE, anion exchange; BILND, bilateral inguinal lymph node dissection; BSO, bilateral salpingo-oophorectomy; CAM, cellular adhesion molecule; CEA, carcinoembryonic antigen; EMA, epithelial membrane antigen; FIGO, International Federation of Obstetrics and Gynecology; GFAP, glial acidic fibrillary protein; SMA, smooth muscle actin; NA, not available; PLND, pelvic lymph node dissection; TAH, total abdominal hysterectomy.

are typically well circumscribed, non-encapsulated tumors without dermal connections on histology (Frost et al., 2014). Microscopic features typically show a reticular growth pattern with intersecting chords of epithelial, ovoid, or spindled cells separated by chondromyxoid or collagenous stroma that vary in prominence. Some tumors also have a mix of both epithelioid and spindled cells. Our patient's pathology was consistent with this morphology, showing a densely cellular spindle cell neoplasm with fascicular and lobulated growth. The cells were predominantly spindled with ample eosinophilic cytoplasm, with a minor component of more epithelioid to rhabdoid morphology. Scattered multinucleated cells, frequent mitotic activity and multifocal necrosis were also identified. Immunohistochemistry staining is important for making the diagnosis because some of the morphologic features can be confused with other more common tumors (Miyata et al., 2011). In Hornick and Fletcher's review, all tumors were positive for cytokeratin and/or epithelial membrane antigen (EMA). 93 % were positive for keratin, usually AE1/AE3, PAN-K and/or CAM5.2. EMA was seen in over half the tumors focally in spindled or epithelioid myoepithelial calls. The myogenic markers calponin, SMA, and desmin were also seen in several tumors with calponin being the most sensitive, present in 87 % of tumors. The myoepithelial marker p63 was seen in 23 % of tumors. Other important IHC markers are S-100 and GFAP seen in 87 % and 46 % of tumors respectively (Miyata et al., 2011). The most significant histologic characteristic that has been correlated with recurrence or distant metastasis is moderate to severe cytologic atypia (Hornick and Fletcher, 2003). Our patient's pathology showed staining mostly consistent with these findings. Staining was positive for EMA, calponin, and smooth muscle actin; however, staining was interestingly negative for desmin, myogen, S100, p63 and keratins including AE1/AE3 and PAN-K. Myoepithelial carcinomas typically follow a lymphatic drainage pattern of metastasis. A retrospective review of 51 patients with salivary myoepithelial carcinoma found regional lymph node metastasis in 7 out of 17 patients in which lymph node dissection was performed (41 %). Additionally, 4 of the patients with regional metastasis had recurrence and 3 had distant metastasis to sites including thigh musculature, brain and vertebral bones (Kane and Bagwan, 2010).

Most data regarding myoepithelial carcinoma have been gathered from reports in sites other than the vulva, making information regarding vulvar primary myoepithelial carcinoma scarce. Including this case, we have compiled 8 cases of primary myoepithelial carcinoma (Table 1) and will now review these cases. In 1999, Heniz et al reported the first case of primary myoepithelial carcinoma in the English literature in an 81-year-old female with a 40 mm tumor in the left labia majora (Hinze et al., 1999). The patient underwent surgical resection and one week later had a bilateral vulvectomy. Gross examination of the tumor showed a yellow-tan glistening mass with focal hemorrhages. The tumor showed multinodular growth pattern histologically and was unencapsulated with a destructive and infiltrative growth pattern. Appearance of the neoplastic cells were myoid, spindled, epithelioid, and/or plasmacytoid. The cells were also immunoreactive for cytokeratin and vimentin. Other positive stains in some cells included S100 protein antibody and alpha-smooth muscle actin antibody. Cells were negative for epithelial membrane antigen, desmin, CD34 and myoglobin. The patient also had resection of abdominal metastasis 18 months after surgery (Hinze et al., 1999).

Fukunaga reported a case of a 3 × 2.5 cm tumor on the right labia majora in a 52-year-old woman than had been enlarging slowly (Fukunaga, 2003). Examination showed a small, well defined, non-fixed and non-tender rubbery mass not involving the overlying skin and without palpable lymphadenopathy. The patient underwent wide excision of the vulva with total abdominal hysterectomy, bilateral salpinogo-oophorectomy, and pelvic and inguinal lymph node dissection. Gross examination of the tumor showed a well demarcated, nonencapsulated solid white rubbery tumor without hemorrhage or necrosis. Microscopically it was unencapsulated and showed a multinodular growth pattern. Appearance of the neoplastic cells were epithelioid, trabecular, cord, solid, or reticular arrangements with hyalinized stroma. Moderate cellular atypia was present with 4 mitotic figures per high powered field. The cells were strongly immunoreactive for vimentin, epithelial membrane antigen, alpha smooth muscle actin, and glial acid fibrillary protein. Some cells were also weakly immunoreactive for keratin and S100p protein. Cells were negative for cytokeratins 7, 8, 18, & 20, desmin, muscle-specific actin, CD10, CD31, CD34, HMB45, and calponin (Fukunaga, 2003).

A case presented in 2006 by Noronha et al included a 37-year-old woman with a 9-month history of a mass in the right labia majora. After an incisional biopsy showing poorly differentiated carcinoma, the patient underwent MRI that revealed a 3.5 × 3.1 × 4.5 cm mass in the right vulva and two enlarged lymph nodes in the right groin. Excision of the right labia and bilateral inguinal lymph nodes was performed. On histologic examination, the right inguinal lymph nodes had sheets of high-grade malignant cells and myxoid stroma. Tumor cells had a rhomboid appearance without glandular configurations. The cells were strongly immunoreactive to cellular adhesion molecule 5.2 and CD68. They were focally positive for CD34 and carcinoembryonic antigen. Only a few cells were positive for anion exchange protein 1/3, calponin, and S100 protein. Cells were negative for alpha smooth muscle actin, desmin, myogenin, myogenic differentiation 1, vimentin, keratin type II cytoskeletal 7, keratin type 1 cytoskeletal 20, and major glycoprotein GP100. The patient also received radiation therapy to the pelvis and bilateral groins. CT scan 1 month after completing of therapy showed a 0.6 cm left lung nodule. A repeat CT 2 months later showed stability of that nodule, but also a new 1.2 cm nodule in the left lung base that showed metastatic myoepithelial carcinoma on fine needle aspiration. Because the authors were not able to fine recommendations on how to treat metastatic myoepithelial carcinoma, they chose carboplatin, paclitaxel, and gemcitabine given their use in carcinoma of unknown primary origin. Gemcitabine was discontinued due to neutropenia, but the patient received three cycles of carboplatin and paclitaxel. Repeat CT showed complete resolution of the left lung base nodule and stability of the other nodule. The patient received a wedge resection of the stable nodule that showed adenofibroma and the patient remained disease free 42 months after diagnosis.

A case series by Meenakshi and McCluggage in 2009 discussing four neoplasms of the vulva and vagina included two more cases of vulvar myoepithelial carcinoma (Meenakshi and McCluggage, 2009). The first patient was a 40-year-old woman who presented with a 3 cm nodule in the right labia majora that underwent wide excision with no evidence of recurrence or metastasis 3 years later. On gross examination, the tumor was a solid, well circumscribed, and white colored. Histology showed the lesion within the deep dermis and subcutaneous tissue. It was unencapsulated and showed marginal irregularities. The tumor cells had spindle shaped nuclei and showed moderate atypia with 4 mitotic cells per high powered field. There were also focal areas of stromal hyalinization and myxoid change with no necrosis or vascular invasion. Immunohistochemistry was diffusively positive for cytokeratin anion exchange protein 1/3, cellular adhesion molecule 5.2, and estrogen receptor. It was also focally positive for epithelial membrane antigen, Desmin, alpha smooth muscle actin, CD34, and calponin. It was negative for S100, p63, and glial fibrillary acid protein (Meenakshi and McCluggage, 2009).

The second patient was a 44-year-old woman that presented with a 2.5 cm nodule in the right vulvar region that underwent surgical excision and with no evidence of a primary tumor in another location after extensive investigation (Meenakshi and McCluggage, 2009). Histology showed a well circumscribed but unencapsulated lesion with epithelioid tumor cells containing eosinophilic cytoplasm. Tumor cells were arranged in a sheeted pattern with focal areas of nested arrangement. Atypia was present with 20 mitotic figures per 10 high powered fields. Again, there was stromal hyalinization and myxoid change with no necrosis or vascular invasion. Immunohistochemistry was diffusely positive for epithelioid membrane antigen, alpha smooth muscle actin,

calponin, and estrogen receptor. It was focally positive for cytokeratin AE1/3 and was negative for cellular adhesion molecule 5.2, desmin, S100, p63, glial fibrillary acid protein, and CD34 (Meenakshi and McCluggage, 2009).

The next case came in 2011 reported by Miyata et al and was a 49-year-old woman who presented with a 20 × 30 mm rubbery, mobile, irregular tumor on the right labia majora that had been slowly growing for the past two years with bilaterally enlarged inguinal lymph nodes (Miyata et al., 2011). She was seen by the authors after undergoing left inguinal lymph node biopsy by another physician that showed poorly differentiated carcinoma. The patient underwent CT scans that showed the 20 × 30 mm tumor of the right labia majora as well as right inguinal lymph node enlargement. Ga scintigraphy revealed anormal uptake in the inguinal region and area of the tumor. Wide local excision of the right labia majora and bilateral dissection of the inguinal lymph nodes was performed. Gross examination revealed a dark tumor with central necrosis. Histology showed an unencapsulated tumor with an infiltrating growth pattern with areas of nested cells and hyalinized stroma with myxoid changes. Appearance of neoplastic cells were epithelioid or trabecular. Atypia was present with over 20 mitotic figures her 10 high-powered fields. Immunohistochemistry of tumor cells was diffusely positive for cellular adhesion molecule 5.2, epithelial membrane antigen, S100 protein, and vimentin. It was focally positive for carcinoembryonic antigen and p63 and negative for alpha smooth actin antigen. The patient also received adjuvant radiation therapy with 50 Gy to the pelvis and groin and had no evidence of disease at 56 months (Miyata et al., 2011).

The most recent case was reported by Khazeni et al and discussed a 33-year-old female that presented with pelvic pain (Khazeni et al., 2018). A CT scan of the abdomen and pelvis reveled a 3.6 × 3.1 cm right inguinal mass and the patient underwent an excision and biopsy. Histology showed intermediate-sized polyhedral cells with eosinophilic cytoplasm and hyalinized to myxoid stroma. Immunohistochemistry stained diffusely positive for epithelial membrane protein and alpha smooth muscle actin. Tumor cells were negative for S100, CD34, SOX10, p63, and glial fibrillary acid protein. The patient was later seen due to pain and swelling at the operative site and CT showed growth of a right labial mass with extension into the subcutaneous tissue and right rectus musculature as well as an enlarged 1.5 cm right inguinal lymph node. The patient received one round of carboplatin and paclitaxel but continued to have tumor progression. She subsequently underwent resection of a 26 cm right groin mass with radical vulvectomy and right superficial inguinal ly-mphadenectomy-two months after initial surgery. Histologic examination showed high grade myoepithelial carcinoma with necrosis and hemorrhage with venous invasion but negative surgical margins. Two resected lymph nodes showed metastasis. Chemoradiation with cisplatin was began six weeks after surgery but the patient later had recurrence in bilateral groin, pelvic, and paraaortic lymph nodes. Radiation therapy was stopped and ifosfamide without doxorubicin was initiated. The patient also underwent whole genome sequencing that revealed a loss of SMARCB1. Although the patient initially responded well to ifosfamide, the patient had recurrence of pain with abdominal distention likely due to abdominal metastasis. The patient expired 9 months after symptom onset (Khazeni et al., 2018).

The above reports demonstrate all cases of primary myoepithelial carcinoma in the literature. Each case was treated with surgical resection. Two patients were given chemoradiation therapy and one was given adjuvant radiotherapy without chemotherapy. Three of these patients had either recurrence or metastasis of their cancer. These cases suggest that while slow growing, myoepithelial carcinoma of the vulva can be aggressive with unpredictable biologic behavior. The most likely route of metastasis through lymphatic spread. Additionally, histologic examination and immunohistochemistry staining are essential for the diagnosis.

There is no current accepted standard treatment for myoepithelial carcinomas. Recommended initial treatment for any cutaneous myoepithelial carcinoma is typically wide excision with free margins (Frost et al., 2014). Based on the reported cases of primary myoepithelial carcinoma, surgical excision with clear tumor margins also seems to be the best initial treatment for these tumors. Several different surgical techniques have been used including radical vulvectomy, bilateral vulvectomy, unilateral vulvectomy, and wide local excision. There is insufficient evidence to recommend one surgical approach as superior. Given the rate of metastasis of these tumors through lymphatic spread, there may be benefit in preoperative imagining as well as sentinel lymph node biopsy to assess tumor spread. There have been inconsistent reports of successful treatment with chemotherapy. Of the two cases of primary vulvar myoepithelial carcinoma in which chemotherapy was used, one patient remained disease free at 42 months. This patient was also treated with right labia majora vulvectomy and bilateral inguinal lymph node dissection with chemoradiation, which could confound the role chemotherapy played in eradication of the cancer (Noronha et al., 2006). However, chemotherapy was only initiated in this patient after discovering lung metastasis and resulted in complete resolution of the lung nodule. The authors used a chemotherapy regimen including carboplatin, paclitaxel, and gemcitabine as this has been shown to be effective in carcinoma of unknown origin. Although gemcitabine was discontinued early due to side effects, this may be a reasonable option as adjuvant therapy. There has also been a report of successful treatment of primary myoepithelial carcinoma found to have a BRAF V600E mutation with vemurafenib-cobimetinib (BRAF and MEK inhibitors), suggesting a possible role of targeted therapy and broad molecular analysis in these tumors (Guidry et al., 2020).Cases of primary myoepithelial carcinoma are scarce and data to support a standard treatment is not widely available. As this tumor can often be mistaken for other more common malignancies, it may be more common than previously realized. It is therefore important to identify these cases and their responses to treatment to improve the prognosis for this potentially devastating diagnosis.

4. Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

CRediT authorship contribution statement

Christopher Castelow: Investigation, Writing – original draft, Project administration. **Elliott Carter:** Conceptualization, Methodology, Investigation, Writing – original draft, Supervision. **Stephen DePasquale:** Conceptualization, Methodology, Validation, Writing – review & editing, Supervision.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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