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Original contribution

Myoepithelial neoplasms involving the vulva and vagina: report of 4 cases

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Summary We report 4 myoepitheial neoplasms involving the vulva (2 cases) or vagina (2 cases) in patients aged 40 to 45. Two tumors were composed entirely of ovoid or spindle-shaped cells, one entirely of epithelioid cells, and in the other, there was a mixture of spindled and epithelioid cells. Small foci of ductal differentiation with squamous metaplasia were present in one case and a minor stromal component, which varied from myxoid to hyalinized, in all cases. In all cases, the tumor cells were positive for epithelial markers (cytokeratins and/or epithelial membrane antigen) and the myoid markers α smooth muscle actin and calponin. Desmin was positive in 3 cases. S100 and p63 were positive in 1 of the 4 neoplasms. On the basis of the nuclear features and degree of mitotic activity, 2 neoplasms were classified as benign myoepitheliomas and 2 as myoepithelial carcinomas. Judging by the paucity of cases in the literature, myoepithelial neoplasms appear extremely rare in the vulvovaginal region with only 3 previous case reports of primary vulval tumors. As far as we are aware, this is the first description of a primary vaginal myoepithelial neoplasm. At these sites, myoepithelial tumors are liable to be misdiagnosed as a variety of other neoplasms because the pathologist may not think of the diagnosis. In reporting these cases, we discuss the criteria for diagnosis and the differential diagnosis.

1. Introduction

Neoplasms with a component of myoepithelial cells most commonly occur within the salivary glands. The most common salivary neoplasm to contain myoepithelial cells is the pleomorphic adenoma that contains, in addition to the myoepithelial component, areas of ductal differentiation. Pure myoepithelial neoplasms are more uncommon within the salivary glands and may be benign (myoepithelioma) or malignant (myoepithelial carcinoma) [1,2]. In recent years, the occurrence of myoepithelial neoplasms in cutaneous and soft tissue sites has been highlighted [3,4]. In this report, we describe 4 myoepithelial neoplasms involving the vulva (2 cases) or vagina (2 cases). There have been only occasional previous reports of a pure myoepithelial neoplasm involving the vulva [5-7], and given the wide array of mesenchymal neoplasms that potentially occur in the vulvovaginal region, pathologists may not consider the diagnosis. In reporting these cases, we discuss the criteria for diagnosis together with the differential diagnosis and stress that when dealing with a mesenchymal neoplasm in the vulvovaginal region, pathologists should consider a wide range of entities and not just those mesenchymal lesions characteristic to this site.

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2. Materials and methods

The cases were derived from the consultation files of one of the authors (WGM). Clinical information was obtained from the letters sent by the referring pathologists and communication with these pathologists. Hematoxylin and eosin–stained slides were examined (the number of slides was 1 in cases 1 and 2, 8 in case 3, and 2 in case 4). Immunohistochemical staining for AE1/3, CAM5.2, epithelial membrane antigen (EMA), desmin, α smooth muscle actin (αSMA), calponin, S100, p63, glial fibrillary acidic protein (GFAP), CD34, and estrogen receptor (ER) was performed during the workup of the cases (not all stains were performed on each case—see Results and Table 1). Immunoreactivity was classified as negative (N), focally positive (F) (<50% cells positive), or diffusely positive (D) (50% or more cells positive).

3. Results

3.1. Clinical details

The patient's ages in cases 1 to 4 were 45, 41, 40, and 44 years, respectively. In case 1, the patient presented with stress incontinence. A 1.5-cm vaginal nodule was identified and removed. The patient in case 2 presented with a 2-cm nontender vaginal nodule that was removed. In case 3, the patient presented with a 3-cm lump on the right labium minus that was removed. Three months after the original resection, the patient in case 3 underwent wide excision of the same area. No residual tumor was present. Three years after this, there is no evidence of tumor recurrence or metastasis. The patient in case 4 presented with a 2.5-cm lump on the right side of the vulva that was removed. After this, the patient was extensively investigated to look for a primary tumor elsewhere, but none was found. We have no follow-up in cases 1 or 2.

Table 1 Immunohistochemistry results

Antibody	Case 1	Case 2	Case 3	Case 4
AE1/3	D	D	D	F
CAM 5.2	ND	D	D	N
EMA	N	ND	F	D
Desmin	F	D	F	N
α SMA	F	F	F	D
Calponin	F	F	F	D
S100	N	F	N	N
p63	F	N	N	N
GFAP	ND	ND	N	N
CD34	ND	D	F	N
ER	D	ND	D	D

Abbreviations: ND indicates not done; N, negative; F, focally positive; D, diffusely positive.

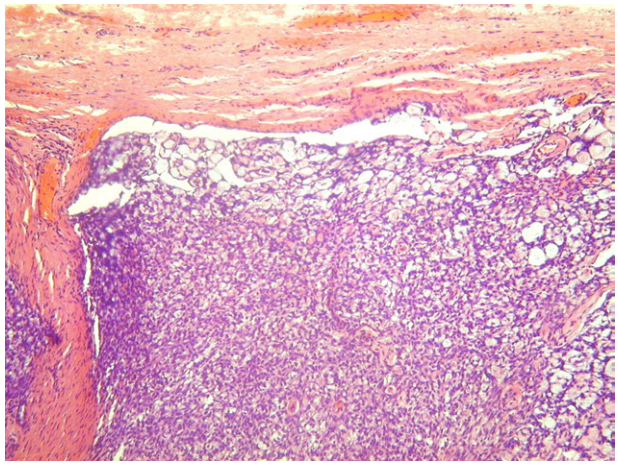

Fig. 1 Case 1 comprising well-circumscribed lesion composed of cells with ovoid to spindle shaped nuclei and scanty cytoplasm.

3.2. Pathologic findings

In case 1, the surgical specimen consisted of a well-circumscribed lesion measuring 1.5 cm in maximum dimension with a solid cream-colored cut surface. The specimen in case 2 comprised a 2-cm well-circumscribed solid gray-colored lesion with overlying vaginal mucosa. In case 3, the specimen consisted of a well-circumscribed solid white-colored lesion measuring 3 cm in maximum dimension with an overlying skin ellipse. In case 4, the specimen consisted of a 2.5-cm well-circumscribed solid tan-colored lesion.

Histologic examination of case 1 showed a well-circumscribed but unencapsulated lesion with a relatively uniform appearance throughout. For the most part, the lesion was densely cellular and composed of cells with ovoid to spindle-shaped nuclei and a scanty amount of eosinophilic cytoplasm (Fig. 1). There was minimal nuclear pleomorphism with evenly dispersed chromatin and an

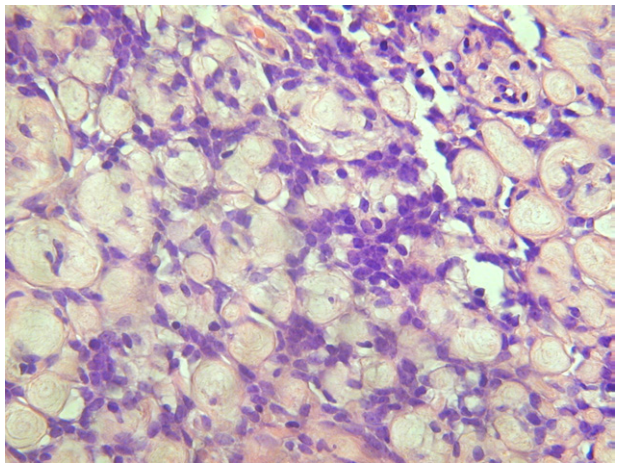

Fig. 2 In areas in case 1, whorls of collagen are surrounded by tumor cells.