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Malignant Adenomyoepithelioma of the Vulva: One Case Report and Literatures Review

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Introduction

Adenomyoepithelioma (AME) is uncommon characterized by biphasic proliferation of both epithelial and myoepithelial cells. More than half of the adenomyoepitheliomas (AME) are benign, but in rare instances, it may become malignant. Malignant adenomyoepitheliomas almost always occur in the breast and salivary gland. They are unusual in vulva. We report this case and provide some information about symptoms, microstructure, treatment，and a discussion about this kind of disease.

Case Report

A 35-year-old female was admitted to the hospital on January 11, 2007 due to a neoplasm of the vulva for three years. The patient felt a neoplasm of the vulva three years ago. It was the size of a grain of rice and there was no flaring of the mucosa or skin. The patient felt neither pain nor itching and did not pay any further attention on the discovery. One week before she was admitted to the hospital, she felt discomfort but no pain or itch. On examination, we found a 3 cm × 2 cm × 2 cm neoplasm, located in the underside of the labium majus, near the vaginal orifice. The surface of the neoplasm was smooth without granulation or flaring. The patient underwent a simple neoplasm excision. The pathology was diagnosed as: “malignant adenomyoepithelioma of the vulva, and there are a few cancer cells in vascular space. IHC: SMA (++), AE1/AE3 (++), S-100 (-), the positive percentage of ki-67 is about 5%. CD34 of Few cell is positive”. One week later, the patient underwent a radical excision and inguinal lymphadenectomy. She recovered well and discharged five days after the surgery and has not reccured within one year.

Discussion

Adenomyoepithelioma (AME) is uncommon characterized by biphasic proliferation of both epithelial and myoepithelial cells. More than half of the adenomyoepitheliomas are benign, but in rare instances, the epithelial, the myoepithelial or both components of an AME may become malignant. There have only been case reports of malignant adenomyoepitheliomas of the breast and salivary gland, but no reports of malignant adeno myoepithelioma of the vulva.

Myoepithelial cells are clear to spindle and occur in nests or sheets, occasionally pushing aside or engulfing the epithelial component. Apocrine change can be seen frequently and occasional cases contain associated hyaline basement membrane-like material. Occasionally, calcifications can be seen (Fig.1) Myoepithelial neoplasms of skin and soft tissues comprise cutaneous mixed tumor (chondroid syringoma), mixed tumor of subcutaneous and deep soft tissues, myoepithelioma and rare malignant myoepithelioma. Myoepithelial tumors of the skin and soft tissues are characterized by an extreme clinicopathological heterogeneity, just as they are in other anatomical locations. The neoplasms arise in children as well as in adults and are composed of epithelioid, histiocytoid, spindled, plasmocytoid and/or clear tumor cells in varying combinations, and are set in a myxoid or hyalinised intercellular matrix[1].

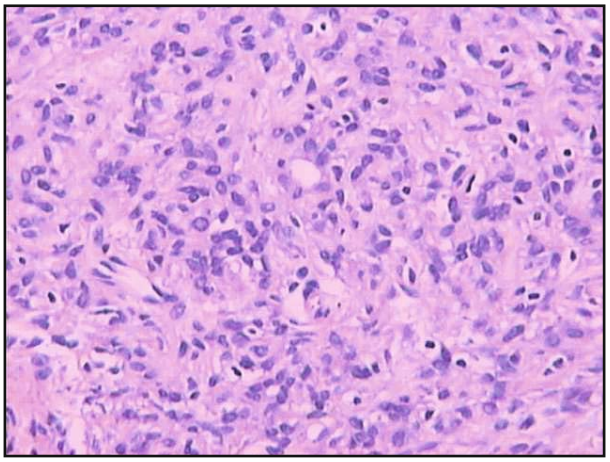

Fig.1. Myoepithelial cells are occurred in nests and Apocrine change can be seen (H&E stain, × 400).

Immunohistochemically, neoplastic cells stain positively for epithelial markers (pancytokeratin and/or epithelial membrane antigen), and often for S-100 protein. Epithelial elements stained with cytokeratins, while the myoepithelial elements often stained weakly with cytokeratin AE1/3 and variably with muscle markers including smooth muscle actin, muscle specific actin, vimentin, calponin, S-100, cytokeratin 14, and p63[2].

The nuclear staining of p63 is usually intense and consistent, staining most of the myoepithelial cells in most sections, even in the oldest archived paraffin tissue. p63, which recently characterized p53 homologue, is consistently expressed in myoepithelial cells of the human breast; however, no assessment of its immunoreactivity in vulvar tissues has been reported so far in vulva tissues. Some research indicated that some factors may lead to malignant AME (e.g.), mutation of the p53 gene (T→G transversion at codon 270)[3]. p53 is consistently negative in benign lesions and is overexpressed in malignant myoepithelioma. The synergy of EGFR and TGFR beta have been shown to have a significant relationship to the proliferative and differentiation processes of myoepithelial tumors. The activated c-erbB-2 oncogene which is similar with EGFR in structure, plays an important promoting role in unrestricted progression of malignant myoepithelioma[4].

We can see some cases like adenomyoepithelioma of breast and salivary glands, but rarely of the vulva. Adenomyoepithelial tumours comprise a spectrum of neoplasms consisting of an admixture of glandular and myoepithelial differentiation patterns. Smooth Muscle Actin positive (SMA-positive) cells co-expressing cytokeratins could be identified as a key component[5]. Kapoor and Chinoy[6] reported two cases of pure malignant myoepithelioma of the breast, utilizing light microscopy and immunohistochemical methods for diagnosis. Both cases were manifested as breast lumps. Hematoxylin and Eosin (H&E) stained microscopic sections predominantly revealed spindle cell tumor morphology. An immunohistochemical work up was done. Case number one expressed positivity for vimentin, Smooth Muscle Actin (SMA), S-100 and CD10. Case number two tested positive for Vimentin, CD10 and p63[6]. This led to the diagnoses of malignant myoepithelioma.

About 90% of primary vulvar malignancies are squamous cell carcinomas. Less common tumors include melanomas, adenocarcinomas (often underlying Paget' s disease), basal cell carcinoma, Bartholin's gland carcinomas, sarcomas, verrucous carcinomas, and lymphomas. The relative proportion of vulvar malignancies is seldom stated in papers, especially malignant adenomyoepithelioma.

Surgery is the mainstay of treatment for malignant adenomyoepithelioma of the vulva. Both physical and psychological morbidity was high with this radical surgery, and in recent years a more conservative surgical procedures have come to be advocated, particularly for the primary tumor. With careful patient selection, survival does not seem to be compromised. Adenomyoepithelioma can metastasize to any site. Nadelman described 2 cases of metastasis of histologically "benign" adenomyoepitheliomas of the breast from the lung[7].

The prognosis for malignant adenomyoepithelioma is better with radical excision. Malignant tumors of the vulva should be treated by radical excision and subsequent inguinal lymphadenectomy. Chemotherapy and radiation therapy as adjuvant or complementary therapies may then further improve the prognosis.

Conclusion

Alhough malignant adenomyoepithelioma of the vulva occurs rarely, we can use surgery, pathology and immunochemistry to diagnose it. Immunohistochemical index such as SMA, CD10, S-100 are used as reference with little specific. Further therapy can be given based on clinical stage and pathological report.

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