Bilateral Fibroepithelial Polyps of The Lower Female Genital Tract With Atypical Stromal Cells (Pseudosarcoma Botryoides): A Case Reports

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A case of female genital tract fibroepithelial polyps containing atypical stromal cells arising from the vulva is described. Fibroepithelial stromal polyps of the vulvar vaginal lesion are benign lesions that, when bland or hypocellular, are readily recognized. The tumor was multiple and the highest one measured 4 centimeter in diameter. Cytologic atypia has been a previously recognized feature in these lesions; however fibroepithelial polyps with stromal atypia are an uncommon tumor of the vulva, are rarely bilateral and combination of these features has only rarely been documented. These tumors must be differentiated from sarcoma botryoides, which resemble both grossly and microscopically.

Key Words: Fibroepithelial polyps, Atypical stromal cells, Female genital tract

Fibroepithelial stromal polyps are benign lesions that occur in a wide variety of sites including the lower female genital tract. Polypoid or grape-like masses of the vagina or cervix are always worrisome because they raise the suspicion of sarcoma botryoides. More recently, larger series showed that reported lesions in the vulva remain an atypical site for these diseases. Although they are rare at any age, about one third have been reported in pregnancy. They have a gross appearance of soft, translucent pedunculated mucosal lesions. These lesions could contain atypical stromal cells and the presence of such cells should not be mistaken for malignancy. Both are rare and it is important to distinguish between them.

We describe a vulvar example of the condition together with atypical stromal cells. The appearance of an identical tumor on the contralateral labia compounded the difficulty in both clinical and pathological diagnosis.

Case Report

A 18 year-old unmarried girl, first presented in 1994 with polypoid swelling of the right and left labium, which had been presented for 5 years. They were excised and no further treatment instituted. The pathological material submitted for assessment. She returned in 2005 with large polypoid lumps on both sides of the clitoris and lower portion of labium majus. The clinical examination revealed to be circumscribed tumor mass of the vulva.

At operation, the polypoid tumor was attached to the left and right labium majus by a broad base. The lower genital tract was otherwise normal and the inguinal nodes were not enlarged. None of the lesions showed locally aggressive behavior. The tumors were removed by local excision and submitted for pathological assessment.

Pathologic Findings

Macroscopic features: The lesions ranged in size from 1 to 4 cm in maximum dimension. The fresh specimen was described as polypoid or pedunculated in appearance which had either smooth or fissured surfaces. The cut surface was oedematous, uniform and soft. They were described as multiple cysts that were gelatinous in appearance (Figure 1).

Figure 1. Fibroepithelial sessile vulvar polyps with fissured surfaces.

Microscopic features: The histologic features revealed loose, acellular myxoid stroma, with irregular dilated blood vessels, covered by normal squamous epithelium. On high power we observed pleomorphic fibroblast-like cells, with hyperchromatic nuclei and elongated cytoplasmic processes and sparse multinucleated giant cells (Figure 2). The micro-
copic features of the right labial lesions were identical to those of the original left labial tumor.

Discussion

Pseudosarcoma botryoides of the female genital tract are benign lesions that occur generally in young to middle age women in their reproductive years. The term is not reserved for embryonal rhabdomyosarcoma, a highly malignant tumour with poor prognosis. The term “pseudosarcoma botryoides” was applied by Elliott et al. to emphasize its gross and microscopic resemblance to the malignant tumor and the fact that it has been confused with latter. They occur most commonly in the vagina—the site of their original description—but can also occur in the vulva and less commonly the cervix. Fibroepithelial stromal polyps can occasionally recur, sometimes more than once.

The pathogenesis of fibroepithelial polyps is poorly understood. These tumors are composed of an intimate admixture of myxoid stroma and vessels reminiscent of the submucosa of the normal uterine cervix, vagina and vulva. The occurrence of morphologically similar cells in normal tissue and reactive processes at other anatomic locations such as breast, bladder, nasal polyps and polyps and ulcers of the gastrointestinal tract add additional support to the likely reactive nature of pseudosarcoma botryoides of the lower female genital tract. The stromal cells of fibroepithelial polyps can express estrogen and progesterone receptors, which suggests that hormonal influences may potentially play a role in the pathogenesis of these lesions. Approximately one-third of the cases of fibroepithelial polyps occur in pregnant women and another third in those who have taken sex hormone preparations. It may be that such a hormonal milieu predisposes to their formation.

In summary, pseudosarcoma botryoides of the lower female genital tract can exhibit a spectrum of worrisome morphologic features, with polyps occurring during pregnancy being especially pseudosarcomatous. Cytologic atypia has been a recognized feature in these lesions. Patients with a combination of atypical features may have greater recurrent potential. The treatment of choice is local excision. Recurrence may occur if resection is incomplete. To ensure completeness of surgical removal, it is urged that pathologists pay special attention to excision margins.

References