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Case Report

Giant fibroepithelial polyp of vulva with fibroid uterus with primary lymphedema (Meige disease/lymphedema praecox): a rare case report

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ABSTRACT

Fibroepithelial polyps or FSPs develop in the reproductive years of young to middle-aged women. They primarily affect the vagina, and their occurrence in the vulva is less common. Giant FSP are very rare. We present a case of a patient with giant FSP of the vulva with unilateral lymphedema. A forty year old patient presented with history of mild, dull, aching, continuous pain in lower abdomen. After her first delivery, she had developed pruritis over vulva followed by appearance of a growth. A non-tender, mobile warty growth was found on the examination of vulva, and it involved the mons pubis, clitoris and left labia minora and major. Dermatological consultation made a preliminary diagnosis of nevoid growth vulva. Operative findings from exploratory laparotomy confirmed that uterus had enlarged to 28 weeks size. The diagnosis of fibroepithelial polyp was confirmed by histopathological examination. A common pathogenetic background of lymphedematous FSPs is persistent lymph stasis, along with consequent injury of microcirculation and stromal hyperplasia. The role of hormonal factors in the development of these lesions in also supported by the Estrogen and progesterone receptor positivity. The association of FSPs with unilateral lymphedema, as in our patient, is very rare. This case provides some evidence that chronic lymph stasis can lead to microcirculation injury which further lead to stromal hyperplasia.

Keywords: Fibroepithelial stromal polyps, Lymphedema, Stromal hyperplasia

INTRODUCTION

Fibroepithelial polyp/acrochondron/skin tag/fibro-epithelial stromal polyps (FSP) often occur in a variety of sites, which includes the lower female genital tract. Their histological appearance has a wide range, andthey follow a benign clinical course.[1] Their incidence is approximately 46% in general population butgiant FSP are very rare. Common sites are skin creases e.g. neck, groin, armpits & eyelids. They are usually solitary but occasionally multiple lesions can occur. The lesion is generally 1-2 cm in size, but in rare cases it can reach large sizes, even up to 15-20 cm. There is a tendency for local recurrence after incomplete excision.[2]

Fibroepithelial polyps often develop in young to middle-aged women in their reproductive years. They primarily affect the vagina, less commonly the vulva, and the cervix is rarely affected. Since patients suffering from FSP are sometimes pregnant, or have a history of tamoxifen therapy or hormonal usage, there is some evidence of hormonal association. Fibroepithelial polyps often present with varying histological appearances, and thus may cause diagnostic difficulties.[3] Microscopically, the stroma in typical cases is edematous with prominent, often dilated, vessels. Rarely the stroma is myxoid. The constituent cells are spindle orstellate shaped and contain pale, tapering cytoplasmic processes. Multinucleated cells are often present. When associated with the cytologic pleomorphism, high mitotic activity (more than

10 per 10 HPFs), and atypical mitotic figures that may be seen in these polyps, the potential for misdiagnosis as sarcoma is significant.[4] Uponimmunohistochemical stains for these polyps, diffuse staining of the subepithelial spindle cells for estrogen receptor (ER) and progesterone receptor (PR) as well as vimentin is found.[5]

CASE REPORT

A 40 years P2002 presented with history of pain in lower abdomen since last one year which was continuous, dull aching, mild in intensity, progressive, radiating to back without any relieving or aggravating factor with no any diurnal variation. The patient did not have any urinary, bowel, or menstrual complaint. She had pruritis over vulva followed by appearance of a growth after her first delivery. She had noticed hypertrophy of her left half of body since childhood, and it was non-tender and gradually progressive without any impairment of her activity. She was average built with unremarkable general physical examination except left half of body below neck was hypertrophied as in comparison to right half (Figure 3). Per abdominal examination founda mass arising from pelvis around 28 weeks size, firm, irregular, non-tender, mobile from side to side, lower limit not reached and no free fluid. Examination of vulva revealed large warty growth involving the mons pubis, clitoris and left labia minora and major. The growth was non tender, mobile with multiple watery lesions on surface, and did not bleed on touch.

Another lesion involving left labia majora around 2.2 cm, multiple similar small lesions were seen around anal opening measuring 1.1 to 3.3 cm (Figure 1). Per vaginal examination showed cervix was high up behind the pubis symphysis. Uterus was enlarged to 28 week size, firm irregular, non-tender, mobile from side to side, and seemed continuous with cervix. Dermatological consultation was sought, and a preliminary diagnosis of nevoid growth vulva was made. Routine investigation was normal; serology for HIV and HbsAg was negative; CA-125 testreturned a value of 11.5 U/m; HPV DNA negative, ECG and ECHO were normal. Ultrasound showeda large heterogeneous echotexture abdomino-pelvic mass, a possibility of subserous/broad ligament fibroid.

CECT scan of abdomen and pelvis showed a large lobulated soft tissue exophytic growth seen in perineum arising from vulva with component in the vagina with small foci of calcification and increased vascularity, a sign of giant condyloma. A large subserosal leiomyoma arising from uterine fundus with abutment to anterior abdominal wall was noticed, with heterogenous enhancement with calcification and necrosis. Possibility of sarcomatous changes could not be ruled out. Patient underwent total abdominal hysterectomy with unilateral salpingo-oophorectomy along with wide excision of vulval growth in a single sitting. Operative findings confirmed that uterus had enlarged to 28weeks size. It was firm in consistency, had bosselated surface, and left tube and ovary was adherent to omentum and gut. Right ovary and tube were grossly normal. Cut section view revealed large fundal fibroid measuring 15.15cm, with 2-3 small fibroids around 2.2cm, no evidence of degeneration. Endometrium was normal, excision of vulval growth with wide margin (Figure 2) measuring 10.10 cm with papillary projection on surface another growth on left labia majora and perianal region were also excised. All specimens were sent for HPE. She received one unit of blood intraoperative. Her postoperative period was uneventful. She was discharged on 10th day and advised regular follow-up. Histopathological examination confirmed diagnosis of fibroepithelial polyp (Figure 4).

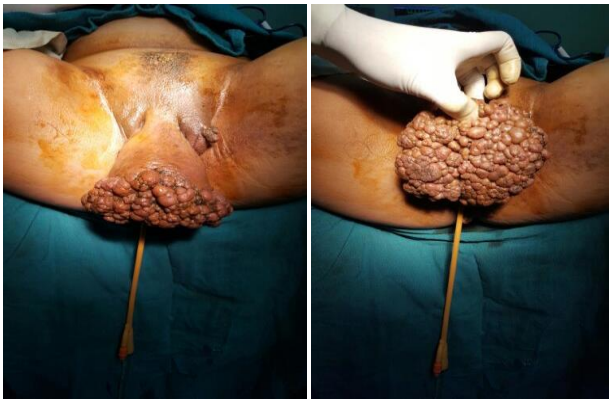

Figure 1: Fibroepithelial polyp of vulva.

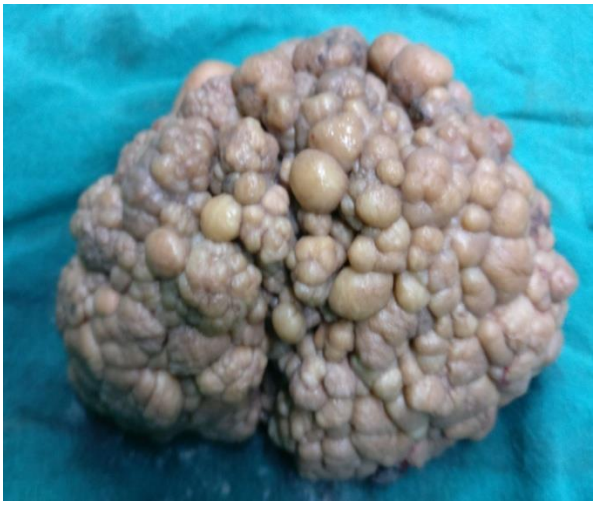

Figure 2: Vulval growth after excision.

DISCUSSION

In 15% reported cases of FSP lesions, they occur during pregnancy and they exhibit greater cellular pleomorphism and atypia. Additionally, they show exhibit multiple occurance in pregnant women. FSP can recur, occasionally, and after delivery spontaneous regression can occur. Hormonal influences probably factor in pathogenesis of these lesions, since the stromal cells of FSP can express estrogen and progesterone receptors.[1] After incomplete excision the stromal polyps have the

potential for local, non-destructive recurrence. FSPs contain a fibrovascular core and the stromal cellularity of the polyp can be variable. The hypocellular variant is composed of spindle cells set within a finely collagenous myxoid-like stroma. In the hypercellular form, the cells exhibit pleomorphism with mitotic figures.[2] The presence of stellate and multinucleated cells accumulating beneath the surface is not a sign of malignancy, and it has its own diagnostic importance. The stromal cells are vimentin and desmin positive, and can be actin positive.

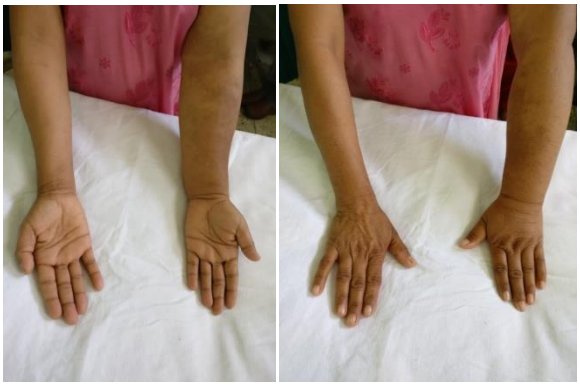

Figure 3: Lymphedema; left side.

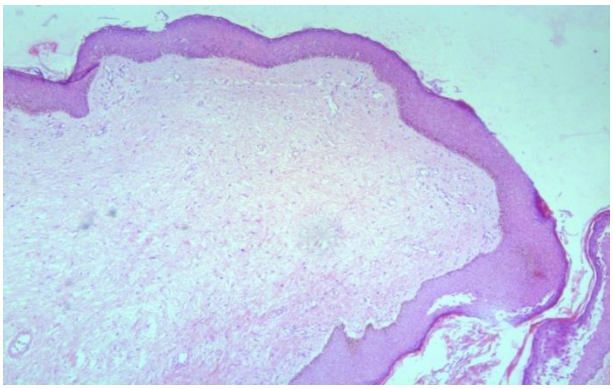

Figure 4: Histopathological slide.

The differential diagnosis of female genital tract tumours includes aggressive angiomyxoma, angiomyofibro-blastoma, cellular angiofibroma, superficial angiomyxoma, leiomyoma, leiomyosarcoma, squamous cell carcinoma, Paget's disease, and malignant melanoma.[3] Our case is most probably due to the persistent lymph stasis with a consequent injury of microcirculation and stromal hyperplasia, which is a common pathogenetic background of lymphedematous fibroepithelial stromal polyp.[2] Also, congenital ipsilateral lymphedema is thought to be the predisposing factor to the development of lymph stasis. The additional role of hormonal factors is suggested by the presentation of the primary lesion in young adulthood. Estrogen and progesterone receptor positivity of the mesenchymal cells further supports the importance of hormonal influence in the development of such lesions.

Their association with unilateral lymphedema, as in our patient, is very rare and only very few cases has been reported in history. Whether it is a coincidence finding or some association between these is not clear. But one hypothesis is proposed that chronic lymph stasis can lead to microcirculation injury which further leads to stromal hyperplasia.

CONCLUSION

It can be concluded from current study that chronic lymph stasis can lead to microcirculation injury which further leads to stromal hyperplasia.

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