Rare Vulvar Lesions: A Case Series

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ABSTRACT

Mass of the vulva include both benign and malignant lesions. Benign mass lesions of the vulva include tumors, hamartomas, cysts, infectious disorders, and non-neoplastic epithelial disorders. Some are rare mass lesions. Here, we presented three rare mass lesions of the vulva. The first case presented with the complaint of vulvar lesion since childhood, while the two cases presented in reproductive age and perimenopausal age. All three cases presented as mass lesions in the vulva with pain or uneasiness during work. In all these cases, excision was done. On histopathological examination, the lesions have different diagnoses which are common in other parts of the body, but rarely present at the vulva. A definitive diagnosis of a vulvar mass lesion is difficult to make, especially in the case of rare mass lesion. Histopathology is the principal tool of diagnosis.

Keywords: vulvar lesion, lymphangioma, fibroadenoma, histopathology, excision

INTRODUCTION

Mass lesions of the vulva include both benign and malignant lesions.[1] Some are common in other sites of the body but rare at the vulva like lymphangioma circumscriptum, fibroadenoma, and epidermal cyst.[1]

Lymphangioma circumscriptum (LC) is a common form of cutaneous lymphangioma.[2] It may be primary which presents since birth or childhood, or secondary or acquired which is due to blockade in lymph flow by any reason.[2,3] It presents as small fluid-filled vesicles and often mimics condylomata acuminata, molluscum contagiosum, and tubercular lesions.[3,4]

Epidermal cysts are intradermal or subcutaneous masses within the epidermis due to invagination of squamous epithelium. It mainly presents on the face, torso, extremities, and scalp, but is rarely found on the vulva.[5,6]

Fibroadenoma is a common entity in breast tissue but very rare in the vulva. Fibroadenoma responds to hormonal changes, and therefore commonly presents in pregnancy and lactation.[7]

As these cases are very rare and very few case reports related to these vulvar lesions are in the literature, we compiled and presented this case series.

Informed consent was secured from all the patients for the photographs and publication of this case series.

CASE 1

A 20-year-old, nullipara, unmarried female presented to OPD with fluid-filled, vesicular, not very painful lesions in the vulva since childhood which increased in size with time. It was causing her uneasiness and difficulty while walking. The swelling on the vulva increased after prolonged standing.

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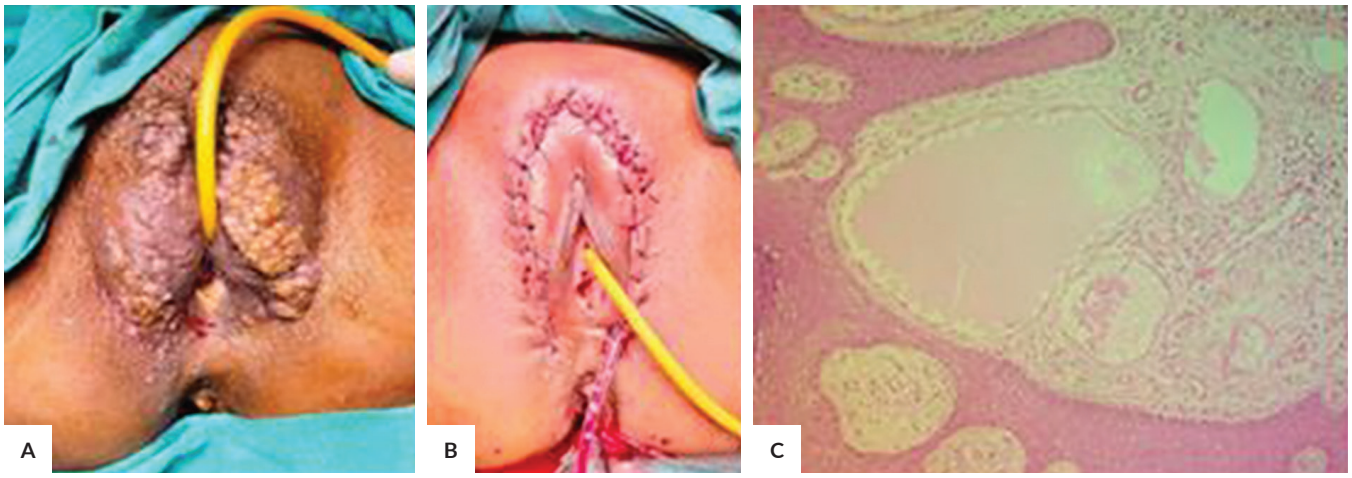

Figure 1. Lymphangioma Circumscriptum **(A)** pre-operative, **(B)** post-operative, and **(C)** microscopic (HE, 10x) pictures.

There was no history of trauma or any operative procedure. Medical, family, and psychosocial histories were not significant. On examination, fluid-filled vesicular as well as verrucous lesions, like genital warts in appearance, were seen involving bilateral labia majora and mons pubis extending from clitoris anteriorly to forchette posteriorly. There was no crusting, oozing, or discharge from the lesion. The rest of the physical examination was normal. Her laboratory tests such as complete blood count, erythrocyte sedimentation rate, and chest x-ray were within normal limits. Informed consent for doing the biopsy was secured from the patient before the procedure. A punch biopsy from the lesion was done and tissue was sent for histopathological examination which showed a lining of hyperkeratotic acanthotic stratified squamous epithelium, sub-epithelium showing large dilated lymphatic channels, lined by plump endothelial cells surrounded by dense mononuclear infiltrates (Figure 1). The features were consistent with Lymphangioma circumscriptum. As the lesion was extensive involving bilateral labia majora and mons pubis, simple vulvectomy was performed after securing informed consent. Post-operative period was uneventful. Patient was discharged four days after the procedure in satisfactory condition. Patient followed-up after two months in OPD without any complaints and examination showed apparently healthy scar tissue.

CASE 2

A 36-year-old female, parity two, presented with a mass arising from the vulva for three years which progressively increased in size. She had difficulty walking and sometimes felt pain in the lesion. There was no history of trauma or any surgery on the vulva. Medical, family, and psychosocial histories were not significant. On examination, a 6 x 5 cm, firm, non-tender mass, with restricted mobility, without any signs of inflammation was seen arising from the left labia majora (Figure 2). The rest of the physical examination

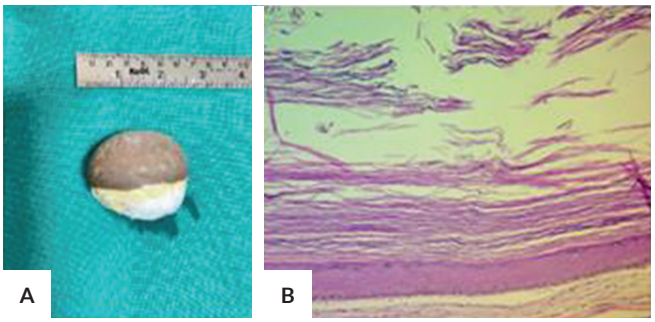

Figure 2. Epidermal cyst **(A)** gross and **(B)** microscopic (HE, 10x) pictures.

were normal. Informed consent for performing excision of vulvar lesion was secured. Excision of mass was performed and tissue was sent for histopathological examination. The mass removed was 5 x 5 x 5 cm. It was covered with skin on one side. Histopathological examination confirmed the diagnosis of an epidermal inclusion cyst. The post-operative period was uneventful and the patient was discharged on day 2 in satisfactory condition. On follow-up, after two months, the patient is well without any complaints and examination showed a healthy scar.

CASE 3

A 43-year-old parity three including two full-term normal deliveries and one first-trimester spontaneous abortion, presented with swelling in the perineal region for seven months and complained of dull pain in that area for one month. There were no other associated complaints. Medical, family, and psychosocial histories were not significant. On examination, a 3 x 3 cm, elevated, nodular mass was noted on the left labia majora in the peri-urethral area (Figure 3). The mass was firm, immobile, and non-tender without any signs of inflammation. No inguinal lymphadenopathy

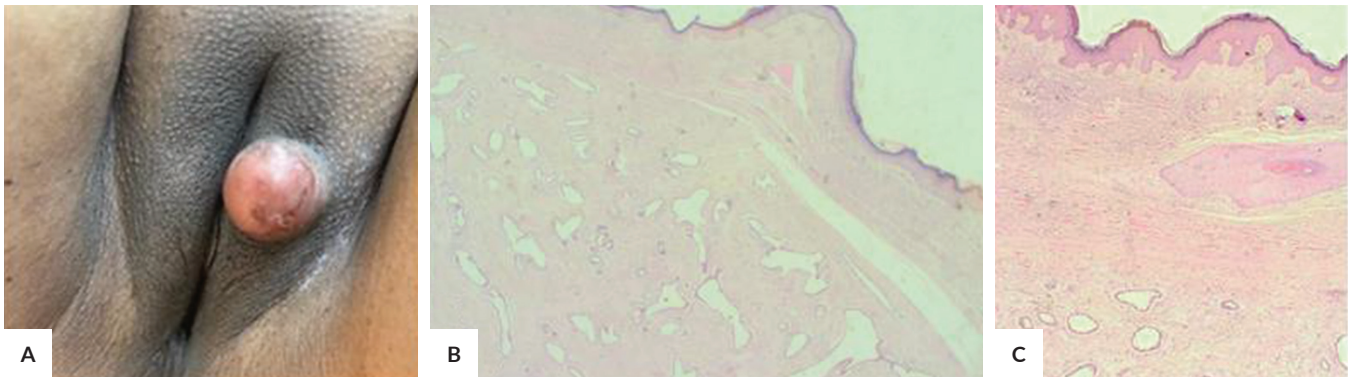

Figure 3. Fibroadenoma **(A)** gross and **(B,C)** microscopic pictures – microphotographs showing low magnification view of vulval epidermis with appendages and dermis showing a well-circumscribed lesion composed of both glandular and stromal elements in pericanalicular pattern. **(B)** – HE, 20x and **(C)** – HE, 40x.

Table 1. Description of Cases

Case	Age	Symptom	Duration	Treatment	Post-operative period	Histopathology
1	20 years	Mass at vulva, uneasiness during walking and work	Since childhood	Excision	uneventful	Lymphangioma circumscriptum
2	36 years	6 x 5 cm mass at vulva, pain and uneasiness during work	3 years	Excision	uneventful	Epidermal inclusion cyst
3	43 years	Mass at vulva 3 x 2 cm and pain	7 months	Excision	uneventful	Vulvar fibroadenoma

was noted on clinical examination. The rest of the physical and laboratory examinations were within normal. Informed consent for the excision of the vulvar lesion was secured before the procedure. Excision of the lesion was done and tissue was sent for histopathology. On gross examination, a 2 x 3 x 2 cm mass was removed. It had a glistening surface. On cut section, a fleshy, white, solid mass was seen. Histopathological examination showed a biphasic tumor composed of epithelial and stromal components. The epithelial component was in the form of round, tubular to dilated glands, lined by monomorphic round nuclei, surrounded by myoepithelial cells, embedded in the hypocellular stroma with no increase in cellularity or mitosis. These features were suggestive of vulvar fibroadenoma. The patient was well and discharged on the same day of surgery. The post-operative period was uneventful. The patient was well one month and two months of follow-up without any complaints and on examination showed a healthy scar.

All patients presented with a mass in the vulva. The first case presented as a swelling in the vulva since childhood while the other two presented during reproductive and perimenopausal age. In all cases, excision of masses was done and on histopathology, the diagnoses were different rare lesions (Table 1).

DISCUSSION

LC is characterized by small clusters of vesicular lesions containing clear lymph fluid.[2] It may be associated with lymphedema of the lower limbs.[8] Diagnosis of LC is difficult to make just by inspection as its lesions may look like condylomata acuminata, verrucous carcinoma vulva, or herpes genitalis therefore diagnosis is confirmed through a histopathology report. It arises due to blockage of lymphatic drainage. In acquired cases, it may be after surgery, or radiation therapy for carcinoma cervix. Various treatment modalities are available like electrocoagulation, superficial radiotherapy, cryosurgery, or surgical excision. The recurrence rate is high which makes it difficult to treat.[3,8] However, in this case, a simple vulvectomy was performed and the patient was well in the 6-month follow-up period without recurrence. Most case reports on lymphangioma circumscriptum in literature are of the reproductive or perimenopausal age group. In our case, the patient had a complaint since childhood. In the study by Valente et al., the lesion presented after radiotherapy for carcinoma of cervix.[4] While in the case presented by Sinha et al., the patient presented with wart-like lesion at 60 years of age.[9]

Sluga et al. also presented a series of four cases of lymphangioma circumscriptum wherein one case was acquired and three were congenital but presented at the age of more than 60 years. CO_2 vaporization was done in two cases, surgical removal in the acquired case, and only followed the patient for observation in the other case.[10] As vulvar LC is rare, there are no reports in the literature regarding its recurrence. However, in the case of cutaneous lymphangiomas, recurrence rate is around 23.1% during the 6-month to 81-month follow-up period. The recurrence rate is almost similar for all treatment modalities used.[4]

The most common site for epidermal inclusion cysts of vulva is the clitoris and these are formed after surgery or trauma.[5] In some communities, it is due to circumcision. In this case, the patient has no history of trauma or surgery and a lesion was present on labia majora. The treatment of choice is excision of mass and histopathological examination in case of large lesions or lesions which are problematic to the patient. Small cysts without any complaint can be left without any intervention.

Ectopic breast tissue is the presence of breast tissue outside the pectoral region. The most common site of ectopic breast tissue is the axilla followed by the vulva. Therefore, it may present rarely as a painless or painful mass lesion at the vulva. Milk secretion may or may not be seen from the mass. The histogenesis of vulvar fibroadenoma is not well known and controversial. The prognosis of vulvar fibroadenoma is good after excision. A case reported by Kapur et al. showed lactational changes in pre-existing vulvar fibroadenoma after four days of childbirth.[11] Most of the cases of vulvar fibroadenoma present in literature are in pregnancy or postpartum period, whereas in the present case, the patient presented with a mass lesion in the vulva in the perimenopausal age.

No unanticipated or adverse events were observed in any case in this series.

As this was a case report and few cases were reported in literature, data regarding change of intervention, follow-up, and recurrence were not available. This case series will definitely contribute to literature.

CONCLUSION

Definitive diagnosis of a mass lesion of the vulva on examination is difficult to make especially in the case of rare mass lesions. All the cases in this series were presented with mass lesions in the vulva with pain or uneasiness during working. Excision and histopathological examination of the lesions were done and the diagnoses were found to be different in all cases. Therefore, a high level of suspicion is needed for exact diagnosis. Excision or biopsy of the lesion should be performed in case of any suspicion. Histopathology is the principal tool of diagnosis. These rare vulvar lesions have good prognosis after excision without recurrence.

Statement of Authorship

All authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

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