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Original Research Paper

Gynaecology

ISOLATED NEUROFIBROMA OF VULVA AND VAGINA: A RARE CASE REPORT

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ABSTRACT Neurofibroma is a disease of peripheral nervous system that involves most commonly the extremities. Amongst female genital tract, neurofibroma involves most commonly the vulva, clitoris and labia but rarely may be seen in vagina, cervix, endometrium and myometrium as well as urinary tract. Isolated neurofibroma of vagina is a rare association which is usually a part of Von Recklinghausen's disease. We present here a rare clinical entity of vulval neurofibroma in a 52-year-old female with no other features of Von Recklinghausen's disease. The mass was excised surgically and sent for histopathological examination which confirmed the diagnosis.

KEYWORDS :

INTRODUCTION

Neurofibroma in isolation is a rare entity that involves the peripheral extremities most commonly. It usually occurs in patients with neurofibromatosis. Neurofibromatosis is an autosomal dominant inherited disorder with an incidence of approximately 1 in 3000 live births. It is characterized by hyper-pigmented lesions (café-au-lait spots), neurofibromas, iris hamartomas, macrocephaly, central nervous system tumors, defects of skull and facial bones and vascular lesions. These neurofibromas in the female gentital tract involve most commonly the vulva, clitoris and labia. They are rarely seen to involve the vagina, cervix, endometrium, myometrium and the urinary tract.[1]

Neurofibromas are categorized as cutaneous, intraneural, massive soft tissue type and sporadic neurofibromas or those in association with neurofibromatosis type1[2, 3]. They are commonly asymptomatic. Vulva is a common site for epithelial and mesenchymal tumors. Vulval neurofibroma makes up to 5% of all benign vulvar lesions.[4]

Majority of these are associated with Neurofibromatosis type 1 (NF 1) or Von Recklinghausen's disease. Vulval involvement is seen in 18 % of these cases.[5] A few of these are seen after surgical trauma. They are thought to occur spontaneously in morphologically abnormal tissues that are subjected to trauma, exact etiology is however, unclear. Significant complications from neurofibroma of vulva are generally not noted because it is benign tumor[6].

Although benign, some of the following are worrisome observations with them.

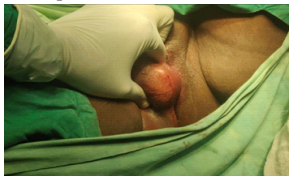
- Stress of a neoplasm
- Cosmetic concerns
- Dyspareunia and bleeding
- Injury to vital structures while excision and surgical site infectionslater
- Recurrence
- Neurofibromatosis type 1 related morbidity

Management strategies are as follows –

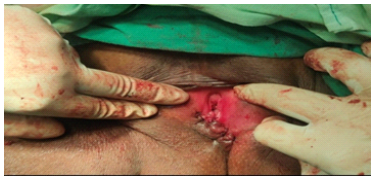
- Surgical excision -
- Vulvectomy
- Myolysis –Procedure includes inserting a needle wherein the neoplasm is subjected to electric current or cryoablation.
- Tumor embolization by interventional radiology
- Radiofrequency ablation –Herein the tumor cells are subjected to radio waves

CASE REPORT

A 52-year-old nulligravida hypertensive female presented with a history of an insidious onset progressively increasing mass involving the vulva. The mass was not associated with pain. There was no history of trauma preceding the onset. There was no associated vaginal discharge, urinary symptoms or any such similar swellings in other areas. There was history of left sided hemiparesis and Bell's palsy 7 years ago. She also had Bronchial Asthma. There was also history of cervical lymphadenopathy 35 years ago for which AKT was given. She achieved menopause 3 years ago. On examination, there was a pedunculated, soft, non-tender, mobile mass on right labia majora about 4 X 3 cm. It was seen to involve labia minora and lower third of vagina. The left side was normal. Her routine blood workup was unremarkable. Ultrasound of abdomen and pelvis showed no abnormality. X-ray Pelvis did not show any bony involvement. MRI Pelvis revealed a large well defined lobulated soft tissue mass in right labial region arising from circumferentially thickened mucosa of lower one third of vagina. Focal loss of fat planes with adjacent anterior wall of anal canal and adjacent right perineal muscles was observed. Few insignificant lymph nodes in bilateral internal iliac region were also noted. The mass biopsy was taken and sent for histopathological examination. On histopathological examination the mass was benign. It was spindle shaped with cellular arrangement as fascicles and bundles. Individual tumor cells had spindle shaped plump nuclei showing waviness at certain places. Cytoplasm was eosinophilic with indistinct cellular margins. This was diagnostic of a Neurofibroma. The mass was completely excised under spinal anesthesia and patient made an uneventful recovery. She is on routine follow-up in the outpatient clinic and is doing well with no signs of recurrence at one month follow up.



Vulval mass 6x4 cm in Right Labial fold and lower one third of vagina



Vulval mass excision under spinal anaesthesia

DISCUSSION

Vulval involvement is seen in approximately 18% of women with NF1 . Neurofibromas in the female genital tract commonly involve the vulva, clitoris and the labia. Rarely they involve vagina, cervix, endometrium, myometrium and ovaries. They may also be seen in the urinary tract. Vulval neurofibromas may result as a sequelae of surgical trauma like episiotomy or other vulval injuries. History of trauma and symptoms related to urinary tract should be sought. Genitourinary system examination including cystoscopy should also be considered while managing. The cases with NF1 may transform into malignant sarcomas. However, solitary lesions are usually benign with no reported cases of malignant transformation. Reported cases have been usually small in size (<3 cm in diameter) and slow growing. Rarely, they may be giant lesions with florid growth pattern.
They are usually asymptomatic but Venter *et al*.[3] and Gordon *et al*.[7] have reported giant lesions with intractable chronic pelvic pain and dyspareunia.

Surgical excision seems to be the sole treatment modality, however, the risk of recurrence entails a close follow-up involving both history and clinical examination.

In this case, the vulvar location of the isolated lesion and its large size in absence of other cutaneous or visceral lesions or positive family history is striking.

Solitary, genital neurofibromas have been described by Venter *et al*.[3]and Gordon *et al*.[7]presenting with intractable chronic pelvic pain and dyspareunia respectively. But the rate of growth of the tumor in our case was not very large compared to two cases reported by Venter et al[3]

Kane *et al*.[8] in their study have reported a case of isolated vulval neurofibroma in an 18-month-old female child. In our report patient was 52 years with both vulval and vaginal involvement Sa'adatu *et al*.[10] have reported case of neurofibroma of labium majus in young female as globular pedunculated cutaneous tissue.

Amita *et al*.[9] and Negi[11]*et al*. have reported a case of giant vulval neurofibroma of 17x8x6 cm presenting as soft tissue mass Hussein L Kidanto[12]*et al*. have reported a large neurofibroma of labia majora presenting as globular pedunculated mass of 20x20 cm Nibhoria S *et al*[13]reported an Isolated Vaginal Neurofibroma presenting as Vaginal Wall Cyst as a rare case report with Review of Literature . Our patient presented with mass in vulva and vagina.

Mourali et al [14]and Yayli et al [15] reported a 71 year old patient with vaginal neurofibroma with Von Recklinghausen disease. Our patient was at 52 years old and hasn't had Von Recklinghausen disease.

Baulies et al[16] reported a 20-year-old woman with a history of type-1 neurofibromatosis with a vaginal nodule neurofibroma. But our patient did not have history of type-1 neurofibromatosis.

Sharma et al[17] reported huge localized vaginal neurofibromatosis as an unusual cause of postmenopausal bleeding . In our report, the bleeding was not a manifestation.

Eusebi and Schönauer[18] reported Pigmented vaginal neurofibroma. In our report there were no pigmentations .

Gold[19] published about Neurofibromatosis of the bladder and vagina. In our report the vagina and vulva was involved.

Drescher and Herzog[20] published about neurofibromatosis of the vulva and vagina. In our report patient did not have any features of von Recklinghausen disease.

Marmey and Lacroix[21] published about Recklinghausen disease and pregnancy. But our patient was not pregnant at presentation and the neurofibroma was solitary.

Belvederi et al[22] reported Anatomo-clinical findings on a case of neurofibroma of the vagina.

During follow up, the patients should be advised to report in case of development of cutaneous lesion or any symptoms of the genitourinary tract.

CONCLUSION

To conclude isolated neurofibromas of female genital tract is not a common entity. The evaluation should consist of history and examination for presence of NF1 features and exclude genitourinary involvement. The rare incidence of these being in isolation with risk of recurrence and rare likelihood of malignant transformation emphasizes formulation of plan for close follow up of these patients post-surgical excision. Although rare, they should be considered as a differential of benign solid tumors of vagina.

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