A Case Report of Isolated Vulvar Neurofibroma Radiologically suggested to be a Lipoma

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Abstract

Neurofibroma is a benign tumor of peripheral nervous system, which can occur in isolation or be a component of neurofibromatosis type 1. It can occur in any part of the body (except brain and spinal cord) but rarely seen in the female genital areas where, if it ever occurs, the labia are the most frequent site.

We report a case of a 46yr old P0^{+2} married woman who presented with a 4 year history of an isolated, slowly growing right sided vulval mass. There were no urogenital or other systemic symptoms. She had no features of neurofibromatosis type 1. She had successful excision biopsy with histologic diagnosis of neurofibroma. The patient had good outcome and is still on follow up.

Neurofibroma of the vulva is rare and may mimic other soft tissue tumours when in solitary occurrence. Excision biopsy, histological diagnosis and keen follow-up are strongly recommended.

KEYWORDS: Neurofibroma, neural lesion, labial mass

Introduction

Neurofibroma is a peripheral nerve tissue tumour which can occur in isolation or be a component of neurofibromatosis 1. The latter also known as Von Recklinghausen's disease is an autosomal dominant systemic condition which presents commonly as peripheral nerve sheath tumor that causes typical distortion of the gross appearance of the contained tissue. It is characterized by café-au-lait spots, cutaneous neurofibromas, freckles of the axillary and ingunal regions, iris Lisch nodules, choroidal freckling etc[1]. These lesions are

mostly mesenchymal or neuroectodermal derivatives [2] and have typical histopathologic appearance described as spindle shaped cells with wavy nuclei arranged in a loose myxomatousstroma. Neurofibromatosis 1 has an incidence of 1 in about 3000 live births[3].
Isolated neurofibroma of the female genital tract is rare. When it occurs in this area, the vulvar is noted to be the commonest site; more rarely vagina, cervix, endometrium, myometrium, ovaries are involved [4,5].

Case Presentation
We report a case of a 46yr old $P0^{+2}$ married woman who presented with a 4 year history of a slowly growing right sided vulval mass. There was no similar swelling on any other external part of her body. She had no antecedent trauma in the vulvar. There were no urogenital or other systemic symptoms. She was not hypertensive or diabetic. There was no family history of tumors.

Clinical examination showed a well oriented, fully developed and healthy looking woman. There was right labia majora mass (measuring about 6cm by 4cm) with a little extension to the lower part of the adjoining mons pubis and distorting the vulval anatomy. There was no demonstrable cough impulse. The mass was firm, irreducible, non tender, relatively mobile, non fluctuant and had no differential warmth or obvious colour disparity with adjacent tissues.

Ultrasonography of the vulvar showed "oval shaped echogenic, soft tissue mass in the right majus. It was well circumscribed, measuring 3.8cmx2.0cmx3.0cm, and had no flow activity on colour Doppler evaluation. Inguinal structures were intact. The features were suggestive of vulva lipoma" Abdominopelvic ultrasound revealed normal findings.

The patient was counseled on the findings and she gave consent for excision biopsy. This was done under spinal saddle block anaesthesia and the specimen sent for histology. The histopathology report showed "a circumscribed piece of grayish white tissue. Cut surface in 2 blocks is solid and grayish. The microscopic sections show an expansive neural lesion composed of bundles of curvy neural fibres with wavy nuclei, dispersed in a myxoid and cellular loose areolar tissue. Diagnosis was neurofibroma of the labium.

The patient had good post operative recovery. She was seen one week post surgery and the wound healing was satisfactory. She was counseled on the need for follow up and was given one month appointment, then followed up six monthly and yearly. She had remained disease/recurrence-free.

Discussion
Isolated vulva neurofibroma, an uncommon benign condition managed successfully in a 46 year old nulliparous woman is presented. The condition has no age predilection as it has been reported in females of various age groups including teenagers and other women within or after reproductive age. [6-9] Its growth can be slow like the patient in this case report or have a rapid course. [7,9,10]It can also be solitary or multiple.[8]

Benign vulvar tumours may not manifest specific symptoms and some of them like neurofibromas may be asymptomatic[6] but could constitute a source of bother to the patients. Chronic pelvic pain and dyspareunia are common complaints when genital neurofibromas present with symptoms.[8]Some patients with large vulvar

tumors may have discomfort walking due to unsuitable location and size[7,8]. Others are concerned about the disfigurement or reduced function[7] of the affected part of the body. Systemic symptoms are unusually associated with isolated lesions[5].

The patient in this case report was only concerned about the distortion of her vulvar anatomy. She had no other local or systemic symptoms and didn't have features indicative of neurofibromatosis 1

Kidanto et al in Tanzania reported an isolated giant neurofibroma of the right labia majora in a 15 year old student causing itching, ulceration, discharge, difficulty in walking and temporary school discontinuation[7]. The patient had no neurofibromatosis and had total excision as definitive management

Dogra and colleagues reported a case of a long standing left labial neurofibroma with recent progressive increase in size causing difficulty in walking in a 15 year old short statured girl diagnosed of neurofibromatosis 1[8].

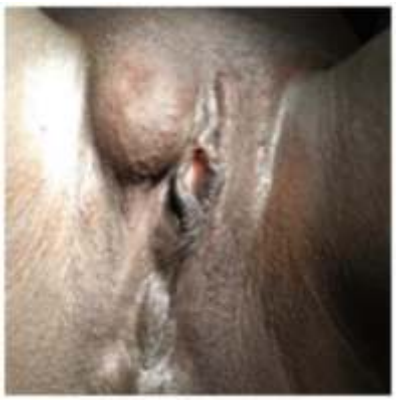
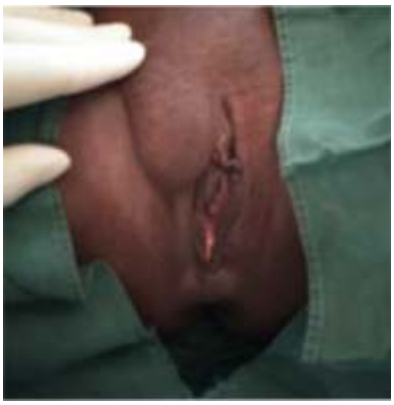
Sa'adatu et al, in northern Nigeria reported an isolated asymptomatic right vulvar neurofibroma that grew slowly over 2 years in a 21 year old nulliparous woman with no features of Von Recklinghausen's disease[9].
A large, painless, isolated right vulvar neurofibroma of 3 month duration was reported by Negi etal in a 25 year old woman[10].

Differential diagnoses of such vulvar masses such as vulva lipoma, Bartholin's cyst, inguinolabial hernia etc should be considered and meticulous attempts made at ruling them out. In the evaluation of such patients, care is also taken to exclude any history or physical trait suggestive of neurofibromatosis 1. Finding of such makes follow up paramount because of chances of malignancy or recurrence. [7,9,12] Whereas cases associated with neurofibromatosis 1 may undergo malignant transformations, [11, 12] isolated genital neurofibromas usually have benign clinical course. The case presented was free of features of neurofibromatosis 1.

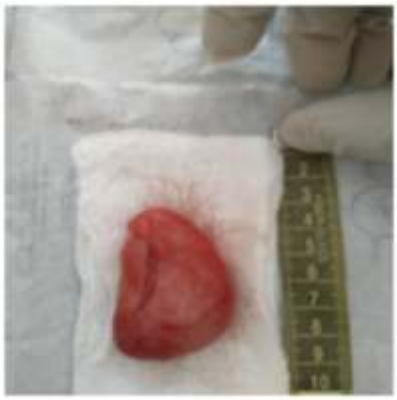
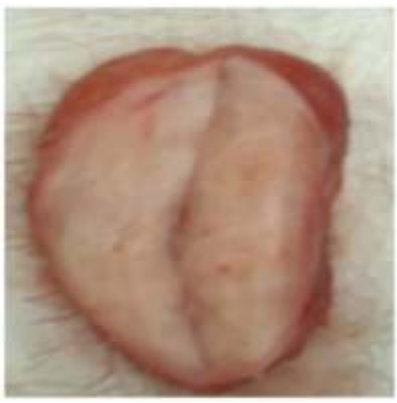
Radiological assessments of genital neurofibromas have been found to be useful especially in distinguishing superficial and invasive tumors. Magnetic resonance imaging (MRI) was favoured by some authors as preferred option preoperatively [8, 12]. Vulvar and abdominopelvic ultrasound were done for the index patient. The vulvar ultrasound findings were suggestive of lipoma. Though this was not accurate in differentiating between neurofibroma and lipoma, it was helpful in preoperative assessment of risks of malignancy. This implies that ultrasound of the vulvar in such conditions is a limited diagnostic tool as it lacks specific features for diagnosis of vulvar lipoma or neurofibroma. This was the case with the index patient.

From the reviewed literatures, the most popular management approach as was also done for the patient in this report include total surgical excision, prompt histologic diagnosis and follow up.

Conclusions:
Neurofibroma of the vulva is a rare condition and may mimic other soft tissue tumours when in solitary occurrence. Excision biopsy, histological diagnosis and keen follow-up are strongly recommended.

Figs 1&2: Pre op Gross appearance of Right labial mass

Figs 3 & 4 Surgical specimens

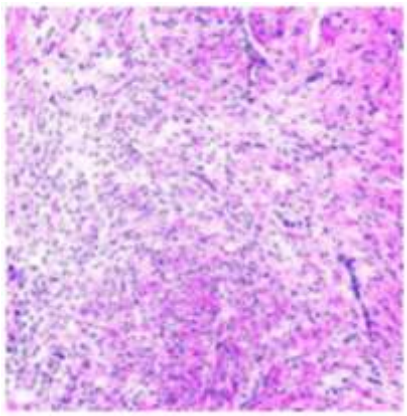


Fig 5: Histologic photomicrograph

Reference

1. Friedman JM. Neurofibromatosis 1 GeneReviews [Internet]. Available @https://www.ncbi.nlm.nih.gov/books/NBK1109/? ga=2.74586962.1139702472.1553944849-1551131474.1553944849. Revised May 2018 and accessed March 2019
2. Shahzad R, YounasFarhana. PlexiformNeurofibroma of Uterus: A Rare Manifestation of Neurofibromatosis 1. Journal of the College of Physicians and Surgeons Pakistan 2014; 24 (Special Supplement 1): S22-S23
3. Nasir A, AbdurRehman L, Ibrahim K, Adejoke M, AfolabiJ,Adeniran J. Genitourinary plexiformneurofibroma mimicking

sacrococcygealteratoma. J Surg Tech Case Rep 2012; 4:50-2.

4. Gómez LAM, Martínez DF, Izquierdo SB, Vicente SEM, Salmerón R; Localized neurofibromatosis of the female genital system: a case report and review of the literature. J ObstetGynaecol Res. 2012; 38(6):953-6.

5. Nibhoria S, Tiwana K K,Kaur M, Phutela. Isolated Vaginal Neurofibroma Presenting as Vaginal Wall Cyst: A Rare Case Report With Review of Literature.J Family Reprod Health. 2016; 10(1): 49–51.

6. Garg C, Agarwal A, Agrawal R, Kumar P. Neurofibroma at Unusual Locations: Report of Two Cases in Teenage Girls. Journal of Clinical and Diagnostic Research. 2015;9(4): ED03-ED04

7. Kidanto Hl, Garrison J,WangweP. Largeneurofibroma of the labia majora: A case report. Tanzania Journal of Health Research. 2013;15(1):1-4 Doi: http://dx.doi.org/10.4314/thrb.v15i1.10

8. Dogra BB, Ahmed S, Kandari A, Virmani R. Plexiform neurofibromatosis of vulva. Int J Res Med Sci 2014; 2:1771-3.

9. Sa'adatu TS, Shehu SM, Umar HS. Neurofibroma of the labium majus: A case report.NigerainJounal of Surgical Research. 2006; 8 (1 – 2): 99-100

10. Negi L, Wahal SP, Gulati A, Kaushal V. Giant vulvalneurofibroma: A rare entity with review of literature. J Can Res Ther 2015;11:1045

11. Spyk SL-v, Thomas N, Cooper DN, Upadhyaya M. Neurofibromatosis type 1-associated tumours: their somatic mutational spectrum and pathogenesis *Human genomics*, 2011; 5(6): 623–690.

12. Musella A, Palaia I, Domenici L, Casorelli A, Martoccia A, Panici PB. Vulvar Malignancy in Neurofibromatosis Syndrome. Case Reports in Obstetrics and Gynecology. Article ID 217924. Available on line @https://doi.org/10.1155/2013/217924 . Accessed on 18/04/2020