Neurofibroma: A Rare Case of Vulva Tumour

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Abstract: Vulva tumors are neoplasms of the vulva that occur in small percentage (3%) of female genital cancers which can be benign or malignant or vulva cancer. Vulva neoplasms are divided into cystic or solid lesions and other mixed types. The most common benign tumors of the vulva are fibromas, papillomas, lipomas, angiomas, and others. A young woman, 25 years old, came to Bintang Amin Hospital OBGYN clinic, complaining of a huge mass in the front area of left labia major from one year before. She never had any examination before and felt the mass become larger and caused pain and obstacle in her daily activity. We found a huge pedunculated tumor, size 9 x 6 x 1.5 cm. After excision and pathological examination, we diagnosed the patient with benign neurofibroma and suggested follow up if another lesion or tumor occur.

1 INTRODUCTION

Vulva tumors are neoplasms of the vulva occur in small percentage (3%) of female genital cancers which can be benign or malignant or vulva cancer. Vulva neoplasms are divided into cystic or solid lesions and other mixed types. Epithelial and mesenchymal tissue are the origin of vulva tumors. Vulva tumors are generally a rare disease encountered in gynecological clinical practice, especially in the elderly young. Vulva tumors, more often occur in the labia majora and rarely in the labia minora, clitoris, vestibule and commissure posterior. The most common benign tumors of the vulva are fibromas, papillomas, lipomas, angiomas, and others.

2 CASE PRESENTATION

Patient complained of a lump on the labia major sinistra. accompanied by itching. The lump is the size of a duck egg (sagging) about 8cm. The lump has been felt for about 3 years. The mass caused. Obstacles when she sits, walks or even does daily activity. The patient claimed to have no history of other diseases such as asthma, hepatitis, hypertension, DM, and heart disease. The patient's family has no history of diseases such as asthma, hepatitis, hypertension, DM, heart disease and lung disease. (Figure 1)

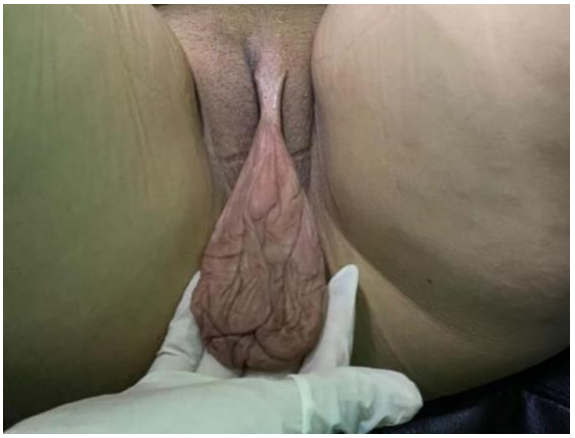

Figure 1. Mass of Vulva.

We diagnosed her with suspected benign vulva tumor and suggested for excision and pathologinal examination, The differential diagnosis were angiomyxoma, malignant peripheral nerve sheath tumor, neurofibroma, perineuroma, dermatofibroma, schwannoma, dermatofibrosarcoma protuberans, and ganglioneuroma. We performed an excision with spinal anesthesia at October 21ST 2023. We managed the wound and treated patient with analgetic. The

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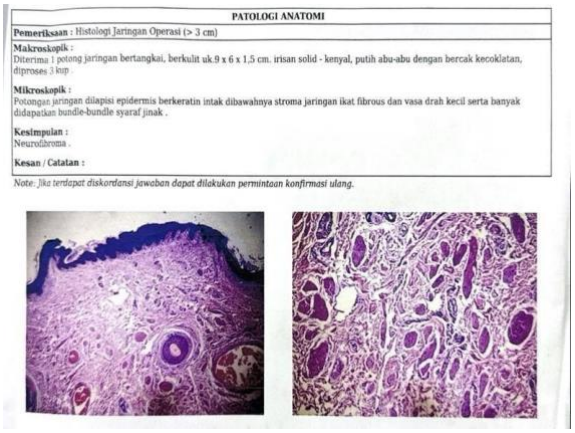
pathological examination showed the tissue was surrounded intact keratinized epidermis with fibrous mesenchymal stroma and vessels and many benign nerve bundle, suggesting neurofibroma. (Figure 2)


PATOLOGI ANATOMI

Pemeriksaan : Histologi Jaringan Operasi (> 3 cm)

Makroskopik :
Diterima 1 potong jaringan bertangkai, berkulit uk.9 x 6 x 1,5 cm. irisan solid - kenyal, putih abu-abu dengan bercak kecoklatan, diproses 3 kup .

Mikroskopik :
Potongan jaringan dilapisi epidermis berkeratin intak dibawahnya stroma jaringan ikat fibrous dan vasa drah kecil serta banyak didapatkan bundle-bundle syaraf jinak .

Kesimpulan :
Neurofibroma .

Kesan / Catatan :

Note: Jika terdapat diskordansi jawaban dapat dilakukan permintaan konfirmasi ulang.

Figure 2. Histopathology result.

3 DISCUSSION

Neurofibroma is the most common benign tumor of the peripheral nerve sheath. It often presents as a soft, skin-colored papule or small subcutaneous nodule, arising from the endoneurium and connective tissue of the peripheral nerve sheath. Neurofibromas are composed of Schwann cells, fibroblasts, perineural cells, and mast cells with a variable myxoid background. (Burger et al, 2002; Tamer et al, 2003)

Isolated neurofibromas of the female genital tract are very rare and mostly associated with neurofibromatosis. Neurofibromas are a disease of the peripheral nervous system and most commonly occur in the extremities. Among the female genital tract, neurofibromas most commonly affect the vulva, clitoris, and labia but are rarely seen in the vagina, cervix, endometrium, myometrium, and urinary tract. (Gordon et al, 1996) The vulva is the most frequent genital location, but it has rarely been reported in the context of the vagina, uterine cervix, or ovaries. Despite its rarity, neurofibroma is a neoplasm that should be considered in the differential diagnosis of pelvic masses, especially in patients with neurofibromatosis. The solitary lesions are rare and usually, they are not associated with any systemic symptom.

Neurofibromas are categorized as cutaneous neurofibromas, intraneural neurofibromas, massive soft tissue neurofibromas, and sporadic neurofibromas or those associated with neurofibromatosis 1. Solitary lesions are rare and usually not associated with any systemic symptoms. (Mourali et al, 2009) About 90% of cases occur sporadically, while the remaining cases are associated with neurofibromatosis type 1 or 2. Mutations in the NF1 gene cause neurofibromas. There are three main types of neurofibromas: localized (most common), diffuse, and plexiform. Although most neurofibromas occur sporadically and have a very low risk of malignant transformation, the plexiform type is pathognomonic for neurofibromatosis type 1 (NF 1). This disease increases the risk of malignant transfo A rare benign tumor called angiomyofibroblastoma typically develops in the vulva. A lipoma, Bartholin cyst, or severe angiomyxoma could be the differential diagnosis. (Sims and others, 2012) The preferred course of action is straightforward complete excision, which is typically curative. The lower female genital tract (FGT) is home to a variety of mesenchymal cancers, such as cellular angiofibroma (CAF), angiomyofibroblastoma (AMFB), and aggressive angiomyxoma (AAM).

In 2022, Haroon et al. With a few minor histological variations, these tumors are histologically formed of stromal cells intermingled with vasculature. Because of the variations in the prognosis, a precise diagnosis of these malignancies is crucial. Verrucous carcinoma resembles cauliflower-like growths resembling genital warts and is a subtype of aggressive squamous cell vulva cancer.

The majority of women who have invasive vulva cancer will experience symptoms. If there is a region of the vulva that appears abnormal—that is, lighter or darker than the surrounding normal skin—or appears red or pink, we need to be aware of it. It's also important to observe any lump or bump, which may be red, pink, or white, feel thick or rough, or have a surface resembling a wart. Additional symptoms to take into account include thickening of the vulva skin, itching, soreness, or burning, bleeding or discharge that is not associated with a typical menstrual cycle, and an open sore (particularly if it persists for more than a month). Vulva melanomas might be white, pink, red, or any other color; however, the majority are black or dark brown. Although they are present throughout the vulva, the majority of them are on the labia majora or minora or in the vicinity of the clitoris. (Dobrica et al. 2021)

Patient should be advised to visit if a new lesion appeared and the treatment was deemed enough. Physical examination and/or excisional biopsy along with microscopic inspection are frequently used in the evaluation of isolated superficial lesions. To

determine the degree of involvement, prepare for surgical excision, and submit larger lesions to the pathology department for histology, further biopsy and/or CT/MRI imaging may be necessary.

Complete surgical excision is the preferred in most situations, and local recurrence is quite uncommon. There are still no substitute treatments available for cutaneous neurofibromas. When comprehensive surgical excision is not feasible due to diffuse or plexiform neurofibromas, the lesion is frequently completely removed for esthetic or symptomatic relief. The clinical practitioner may decide to monitor these patients for rapid growth or recurrence. With varying degrees of success, interferon-alpha has been investigated as an adjuvant treatment for plexiform neurofibromas. (Legius et al, 2021).

The following minor complications are linked to surgically excising the lesion and are commonly connected with localized neurofibromas: discomfort, hemorrhage, scarring, and local infection. Complications with plexiform lesions stem from the inherent dangers associated with surgery and, in rare instances, from the inability to remove the lesion entirely. There is a higher chance of malignant development into malignant peripheral nerve sheath tumors in patients with NF1 and chronic lesions. (Legius et al, 2021).

4 CONCLUSIONS

Vulva tumors are generally a rare disease encountered in gynecological clinical practice, especially in the elderly young. Vulva tumors, more often occur in the labia majora and rarely in the labia minora, clitoris, vestibule, and commissure posterior. The most common benign tumors of the vulva are fibromas, papillomas, lipomas, angiomas, and others. Despite its rarity, neurofibroma is a neoplasm that should be considered in the differential diagnosis of pelvic masses, especially in patients with neurofibromatosis. The solitary lesions are rare and usually, they are not associated with any systemic symptom.

REFERENCES

Gordon MD, Weilert M, Ireland K. Plexiform neurofibromatosis involving the uterine cervix, endometrium, myometrium, and ovary. Obstet Gynecol 1996;88:699-701.

Burger PC, Scheithauer BW, Vogel FS. Surgical Pathology of the Nervous System and Its Covering vol 1, 4th edn. Philadelphia: Churchill Livingstone 2002:594–611.

Tamer E, Yalein B, Alli N. Multiple nodular plexiformneurofibromas in a neurofibromatosis1 family: A familial tendency? A case report and review of the literature. Int J Dermatol 2003;42:357–8.Finkel G, Lane B. 2019. Granular cell variant of neurofibromatosis: ultrastructure of benign and malignant tumors. Hum Pathol. 13(10):959-63.

Mourali M, Hammedi N, Fekih C, Essoussi-Chikhaoui J, Binous N, Ben Zineb S, et al. Vaginal neurofibroma. Tunis Med 2009; 87: 411.

Raffensperger J, Cohen R. 2020. Plexiform neurofibromas in childhood. J Pediatr Surg.;7(2):144-51.

Ros Briones, R., Arredondo Montero, J., Bronte Anaut, M., Hernández-Martín, S., & Guarch Troyas, R. (2023). Pediatric Vulvar Superficial Angiomyxoma: A Case Report With Clinical, Radiological, and Anatomopathological Characterization and a Comprehensive Review of the Literature. International Journal of Surgical Pathology, 31(5), 852-860.

O'Brien, K. L. O. F., Cortes-Santiago, N., Patil, N. M., & Bercaw-Pratt, J. L. (2020). A rare case of vulvar superficial angiomyxoma in a pediatric patient. Journal of Pediatric and Adolescent Gynecology, 33(6), 727-729.

Haroon, S., Irshad, L., Zia, S., Ali, A. H., Dowlah, T. U., Rashid, K., ... & Khan, A. (2022). Aggressive Angiomyxoma, Angiomyofibroblastoma, and Cellular Angiofibroma of the Lower Female Genital Tract: Related Entities With Different Outcomes. Cureus, 14(9).

Dobrică, E. C., Vâjâitu, C., Condrat, C. E., Crețoiu, D., Popa, I., Gaspar, B. S., ... & Varlas, V. N. (2021). Vulvar and vaginal melanomas—the darker shades of gynecological cancers. Biomedicines, 9(7), 758.

Harder A, Wesemann M, Hagel C, Schittenhelm J, Fischer S, Tatagiba M, Nagel C, Jeibmann A, Bohring A, Mautner VF, Paulus W. 2022. Hybrid neurofibroma/schwannoma is overrepresented among schwannomatosis and neurofibromatosis patients. Am J Surg Pathol.;36(5):702-9.

Sims, S. M., Stinson, K., McLean, F. W., Davis, J. D., & Wilkinson, E. J. (2012). Angiomyofibroblastoma of the vulva: a case report of a pedunculated variant and review of the literature. Journal of Lower Genital Tract Disease, 16(2), 149-154.

Hornick JL. 2019. Limited biopsies of soft tissue tumors: the contemporary role of immunohistochemistry and molecular diagnostics. Mod Pathol ;32(Suppl 1):27-37.

Sims SM, Stinson K, McLean FW, Davis JD, Wilkinson EJ. Angiomyofibroblastoma of the vulva: a case report of a pedunculated variant and review of the literature. J Low Genit Tract Dis. 2012 Apr;16(2):149-54.

Baulies S, Cusidó MT, Grases PJ, Ubeda B, Pascual MA, Fábregas R. Neurofibroma of the vaginal wall. Clin Exp Obstet Gynecol. 2008;35(2):140-3.

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

Sharma JB, Wadhwa L, Malhotra M, Arora R, Garg A, Singh S. Huge localized vaginal neurofibromatosis: an

unusual cause of postmenopausal bleeding. J Obstet Gynaecol Res. 2004 Apr;30(2):96-9.

Gómez-Laencina AM, Martínez Díaz F, Izquierdo Sanjuanes B, Vicente Sánchez EM, Fernandez Salmerón R, Meseguer Peña F. Localized neurofibromatosis of the female genital system: a case report and review of the literature. J Obstet Gynaecol Res. 2012 Jun;38(6):953-6.

Işık E, Onay H, Atik T, Solmaz AE, Özen S, Çoğulu Ö, Darcan Ş, Özkınay F. A Neurofibromatosis Noonan Syndrome Patient Presenting with Abnormal External Genitalia. J Clin Res Pediatr Endocrinol. 2020 Mar 19;12(1):113-116.

Lee TJ, Chopra M, Kim RH, Parkin PC, Barnett-Tapia C. Incidence and prevalence of neurofibromatosis type 1 and 2: a systematic review and meta-analysis. Orphanet J Rare Dis. 2023 Sep 14;18(1):292.

Legius E, Messiaen L, Wolkenstein P, Pancza P, Avery RA, Berman Y, Blakeley J, Babovic-Vuksanovic D, Cunha KS, Ferner R, Fisher MJ, Friedman JM, Gutmann DH, Kehrer-Sawatzki H, Korf BR, Mautner VF, Peltonen S, Rauen KA, Riccardi V, Schorry E, Stemmer- Rachamimov A, Stevenson DA, Tadini G, Ullrich NJ, Viskochil D, Wimmer K, Yohay K; International Consensus Group on Neurofibromatosis Diagnostic Criteria (I-NF-DC); Huson SM, Evans DG, Plotkin SR. Revised diagnostic criteria for neurofibromatosis type1 and Legius syndrome: an international consensus recommendation. Genet Med. 2021 Aug;23(8):1506- 1513.