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An Unusual Case of Vulvar Schwannoma: A Case Report and Review of Literature

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ABSTRACT

Background & aim: Schwannoma seldom occurs in female genitalia; with only a few cases being reported in the medical literature. This tumor can present on labia majora, labia minora, clitoris, and vagina. In this study, a case of vulvar schwannoma, which resembled a leiomyoma, is reported.

***Case report*:** A 44-year-old woman presented with a history of a small palpable mass located on the left labium. Ultrasonography showed a heterogeneous mass suggestive of a leiomyoma. Subsequently, the patient underwent surgical excision of the mass with clear margins. Histopathological examination confirmed the diagnosis of vulvar schwannoma. The patient was discharged in good condition following the excision.

***Conclusion*:** Schwannoma of the vulva represents a rare neoplasm that can be treated by surgical excision. The prognosis is excellent and recurrence is uncommon. Therefore, histopathological assessment is recommended for management of Schwannoma of the vulva.

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Introduction

Schwannoma, also named as neurilemoma, is a rare tumor of the peripheral nerves (1). This tumor is a solitary benign overgrowth of the nerve sheath cells within a well-defined capsule in the peripheral nervous system. It can be developed sporadically or as a component of a familial disorder like neurofibromatosis type 2 (NF2) or Carney complex syndromes (2). Almost 90% of schwannomas are sporadic, while 3% are associated with NF2, 2% with schwannomatosis, and 5% with meningiomatosis (2).

The prevalence of schwannoma peaks between 20 and 50 years of age, with no gender or race predominance (3). The most common site of involvement is upper extremity, followed by head (including the oral cavity, orbit and salivary glands), neck, and lower extremities (1). Other sites including posterior mediastinum and retroperitoneum are also reported to be involved in some cases (3).

This tumor seldom affects the genitalia. In the female genitalia, it usually affects the labia, clitoris or vagina as a small, painless and slow growing mass, that is often diagnosed by biopsy and pathological assessment (3). Immunohistochemistry may be useful to confirm the diagnosis and exclude differential diagnoses, especially malignant conditions (4). Few cases of vulvar schwannoma have been reported so far (1, 3-12). Here, a case of labial schwannoma, initially resembling leiomyoma, is reported.

Case presentation

A 44-year-old woman was referred to our clinic with a painless, and mobile mass located

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in subcutaneous tissue of the vulva, with no changes in the overlying skin, and no ulceration or bleeding.

The tumor was present for several months and was slowly increasing in size; however, she did not seek any medical help. The patient also had previous a history of uterine curettage, appendectomy, and mitral valve insufficiency. Her history was otherwise pointless, with no recent weight loss or irregular menstruation.

Physical examination revealed a 3×2 cm mass palpable on the left labia, with no other visible abnormalities. Ultrasonography reported a well-defined heterogeneous mass, suspicious of leiomyoma. Ultrasound showed that the size of the tumor was 33×21×11 mm. All laboratory blood tests revealed normal results.

The patient underwent a minor surgery for resection of the tumor with a safe margin of the surrounding tissue. The macroscopic appearance of the mass was indicative of a leiomyoma (Figure 1).

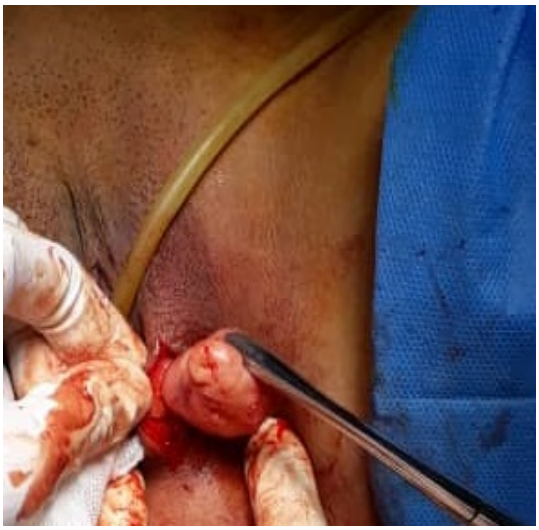

Figure 1. Macroscopic feature of the tumor

Unexpectedly, microscopic histopathologic assessment showed an encapsulated neoplastic growth of mesenchymal cells, as along with a few spindle-shaped monomorphic cells demonstrating no significant mitotic activity (Figure 2)These findings indicated the diagnosis of schwannoma. The patient discharged with good general condition, and showed no complication or recurrence during the 6 months follow up visits after the surgery.

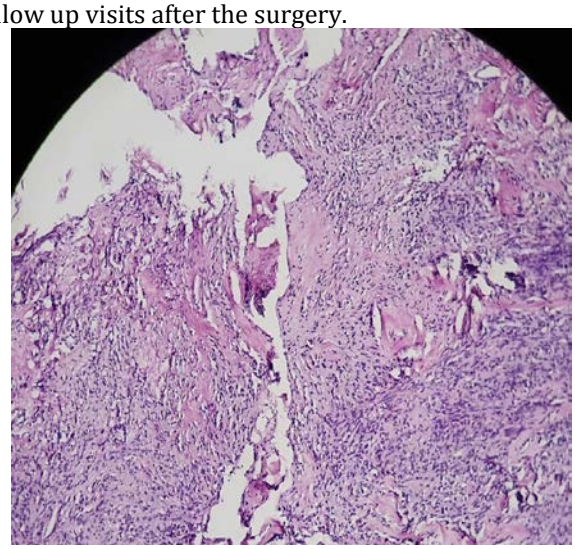

Figure 2. Microscopic feature of the tumor showing Verocay bodies

Discussion

Although the female reproductive system is prone to mesenchymal tumors, they rarely involve vulva. Moreover, the most commonly diagnosed mesenchymal tumor is leiomyoma and other vulvar tumor such as schwannoma seldom occur in female genitalia (13-14). According to the results of literatures review for the vulvar Schwannoma that is shown in Table 1, Schwannoma could affect females of different ages. However, it mostly involves women aged 20 to 50 years (3). Despite the benign nature of this tumor, these excision is essential to exclude the possibility of a malignant differential diagnosis (3).

Schwannomas vary in size, ranging from 0.6×0.5cm (3) to 15×12 cm (5). A literature showed that the tumor can affect different parts of the female genitalia, but involvement of vaginal wall (10) and clitoris (1,11-12) is extremely rare. The reported cases of schwannoma in the female genitalia sowed that labia were the most commonly affected site (3, 7). Moreover, only two cases of recurrence have been reported so far (4). The present case was also consistent with the majority of previous reports in this regard, as a relatively small mass was found in the left labium majora.

Table1. Review of Vulvar schwannoma Cases inthe literature

Authors/year	Age	Tumor Location	Swelling duration	recurrence	size
Kocak A et al /2021 (1)	46	Clitoris	2 years	NR	2 × 3 cm
Vhriterhire R ,et al/2019 (3)	18	Right labium majus	2 years	NR	10.5 × 7 × 6.5 cm
José Y et al/2017 (4)	45	Left labium majus	2 years	2 years after first excision	4 to 5 cm
Bozkurt M et al/ 2013 (5)	65	Left vulva	Several years	NM	15x12 cm
Sammarco AG et al/2017 (6)	38	Clitoris	28 years	NR	15 cm × 6 cm
Tan J et al/2018 (7)	58	Left labium majus	10 years	NM	1 cm × 1 cm
Yu YS et al /2011(8)	76	Clitoris	Not stated	NR	2.0 × 1.7 cm
An X et al/2017(10)	48	Left side of the vagina	1 year	NR	3.5×2.5 cm
Chuang WY et al/2007(11)	41	Clitoris	Not stated	NR	2×2 cm
Park ST et al/ 2015(12)	37	Clitoris	3 years	NR	3-cm
Ayati E et al/2022(13)	31	Right labia majora	Few months	NR	10 × 4 cm
Woodruff, J.M et al /1983 (16)	26	Vulva	Not stated	2 times	Not stated

The preoperative diagnosis of schwannoma is challenging. Medical history and physical examination are often non-specific and usually the physician encounters a painless, immobile, and non-tender mass. The size of the tumor and the involved nerves can affect the clinical presentation. Pain and neuralgia may be present in large tumors (5), while small tumors like the present case presented with only a painless mass.

Pre-operative radiologic assessment is only important for exclusion of other malignant masses and is not helpful in the diagnosis of schwannoma (5). Ultrasound scan shows a well-circumscribed mass, possibly with displacement of adjacent structures without direct invasion. Moreover, cystic or fatty degeneration may be present that is responsible for heterogeneous feature of the mass in ultrasonography (15). Therefore, diagnosis is mainly based on the histopathologic findings and exclusion of other conditions.

The typical histopathological feature of schwannoma includes s a tightly packed sheath of spindle cells arranged in palisade and swirling patterns, surrounded by areas of loosely packed spindle cells with small round nuclei, referred to as Verocay bodies (3). Cystic degeneration may be present in schwannomas and large masses may even present central necrosis. Accumulation of hyaline bodies around blood vessels can also cause vessel wall thickening (3). Schwannomas are divided into conventional, plexiform, ancient, and cellular forms based on their histopathological appearance, among which conventional is the most common and cellular is the least common (3).

Immunohistochemistry can differentiate malignant cases from a benign schwannoma, as benign cases show high levels of S-100 protein expression while malignant ones barely express it (3,12). Schwannoma cells may also present laminin, type IV collagen, glial fibrillary acidic proteins, and markers such as vimentinor KP1 (CD68) (1,8). Furthermore, benign cases exhibit a low mitotic activity of 3-4 per high-power field at most (3,12). Overall, malignant transformation occurs in less than 1% of schwannomas, and this is even rarer among vulvar schwannomas (5).

Treatment of schwannoma is based on total resection, which leads to a favorable prognosis. Recurrence can rarely happen due to the incomplete resection. Therefore, resection of the tumor with a safe margin of surrounding tissue

is important to prevent recurrence (7). Schwannomas do not infiltrate the adjacent nerve; thus, they can be removed without serious damage to the nerve function. However, the patient may occasionally develop temporary complications such as neuropraxia. Long-term follow-up is therefore advisable after surgical removal (15). Fortunately, the present case had no complication after surgery and during the follow up period.

Conclusion

Vulvar schwannoma is a rare tumor that mainly affects women of reproductive age. Imaging and history are non-specific, and only histopathological assessment can lead to definitive diagnosis. Although the condition is difficult to diagnose, the treatment is straightforward and the patient can be cured with complete resection. Recurrence is not predictable when complete resection of tumor with a safe margin is performed. Further studies and larger number of cases with longer follow up are needed to provide a complete understanding of the diagnosis and management of this condition.

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Conflicts of interest

The authors declared no conflicts of interest.

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