

Case report of vulvar schwannoma and literature review

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ABSTRACT

Schwannoma is a rare benign encapsulated tumor derived from neural sheath cells. The tumor is more common in the head and neck regions and the extremities. Its location in the vulvovaginal region is a rare phenomenon with a few reported cases worldwide. We report a case of a large vulvar schwannoma occurring in an 18-year-old woman, primigravidae at 35 weeks gestation. The ulcerated tumor mass which measured 10.5 × 7 × 6.5 cm and weighed 249.2 grams was located on the right labium majus with extension into the vagina. The tumor evoked strong immunohistochemical reactivity to S-100. There was no evidence of recurrence after several weeks of excision of the neoplasm.

Key words: Pelvic nerve sheath tumor; schwannoma; vulva mass.

Introduction

Schwannoma, also known as neurilemoma or neurinoma, is a benign encapsulated tumor of proliferating differentiated Schwann cells of nerve sheaths. This tumor is most common between the fourth to the sixth decade. There is an equal gender occurrence of schwannomas.^[1]

The tumor has a peculiar characteristic of predilection for the head and neck regions and the flexor surfaces of upper and lower limbs.^[1] Situation in deep regions such as the retroperitoneum and the mediastinum has been recorded in the literature. The growth of this tumor in the vulva or vagina is rare and only a number of cases have been observed.^[1] The tumor is usually small in size in most locations except in the mediastinum and retroperitoneum. We report below a case of a large schwannoma involving the vulva in an 18-year-old woman.

Case Report

An unbooked 18-year-old, primigravidae Nigerian woman, presented with a large ulcerated bleeding mass of

long-standing duration attached mainly to the right labium majus.

The tumor had been present for several months and slowly increased in size. She had previously sought surgical intervention in a rural hospital 2 weeks prior to presentation. There was a markedly accelerated growth of the swelling after the surgical intervention with subsequent enlargement and descent of the mass beyond the vulva, bleeding and pain, features not previously present before the intervention.

She was at 35 weeks' gestation and referred from the rural hospital on account of the accelerated growth of the vulvovaginal lump and uncontrollable hemorrhage. The lump had been noticed 2 years prior to presentation, initially asymptomatic and slowly increasing in size. However, 3 weeks prior to surgical intervention at the rural hospital,

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Access this article online	
Website: www.tjogonline.com	Quick Response Code 
DOI: 10.4103/TJOG.TJOG_62_18	

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How to cite this article: Vhriterhire RA, Swende TZ, Onche EP, Terhemba N. Case report of vulvar schwannoma and literature review. Trop J Obstet Gynaecol 2019;36:147-151.

she had developed paraesthesia in the inner aspect of the right thigh and subsequently started limping with the right lower limb. She was markedly pale, dehydrated, and was in shock with a pulse rate of 142 beats per minute, a thready small volume pulse, and blood pressure of 80/40 mmHg. There was a well - circumscribed but ulcerated gray mass, measuring 10 cm by 8 cm protruding through an incision in the right labium majus and also involving the right vaginal wall. Her urgent packed cell volume was 20%; her blood group was O+ and serum electrolytes were essentially within normal reference ranges except for elevated serum urea probably due to dehydration. The obstetric ultrasound scan showed a live fetus with an estimated fetal weight of 2.1 Kg.

The mass was excised, accompanied by evacuation of a pelvic hematoma collection and repair of lacerated tissues. The appearance of the swelling on presentation is shown on Figure 1. The excised mass was sent to the histopathology laboratory for processing and evaluation.

Her postoperative recovery was remarkable and she was discharged on the 6th postoperative day. She came for follow-up at 2 weeks, 4 weeks, 8 weeks, and 12 weeks after discharge. The patient remained stable on follow-up after discharge from the hospital. The vulvovaginal wound healed without any evidence of recurrence. The baby was delivered at term via a cesarean section.

A large ovoid encapsulated firm tissue specimen, preserved in formalin, was received and accessioned in the histopathology laboratory. On gross examination, the specimen measured 10.5 × 7 × 6.5 cm, weighed 249.2 grams; had a volume of 201 mls (determined by fluid displacement) and a calculated density of 1.24 g/cm³. The cut section showed



Figure 1: Vulva schwannoma: There was ulceration of the vulva and protrusion of a large oval mass which extended to involve the lower part of the vagina at presentation in the hospital

a whitish fleshy surface [Figure 2]. Sections cut from the paraffin wax embedded tissue blocks were routinely stained with hematoxylin and eosin. Histological evaluation revealed a classical poorly cellular (Antoni B) and cellular regions (Antoni A). Most of the tumor consisted of the cellular regions made up of fascicles of spindle-shaped cells with indistinct cell borders and elongated serpentine nuclei with tapered ends. The nuclei of these cells were arranged in characteristic palisades separated by clefts of eosinophilic fibrils (Verocay bodies). Average mitotic counts of 2/10 high power fields (hpfs) were observed. The histological features are shown on Figures 3 and 4. There was a strong positive immunohistochemical reaction of the tumor cells to S-100. The observed histological features were consistent with the diagnosis of a conventional benign schwannoma. The tumor lacked degenerative changes, such as cyst formation, necrosis, or foci of calcification.

Discussion

Mesenchymal tumors in the female reproductive tract range from the commonly occurring leiomyoma to rarely encountered lesions.^[2] Mesenchymal tumors in the vagina and vulva regions are not common, especially when compared with epithelial neoplasms.^[3] Those of neural origin are still a much smaller subset. Schwannomas which arise from the neural sheath are only rarely encountered in gynecological practice and a few cases have been reported.^[4,5] Table 1 below shows some cases involving the vulva and adjacent structures reported in the literature.^[1,6-26] Presented on this table are some important features including the age of the patient, duration of the swelling, histological subtype, specific site, size, and presence or absence of degenerative changes such as necrosis or cyst formation.



Figure 2: Schwannoma of the vulva: The mass consisted of an encapsulated grey brown firm mass with a white fleshy cut surface

In the pelvic region, the tumor may be found attached to the vulva,^[14] in the vagina,^[18] cervix,^[27] uterus,^[5] perineum, or rectovaginal pouch.^[28] Vulvar region schwannomas have been reported to occur in different locations, such as the clitoris,^[11] labia minora,^[1] or labia majora. The tumor in the present reported case was found to be arising from the labium majus, consistent with previous observation that most vulvar schwannomas arise from the labia.^[23] The encounter of the physician with this tumor should prompt further examination of the patient for other neural tumors such as neurofibromatosis.

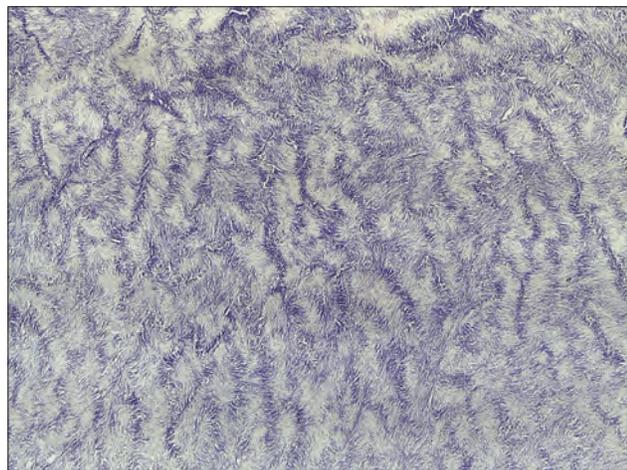


Figure 3: Schwannoma of the vulva: This microscope section of the cellular Antoni A area of the tumor shows the classical fascicles of Schwann cell palisades (Verocay bodies). X4 objective magnification, H and E

The sizes of the reported cases of schwannomas occurring in the perineal region of women vary widely, from the very small, barely palpable to huge masses [Table 1]. Since the tumor grows as a painless swelling, intervention may be sought by the patient either to obviate the swelling metamorphosing into something more sinister, or because movement has become difficult due to pressure effect. The tumor may be present for many years without any further increase in size.^[1] The present case is a large mass measuring 10.5 × 7 × 6.5 cm, a size which unavoidable

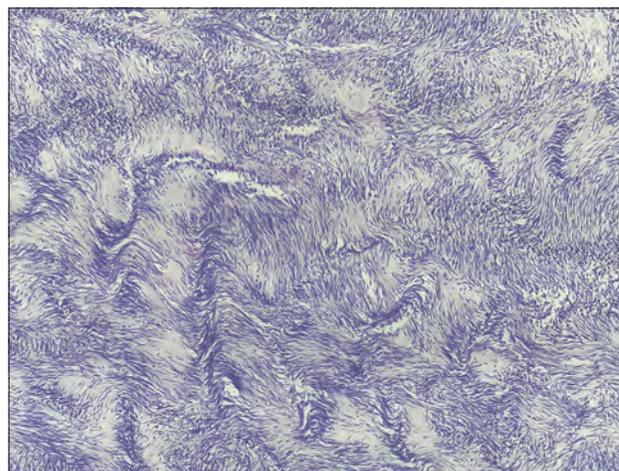


Figure 4: Schwannoma of the vulva: Microscope section of the tumor mass showing the serpentine nuclei arranged in palisades with thin eosinophilic fibrils running across the cleft between the fibrils. X10 objective magnification, H and E

Table 1: Review of the literature on vulva schwannoma

Authors/Year	Age	Subtype	Recurrence	Swelling duration	Location	Size (cm)	Degenerative changes
Huang <i>et al.</i> , 1983 ^[6]	84	Conventional	NR	-	Clitoris	-	Nil
Woodruff <i>et al.</i> , 1983 ^[17]	26	Plexiform	2	-	Vulva	-	Nil
Yamashita <i>et al.</i> , 1996 ^[20]	29	Conventional	NR	-	Vulva	-	Nil
Hanafy <i>et al.</i> , 1997 ^[21]	59	UK	NR	-	Vulva	-	Nil
Quesada <i>et al.</i> , 1998 ^[22]	68	Conventional	NR	-	Vulva	-	Nil
Santos <i>et al.</i> , 2001 ^[23]	5	Plexiform	2	-	Labia minora	-	Nil
Llaneza <i>et al.</i> , 2002 ^[24]	64	Conventional	NR	-	Vulva	-	Nil
Fujimoto <i>et al.</i> , 2004 ^[25]	22	Conventional	UK	-	Vulva	-	Nil
Agaram <i>et al.</i> , 2005 ^[26]	26	Plexiform	NR	-	Vulva	-	Nil
Chuang <i>et al.</i> , 2007 ^[7]	41	Plexiform	NR	-	Clitoris	2×2	Nil
Yegane <i>et al.</i> , 2008 ^[8]	6	Plexiform	NR	6 years	Clitoris	4.5×2.5	Nil
Das <i>et al.</i> , 2008 ^[9]	48	Conventional	NR	-	Vulva	-	Nil
Fong <i>et al.</i> , 2009 ^[10]	53	Ancient	NR	-	Vulva	-	Nil
Yu <i>et al.</i> , 2011 ^[11]	76	Conventional	NR	Not stated	Clitoris	2×1.7×1.5	Nil
Kim <i>et al.</i> , 2011 ^[1]	43	Conventional	NR	2 weeks	Labium minus	0.6×0.5×0.4	No necrosis
Azurah <i>et al.</i> , 2013 ^[12]	6	Plexiform	-	4 years	Clitoris	3×2	Nil
Bozkurt <i>et al.</i> , 2013 ^[13]	65	conventional	-	10 year	Vulva	15×12	Necrosis
Park <i>et al.</i> , 2015 ^[14]	37	conventional	-	3 years	Clitoris	NR	Nil
Bafna <i>et al.</i> , 2016 ^[15]	26	conventional	-	5 months	Labium Minus	1×1 & 1.5×2	No necrosis
Panwar <i>et al.</i> , 2017 ^[16]	48	conventional	-	1 year	Labium Majora	18×16×15	Necrosis
An <i>et al.</i> , 2017 ^[18]	48	Cellular	-	1 year	Vaginal Wall	3.2×0.2	Nil
Tan <i>et al.</i> , 2018 ^[19]	58	conventional	-	10 year	Labium Majus	1×1×1	Cys
Present study	18	conventional	NR	2 years	Labium Majus	10.5×7×6.5	Nil

UK: Unknown; NR: No recurrence

posed movement and other challenges to the patient. An accelerated growth of a schwannoma during pregnancy has been reported in the literature.^[29] The rapid growth and exaggeration of the features of the tumor rarely reported in some cases may be directly due to the physiological changes of pregnancy or pressure effects.^[30-32] Schwannomas may grow into mammoth-sized masses. There was a report of a huge schwannoma incidentally discovered during a caesarean and removed.^[33] It was a huge mass of 40 × 40 cm weighing 6,630 grams, attached by a stalk to the wall of the vagina and extended up into the abdominal cavity.^[33] That case is somewhat similar to the present one herein presented considering the large size of the tumor mass and pregnancy of 35 weeks' gestational age.

Degenerative changes in a schwannoma seem to be related to the size of the tumor. The larger masses may be associated with necrosis or calcification. Sometimes, cystic degeneration may occur. There may be significant deposition of hyaline around blood vessels making the walls appear thickened on microscopy. There was no cystic change, necrosis, or calcification observed in the present case. Huge masses are almost invariably associated with necrosis.^[16]

A high frequency of mitotic cells is only rarely reported in schwannomas cases because the tumor hardly ever becomes malignant.^[34] Most cases of benign vulva schwannoma have low mitosis ranging from none to 3–4 per hpf.^[14,16,18] Malignant schwannomas are associated with frequent mitosis and ultrastructural features such as indented nuclei and absence of Luse bodies.^[35] A mitotic rate of 2/10 hpf was observed in the present case being reported. A Ki67 index of 5% and mitotic rate of 3–4/10 hpf was reported by some researcher who presented a case of pleomorphic schwannoma of the vulva.^[16] Similar to previous reports, this present case demonstrated strong S-100 immunohistochemical reactivity.

Malignant schwannomas are very infrequent and that of the vulvar is a very rare phenomenon.^[35,36] Metastases usually occur by the hematogenous route.^[36]

The patient in the present case was in a stable condition without any evidence of recurrence of the growth several weeks after excision. This is congruent with other reported cases of vulvar schwannomas or those occurring within the pelvic region. Recurrence has occurred only in few instances of reported cases [Table 1]. There seems to be a stable pattern of nonrecurrence once the neoplasm is completely excised.^[1,16]

Conclusion

We have presented a case of a pregnant woman with a large ulcerated vulvar mass of long duration which turned out, on

histological evaluation, to be a conventional schwannoma. This case contributes to the pool of few worldwide reports of this large neural tumor arising in the vulva.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that her names and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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