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Case Report

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Bilateral Giant Mysterious Vulva Tumor in an Adolescent: A Rare Clinical Condition and its Diagnostic Challenges in Resource Constrained Facilities

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Abstract: Vulva tumors are generally rare disease entity encountered in clinical practice of gynecology, and it is especially so at a young age. The spectrum of tumors arising in the vulva may be attributable to inflammatory conditions, infections, dermatological conditions, vulva intraepithelial neoplasia and invasive cancer; thus many specialties in the medical field may be consulted for diagnosis and treatment. Definitive diagnosis requires biopsy (incisional or excision). Other tests may be ordered as supportive or to diagnose infectious or other systemic causes of Vulva swellings. Presented here is a case report and diagnostic challenges of a mysterious vulvar swelling in a 20 years old woman who presented to our Hospital with painless progressively enlarging bilateral vulvar tumor which evolved over a period of four years. **Keywords:** Bilateral, Vulvar tumor, Adolescent.

INTRODUCTION

Tumors of the vulva are rare, few published cases have reported about this condition [1]. Apart from malignant tumors, fibromas fall in the group of the most common benign tumor of vulva though they as well occur very rarely. [2]. Other causes vulvar swellings like infections particularly elephantiasis will have to be ruled out and it is especially so in the tropics where filarial worms' infections are common [3]. When encountered in younger patients, vulvar tumors are more likely to be benign than malignant [4].

Benign growths of the vulva may be composed of any of the tissues that make up the vulvar, ranging from the epithelium to the underlying stroma [3]. The commonest benign tumors of the vulva include; the fibroma, papilloma, lipoma, angioma, neuromas, lymphangiomas, neurofibromas, adenomas, just to mention a few [5]. Of these, fibromas are the most common form of benign masses in young women. When located in the vulva, they occur more often in the labia majora and less frequently they may be found in the labia minora, clitoris, vestibule, and posterior commissure [6]. In tumors of long clinical duration, ulcerations associated with superficial bleeding, often due to repeated trauma or friction, are often observed [6].

Although they may be sessile in the beginning, vulvar tumors show a strong tendency to develop a stalk and become pedunculated and the dependent part

becomes congested, oedematous and ultimately ulcerates and may eventually become infected. Because the presentation may mimic vulvar cancer growths, a biopsy needs to be taken for histological examination to exclude malignancy [7].

Treatment of vulvar tumors (whether the diagnosis is confirmed or is in doubt) is by excision [2]. Generally, vulvar tumors produce non-specific symptoms at initial stages like pruritus and irritation. These minor symptoms may initially be ignored by women, contributing to a delay in diagnosis. Diagnostic delays may compel clinicians to initiate treatment for other conditions such as dermatoses prior to establishing a histological diagnosis. Early detection and biopsy of any vulvar tumor are imperative to exclude vulvar cancer in its early stages [7].

Published material regarding vulvar tumors is scattered throughout the medical literature of various specialties, including dermatology, genitourinary medicine, gynecology, and pathology [8]. The spectrum of involvement of different specialties reflects the complexity of vulvar diseases and the necessity of a multidisciplinary approach to the study and management of vulva diseases.

This case report describes a unique case of huge, mysterious bilateral vulvar tumor in a 20 year old female that had evolved spontaneously over a period of 4 years (since when she was 16 years of age).

CASE REPORT

A 20 year old nulliparous girl presented in the gynecological outpatient Department of our hospital as a referral client from a rural Health Centre in Bahi District Dodoma Region, in Central Tanzania with a complaint of progressively painless bilateral vulvar swellings which started four years prior to seeking medical attention as small itching rashes. The swellings had progressively been increasing in size over a period of four years. For a long period of time the swellings had been painless, soft to firm in consistency (at different sites of the mass) and later became a pedunculated and ulcerated at the dependent part in one side. The patient had not yet been sexually active when the swelling started though she had attained menarche two years earlier (before the illness began) and her menstrual cycles were reported to be regular ever since. She reported minor contact bleeding from the ulcer, difficulty in walking and heaviness in the vulvar region. On general examination, the girl was found to be generally healthy looking and appropriate for age in terms of height and secondary sex characteristics development. Pelvic inspection and palpation revealed bilateral giant pendunculated vulvar swellings with ulceration on the surface on the larger mass (Fig. 1). Speculum examination (though was done with difficulty) revealed nothing out of the ordinary. No lymph nodes were palpable in the inguinal regions. She was admitted to the gynecological ward with a provisional diagnosis of benign vulvar fibroma to

exclude malignancy of the vulva. Vulvar elephantiasis was also earmarked as an important differential diagnosis.

While in the ward, VDRL for syphilis testing and a blood smear for filarial worms were all negative. Other blood tests were also normal. A biopsy was taken from one side of the lesion and pathologists reported: marked epithelial and dermal inflammation, no evidence of neoplasia (Fig. 2). Despite the histological results, the diagnosis of vulvar fibroma was compelling enough to maintain it as a diagnosis pending excision of the whole tumor for further pathological testing.

The final decision was reached to proceed with excision of the tumor and the patient consented for the procedure and without duress, she agreed to be photographed and her case to be published. Prior to the procedure (wide excision), the patient requested to go back home for further preparation and would be back in two weeks for the operation. But it has been three months since we saw her and she has not turned in for the operation and it makes us have the feeling that she may be a lost case.

Ethical issues

Informed consent was obtained from the patient to take photographs and publish the case. The permission to publish patient information was obtained from Hospital Management Team (HMT).



Fig. 1: Showing the shape and extent of the tumor

DO Ro	n 15 Dodoma TZ	Histopathology Report Hosp Reg No: 17-22-52
Cell No. 0784-533513		Surnam
Hospital:	Dodoma Regional Referral	Other N
Doctor: She	emdoua	Postal Residential Bahi
Specimen of: labia majora		Date of Birth Age
Clinical Histor	 Progressive painful swelling of vulva with regions of ulceration. DDx: Vulvar malignancy. 	CHI SANANA GOPD
		Log in No: MHP-0534-14
Date of Reques Report: Spec 1.5x	st 5-3-14 Date Received 5 imen consists of an elipse of thir 1.5x0.6cm. MT,AT,1C.	grey skin 2x1cm. This is over white tissue
Diagnosis:	Marked epithelial and derm [See Comment], bioppsy of	al inflammation, no evidence of neoplas labia majora.
Date Reported	6-3-14	Reported by:
Comment: Due t	a plasma cell infilm the	ascularity syphillus cannot be rulled out.

Fig. 2: Histology report

DISCUSSION

Tumors arising in the vulva commonly begin as small masses that do not normally attract medical attention, but occasionally, they can attain, huge sizes that can impair victim's lifestyle [2] and may in fact affect her sexual and reproductive functions. They appear either as subcutaneous nodules or pedunculated (due to gravitational effects), sometimes superficially ulcerated masses in the dependent part of the tumor [6]. In the present case report, the patient provided information that it started as small itching rashes on the labia that were at first ignored thinking it would spontaneously resolve. This case calls for a careful medical evaluation for every nodule in the vulva and be should be given a deeper thoughtful look before a patient is reassured to be a non-life threatening condition [2] as they have a potential of growing into 'huge and difficult to manage' tumors.

Most often vulva tumors originate in labia majora, but some large benign tumors have been found on the clitoris [9]. They may also originate from smooth or striated muscle (leiomyoma or rhabdomyoma), fibrous tissue (fibroma), fat (lipoma), or they may have vascular (haemangioma), lymphatic (lymphangioma) or neural origin (neurolemoma) [10]. Our patient the tumor emerged from the commonest site (The labia majora).

Tumors of the Vulva, although relatively uncommon, may have a tremendous impact on someone's life ranging from to an extent of social withdrawal to marriage failure or divorce [11] and are often initially referred to dermatologists for evaluation and treatment for the first time in medical consultations because they commonly begin as dermatological conditions. The clinical features of benign tumors may overlap with malignant neoplasms, and therefore, a biopsy is often necessary to make a definitive diagnosis [10]. In the present case the size and the ulceration on the top of the tumor would be, at a quick look point to the possibility of a malignant tumor.

Benign tumors of the vulva may arise from both epithelium and underlying stroma that make up the vulva. Epithelial tumors comprise of a group of common skin changes, which range from small to rather large formations and sometimes may resemble malignant lesions [6] that is why the histological diagnosis is of paramount importance in reaching the definitive diagnosis of vulvar tumors.

As pointed out earlier on, tumors of the vulva are generally rare; few published cases have reported this condition [1]. The rarity of these tumors presents a significant challenge to detailed understanding of their pathological, morphological and epidemiological characteristics, with only short descriptive texts and citations in the literature available [11-13]. This means the diagnosis may easily be missed and ignored, particularly in the early stages of the disease.

Though in this case the initial incisional biopsy histological diagnosis was pointing to simply inflammatory changes and the blood smear for filarial worm was negative, clinical features, and gross structure points more to a vulvar fibroma as supported by the age of the patient and available pictures in literatures [1, 2]. Similarly, other scientists had found that the initial incisional biopsy revealing the vulvar tumor to be a benign smooth muscle hematoma, but after excision of the whole tumor mass, the diagnosis was found to be a fibroma [14]. This implies that the initial incisional biopsy may not reveal the true pathology of the tumor in some cases and therefore a second thought and a high index of suspicion is squarely important.

Fibromas of the vulva are usually asymptomatic in the beginning; however, they develop symptoms as the size of the tumor increases. Associated complications like superficial ulceration present much more obvious alarming clinical signs. Because many new growths in the vulva may present in a similar way, it's particularly important to be aware of the possible differential diagnosis in a patient suspected to have a fibroma. Differential diagnoses include lipoma, inguinal hernia, vulvo-vaginal cyst, and other benign tumors of the vulva [3, 15]. In our case some possible differentials were excluded by the blood smear, clinical appearance and the initial histological findings. Excision would provide a final diagnosis but unfortunately the case was lost. Lost case are common finding in low income countries where patient demographic information are not readily available and therefore it present an enormous challenge to doctors who take care of patient in these settings.

In general fibromas occur more frequently in the eyelids, neck, axillae, submammary regions, and inguino-crural folds, and are unusual in the vulva [16] and this justifies its reporting to expand our horizon of understanding on the possibility of a fibroma to occur in the vulva. Other names for fibroma are connectivetissue benign tumors that have multiple synonyms (naevusmolluscum, acrochordon, Templeton skin tag, and fibroepithelial polyp [9].

Fibromas of the vulva are commonly benign tumors that in some cases, due to their macroscopic appearance, can lead to a mistaken diagnosis of malignancy [9]. Thus histology particularly after excision biopsy forms a cornerstone for the definitive diagnosis. These tumors are predominantly found in women of reproductive age group, though they have been reported in infants, post-menopausal and pregnant women [17].

In the beginning, vulvar fibromas may be asymptomatic but has the potential to grow to huge sizes that may impair sexual function as well as other vital roles of the vulva and the perineum. Giant vulval fibromas like this are quite uncommon and there are very few reported cases in literature [18, 19]. Despite being benign in nature, fibromas may be a cause of dyspareunia, difficulty in walking emotional and psychological disturbances and later patients usually complain of a swelling or bleeding and discharge from the ulcer. Large tumors can as well cause coital difficulties, walking discomfort and complicate labor if the affected young woman is pregnant [2]. While fibromas belong to the most common solid tumors of the vulva while their cause remains unknown [20]. The etiology of these tumors remains unknown [21]. Though excision is the mainstay of treatment, Vulval fibroma may recur after incomplete removal [14, 22]. Our case missed the opportunity for excision which presents an enormous challenge to doctors in resource constrained facilities in establishing the definitive diagnosis of rare and mysterious tumors.

As stated earlier, vulvar pathology is a concern of several specialties, which reflect the complex morphology, and the variety of functions through the life cycle of women. The vulva is the region where numerous local or systemic diseases can be found. The spectrum of abnormalities that can affect the vulva ranges from infections, inflammatory conditions and dermatoses, similar to those encountered in extragenital skin, to vulvar intraepithelial neoplasia and invasive cancer [23].

Treatment of vulvar tumor is usually total excision of the tumor [2]. Recurrence of excision is likely, particularly when it was incompletely removed [14, 22].

CONCLUSION

Vulva tumors are uncommon and much more so in younger patients and the diagnosis at times is challenging and requires a high index of suspicion because initial incisional biopsy findings may be misleading. Furthermore, because of their location, size and its associated complications they can cause severe disturbing symptoms which may be a source of social withdrawal and even divorce due to impairment of sexual and reproductive functions and therefore early diagnosis and treatment is imperative.

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