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Case Report

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VULVAR TUMOR - CASE REPORT AND LITERATURE REVIEW

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Keywords

Vulvar Tumor; Pregnancy; Fibroepithelial polyps; Carcinoma

Introduction

The vulvar region is a complex area because it comprises many elements, besides the skin itself. Therefore, it can present a variety of relatively rare tumors that can be classified based on source tissue (epithelial or mesenchymal). Benign epithelial tumors in the connective tissue are not often diagnosed in the vulvar area, which is mostly affected by fibroepithelial polyps [1].

Fibroepithelial polyps mainly affect obese and diabetic women. However, assumingly, hormonal disorders can enable the development of such tumors, which have no association with human papillomavirus and preferably affect skinfold areas such as armpits and neck. In macroscopic terms, fibroepithelial polyps can appear as a grayish lesion, as a condyloma-like pigmented papillomatous growth, or as a pedicular and hypopigmented tumor. They are often asymptomatic and diagnosed by patients themselves. Their clinical manifestations are associated with skin rashes or ulcers (the large ones) in obese women [1-3]. Other benign tumors, such as leiomyomas, can also affect the vulvar region. In addition, there are reports of granulosa cell tumors that originated from peripheral nerves affecting this region [4,5].

Vulvar cancer is a rare neoplasm that accounts for less than 1% of malignant tumors affecting women, as well as for 3% to 5% of female genital tract malignancies. Squamous cell carcinoma, which accounts for approximately 85% of all cancer cases, is the most often diagnosed histological type of tumor; it is followed by melanoma. It is necessary to emphasize that malignant vulvar neoplasms are associated with HPV in younger women, whereas inflammatory dermatoses, such as scleroatrophic lichen, are mostly diagnosed in older women [6]. In this case, itching is often a symptom.

Since such tumors are rare, it is of paramount

importance discussing about clinical aspects, differential diagnosis and management procedures to help improving routine medical practices.

Case Report

Patient AFA appeared at the Gynecology Service of the Medical School of Barbacena - Santa Cecília Health Center - complaining about a vulvar tumor identified 3 years earlier. She reported having already sought medical attention in another healthcare service, which failed to solve her problem. She observed progressive tumor growth during this period, a fact that prevented her from performing her daily, even sexual, activities.

She was a 52-year-old hypertensive and diabetic patient, G3P3A0, who used amlodipine and 1g of metformin hydrochloride/day. Her menarche happened when she was 10 years old and already presented irregular cycles. At examination time, she was flushed, hydrated, anicteric and acyanotic; her BP was 120/80 mmHg, her breasts did not present palpable nodules and her abdomen was flaccid, globose and painless at superficial and deep palpation. The patient's vulva was trophic, presented *gynecoid hair distribution*, as well as a large tumor (dimension of approximately 7 cm) in the right labia majora; the tumor was hypopigmented and presented long pedicle, whose surface showed exuberant vascularization (**Fig-1A,B**). The palpation of bilateral inguinal lymph nodes did not show abnormalities. Vaginal or cervical lesions were not identified. Cervical samples were collected for colpocytological examination; the tumor was removed with reasonable safety margin (Figures 1A, 1B) and sent for histopathological examination, whose initial diagnosis was compatible with a fibroepithelial polyp. The material was subjected to immunohistochemical analysis, which recorded positive results for the following markers: AE1 / AE3 (in the epithelium), smooth muscle actin - SMA (in the muscle wall), CD34 (in the vascular endothelium), desmin (in the vascular wall), EMA (in the epithelium), Ki67 (in 2% of cells), estrogen and progesterone receptors (in the stroma). The other markers (S100, p16, and myogenin) recorded negative results. Data were compatible with fibroepithelial polyp in association with stromal edema and vascular malformations. However, correlation between clinical and radiological data was suggested to rule out the likelihood of unidentified aggressive residual lesion in the specimen, since the major concern was to rule out aggressive angiomyxoma of the vulva.

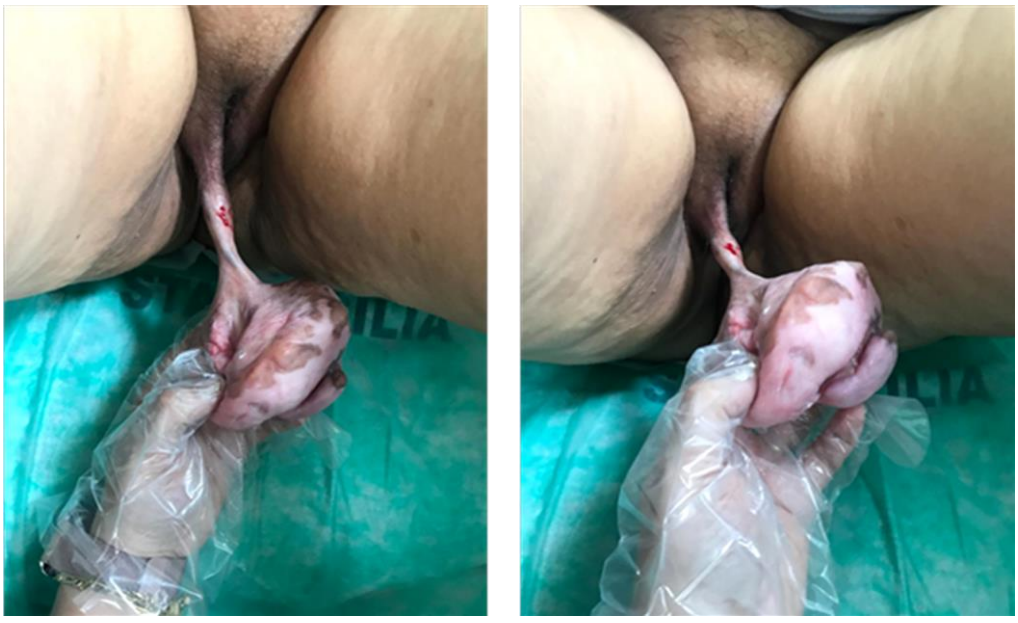

Fig-1(A, B):

Large tumor (dimension of approximately 7 cm) identified in the right labia majora; the tumor is hypopigmented and presents long pedicle, whose surface shows exuberant vascularization.

The patient returned to the outpatient clinic 7, 14 and 30 days after the procedure; she presented satisfactory evolution with no complications and good healing (**Fig-2A**: 7 days after the procedure; **Fig-2B**: 30 days after the procedure). However, she noticed the elimination of serous secretion from a small hole at the exeresis site, 60 days after the procedure. After a new biopsy was performed, the sample was subjected to histopathological analysis, whose result did not identify a residual tumor or atypical material (**Fig-2C**); it only identified the healing process.

Total abdominal and transvaginal ultrasound examinations were carried out to rule out any tumor-associated perineal or pelvic involvement; results did not show expansive tumors, the patient presented uterine dimensions 5.2 x 1.9 x 2.8 cm, centered and regular endometrium (2 mm), and normal volume in the ovaries. The abdominal ultrasound did not identify abdominal masses, except for diffusely increased hepatic echogenicity; the patient was referred to gastroenterology control, without any association with the clinical case.

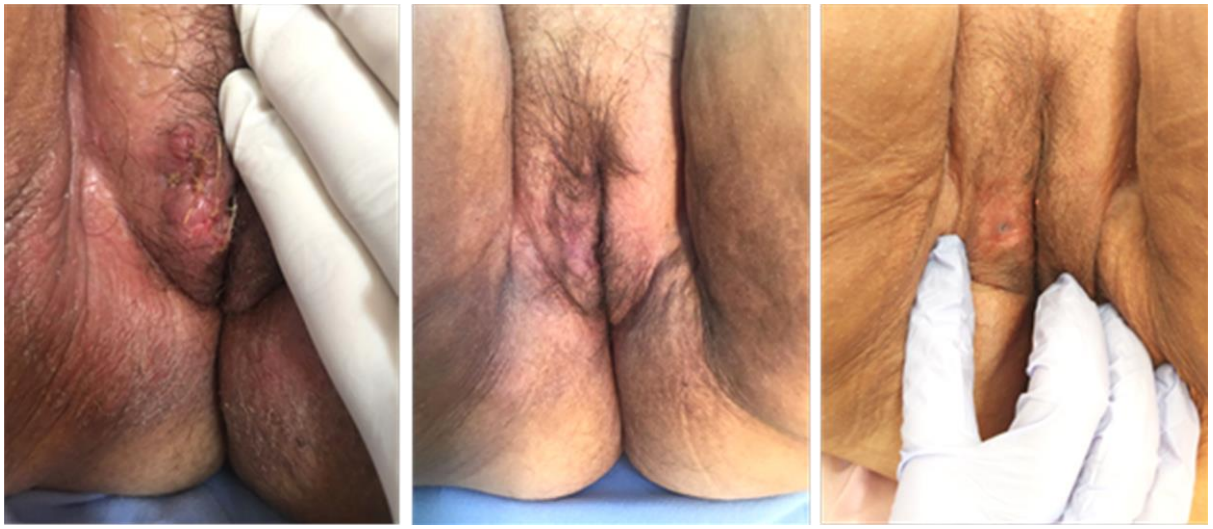

Fig-2:

Control after vulvar tumor excision. Control was performed 7 (2A), 30 (2B) and 60 (3C) days after tumor excision, when a small serous-drainage hole was identified.

Discussion

Vulvar tumors, although rare, can significantly vary depending on the characteristic of the tissue and on the presence of skin. **Table-1** describes the main benign and malignant vulvar tumors [1,7].

The current study reported the case of a patient whose tumor was diagnosed as a long-lasting, large-sized vulvar fibroepithelial polyp. She was a climacteric patient, who already presented irregular menstrual cycles and was not taking hormonal medication. These tumors have been described in menace, and even during pregnancy, when women present high circulating estrogen and progesterone level [8]. Importantly, the patient in the herein reported case was obese and diabetic; her clinical data are often associated with these conditions [8].

The tumor was pediculate and presented discoloration points at analysis time; histopathological analysis result was compatible with a polyp. However, immunohistochemical analysis was recommended. The major initial concern was to rule out aggressive angiomyxoma of the vulva, which may also be pediculate and present long-term evolution. It is more often diagnosed in women, who have worse prognosis due to the possibility of having the tumor invading the pelvis and the perineum, a fact that can lead to a series of severe complications such as compression of pelvic organs. Therefore, complementary pelvic and abdominal evaluation was performed; results came out normal [9,10].

The excision of the lesion at a considerable safety

margin was the recommended treatment. The biopsy performed 60 days after the initial lesion excision did not identify residual tumor, it only identified the healing process [11]. Although fibroepithelial polyp is a benign tumor, local recurrence has been reported; therefore, the patient in the current case report remains under clinical follow-up and no local recurrence has been identified, so far [11].

Table-1: Classification of benign and malignant vulvar tumors based on their origin. Adapted from [1,7].

Tumor type	Origin	Examples	
Benign	Epithelial	Keratinocytic	Fibroepithelial polyp
			Seborrheic Keratosis
			Keratoacanthoma
		Attached	Lymphangioma
		Melanocytic	Nevus
		Ectopic	
	Mesenchymal	Vascular	Hemangiomas, Granulomas
		Fibrous	Fibromas
		Muscular	Leiomyoma
		Neural	Neurofibroma
		Miscellaneous	Lipoma
Malignant	Epithelial	Squamous cell carcinoma	
		Adenocarcinoma	
		Basal cell carcinoma	
	Mesenchymal	Leiomyosarcoma	
		Rhabdomyosarcoma	
	Melanocytic	Malignant melanoma	
	Other tumors	Yolk sac tumors	
		Merkel Cell Tumors	

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