ISSN- O: 2458 - 868X, ISSN –P: 2458 - 8687

International Journal of Medical Science and Innovative Research (IJMSIR)

IJMSIR : A Medical Publication Hub

Available Online at: www.ijmsir.com

Volume – 3, Issue –1, January - 2018, Page No. : 211 - 213

Vulvar leiomyoma - a rare condition and a challenging diagnosis

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Type of publication: Original Research Paper

Conflicts of Interest: Nil

Abstract

Leiomyomas are benign tumours originated from smooth muscle cells. Leyomiomas of the external genitalia are uncommon with fewer than 160 cases of smooth muscle tumors of the vulva having been reported in the literature. The differential diagnosis includes benign and malignant entities such as Bartholin cysts and soft-tissue sarcomas, so surgical excision and histological diagnosis are mandatory. Despite being benign and associated with a low recurrence rate, long term follow-up of vulvar leiomyoma is advisable.

The authors describe a case report of a 60-year-old woman that presented with a mass on the right labia majora. Histological exam after surgical excision concluded it was vulvar leiomyoma.

Keywords: leiomyoma, vulva, Bartholin cysts.

Leiomyomas are benign tumours originated from smooth muscle cells. The most common site for their occurrence is the myometrium, although they can appear in any site where smooth muscle is present [1]. Leyomiomas of the external genitalia are uncommon with fewer than 160 cases of smooth muscle tumors of the vulva reported [1, 2]. Leyomioma of the vulva represents only 0.03% of all gynecologic neoplasms [3]. It is thought to originate from smooth muscle within erectile tissue, round ligament, blood vessel walls, erector pili muscle or the dartos muscle [2, 4, 5]. These tumors are well delimited, solitary and do not present with pain [2].

The differential diagnosis includes benign and malignant entities such as Bartholin cysts, fibromas, soft-tissue sarcomas, lymphangiomas and neurogenic tumors, representing an important diagnostic challenge [4]. Most frequently they are misdiagnosed as Bartholin cyst [1, 4]. They can affect female of any age group but mostly between 30 and 60 years old [2]. Typical vulvar leiomyomas demonstrate spindle shaped cells [2] and immunohistochemistry plays an important role in differential diagnoses: they are positive for actin, vimentin, desmin and anti-smooth muscle antibody; estrogen and progesterone receptors are positive in some of the cases [6]. The only available curative treatment for vulvar leiomyomas is surgical excision. Although recurrence is extremely rare, long-term follow-up of these cases is advised [7].

Case Report

The authors describe the clinical case of a 60-year-old woman with leiomyoma of the vulva. She went to the emergency department because of a painless nodule on the right labia majora which had appeared 3 weeks before (Figure 1). She had no relevant personal history. The first diagnostic hypothesis was a Bartholin duct cyst but for better clarification of the lesion an ultrasonography of the

soft tissues was requested. It revealed a solid, markedly heterogeneous nodular lesion, measuring about 6 cm on the largest axis, with apparently good contours defined. In order to exclude a Bartholin gland neoplasm, histological characterization was recommended. She was proposed for surgical resection of the lesion which revealed a vulvar nodule with about 7 cm of larger diameter with well-defined walls (Figure 2). Anatomopathological exam revealed a well delimited neoplasm composed of spindle cells with elongated nuclei, fine chromatin and eosinophilic cytoplasm, arranged in bundles or fascicles in the sinus of scarce stroma; no atypia, no cell pleomorphisms, mitotic figures or foci of necrosis were identified; the immunohistochemical study revealed positivity for desmin and smooth muscle actin; compatible with leiomyoma of the vulva. Postoperative period was uneventful and the patient was referred to the Gynaecology consultation. One month after surgery, the patient presented no complaints and had a well-healed vulva (Figure 3). The patient was reassessed 6 months later, remaining asymptomatic and without signs of relapse; she will remain under annual surveillance.

Discussion

The case we present is uncommon because leyomiomas of the external genitalia are a rare entity. They tipically arise in women during fertile age [4]. Clinically, they present as a non-painful nodule in the vulva with an average tumor size varying from 0,5 to 15 cm [4] and usually mimic a Bartholin gland cyst [1, 2, 7]. Differential diagnosis with benign and malignant entities is mandatory: despite being rare, vulvar leiomyosarcomas appear in the same location as Bartholin duct cysts and the incorrect initial clinical diagnosis such as Bartholin's duct cyst may delay the diagnosis of a malignant tumor and worsen the prognosis [7]. Best imaging exams for diagnosis of extrauterine leiomyomas are ultrasonography, computed tomography, and magnetic resonance imaging [4]. Surgical ressection of the tumour is mandatory because definitive diagnosis is histological [8]: vulvar leiomyomas have spindled cells with estrogen and progesterone receptor positivity and immunohistochemical staining shows positivity for smooth muscle markers including desmin, and actin [7]. Long term follow-up of all cases is advisable, although recurrence is rare [4].

Conclusion

To summarize, vulvar leiomyomas are extremely rare, usually affect women during fertile age and its importance comes from the need for differential diagnosis with malignant tumours. Surgical excision is the only curative treatment for vulvar leiomyomas. Long-term follow-up of all cases is advisable.

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Acknowledgements

The authors thank the Hospital of Santa Luzia, where they carry out their professional activity, namely the Department of Gynecology and Obstetrics and Pathologic Anatomy.

Declaration of Conflict Of Interest:

There are no conflicts of interest.

Funding: No funding sources

Figures:

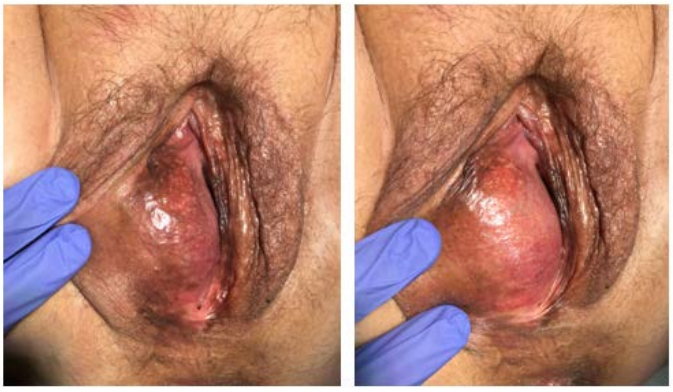

Figure 1: Nodule on the right labia majora.

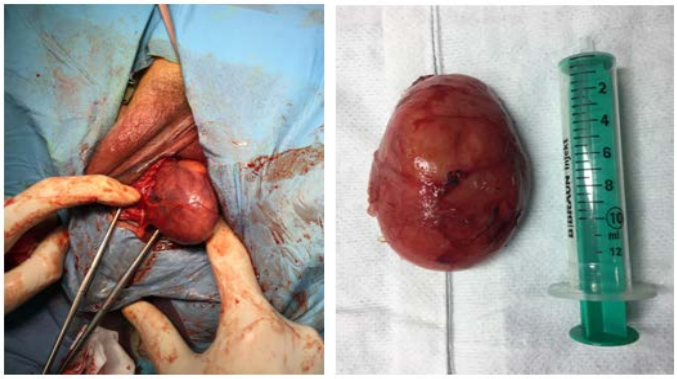

Figure 2: Surgical resection of a vulvar nodule with well-defined walls

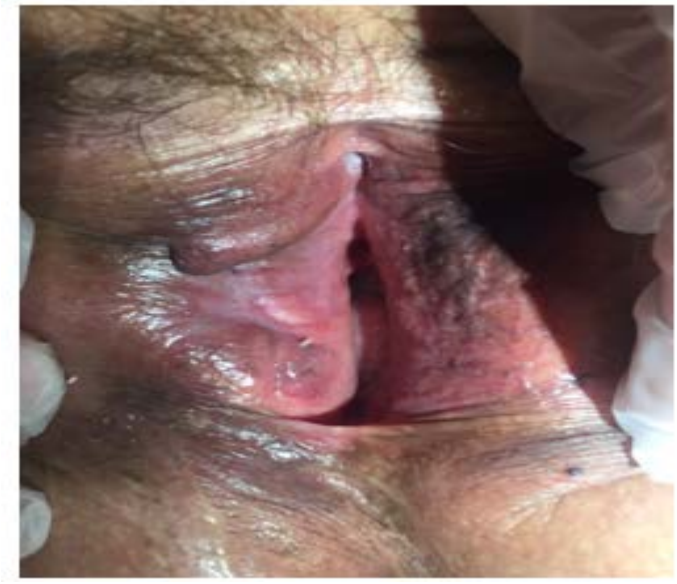

Figure 3: Postoperative Gynaecology consultation showing a well-healed vulva