Case Report ISSN 2639-944X

Journal of Medical - Clinical Research & Reviews

Vulvar Schwannoma: A Case Report in an Elderly Patient

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Received: 05 January 2019; **Accepted:** 28 January 2019

Citation: Andrade Ana Carolina de Almeida, Queiroz Polyanna Mendonça de, Chambô Filho Antônio. Vulvar Schwannoma: A Case Report in an Elderly Patient. J Med - Clin Res & Rev. 2019; 3(1): 1-3.

ABSTRACT

Statement of the problem: *Schwannomas are benign tumors of the peripheral nervous system, originating, more specifically, from Schwann cells. They present in the form of a single nodular lesion and are rarely found on the female genital organs. Schwannomas are generally benign, seldom becoming malignant. When present in the vulva, the incidence is higher in patients of 20 to 40 years of age.*

Case Presentation: *This report describes the case of a 73-year old woman with a single tumorous lesion on her vulva. The lesion was mobile, non-inflammatory and practically painless. Surgical resection was followed by investigation by histopathology and immunohistochemistry, with findings confirming a diagnosis of schwannoma.*

Conclusion & Significance: *Identifying the characteristics of schwannomas is of crucial importance when investigating differential diagnoses of vulvar pathologies. Clinical history and a thorough physical examination help ensure adequate management and correct diagnosis of the condition, with the definitive diagnosis being obtained following histopathology and immunohistochemistry.*

Keywords

Schwannoma, Vulva, Vulvar tumor.

Introduction

Schwannomas generally present as a single, benign, nodular tumor of the peripheral nervous system that originates in the Schwann cells. These lesions are rarely situated in the female genital organs, and reports of such cases in the literature are rare [1-3]. The most common sites of presentation are the head and neck, the flexor surfaces of the limbs and the posterior mediastinum. Irrespective of their location, less than 1% of schwannomas will become malignant. When this type of lesion presents in the vulva, the age group most usually affected is women of 20 to 40 years of age [1-4].

The present paper reports on the case of a 73-year old woman seen at the Department of Gynecology at the Santa Casa de Misericórdia Hospital in Vitória, Espírito Santo, Brazil. The reason for her visit was the appearance of a slow growing mass on her vulva. Following surgical resection, histopathology and immunohistochemistry confirmed a diagnosis of vulvar schwannoma.

The publication of this case report was approved by the institution's internal review board under reference number 3.075.073. The patient gave her written consent.

Case Report

A 73-year old, brown-skinned Brazilian housewife presented at the Gynecology Department with no signs of neurofibromatosis or any other comorbidities and no family history of cancer. An initial examination revealed a single, palpable and almost painless tumor on the vulva that had been slowly increasing in size over the preceding year. The patient was in good general health, reported no significant weight loss during the period in which the lesion had been present, and had no other associated complaints. Gynecological examination confirmed the presence of a solid, well-circumscribed nodule of approximately 4 cm in diameter on the patient's right labium majus. The lesion was mobile, almost painless and not adhering to deep planes. There were no signs of inflammation, dyschromia or any other anatomical abnormalities of the vulva, vagina, cervix or adjacent lymph nodes (Figure 1).

In view of the findings at anamnesis and gynecological examination,

the decision was made to surgically remove the lesion to enable a definitive diagnosis to be reached. Following subarachnoid block, the tumor was completely resected, taking care to ensure good safety margins. The resected material was sent for histopathology.

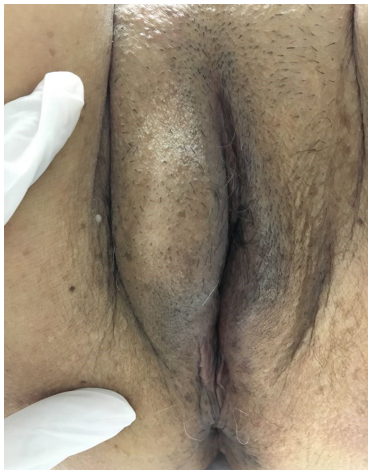

Figure 1: The vulvar lesion prior to surgical resection.

Macroscopy revealed an encapsulated, brown-colored lesion with blackened areas (Figure 2A), while microscopy showed an encapsulated, nodular, spindle cell, mesenchymal tissue tumor, with different areas composed of different cellular densities, with some palisade arrangements (Verocay bodies). Hydropic, hemorrhagic and cystic degeneration was found, with foci of necrosis measuring approximately 3.5 cm. The most striking characteristics included the finding of few mitotic figures and discrete focal pleomorphism (Figure 2B). Immunohistochemistry revealed strong positivity for the S-100 protein, differentiating this type of tumor from neurofibromatosis and supporting the hypothesis of schwannoma.

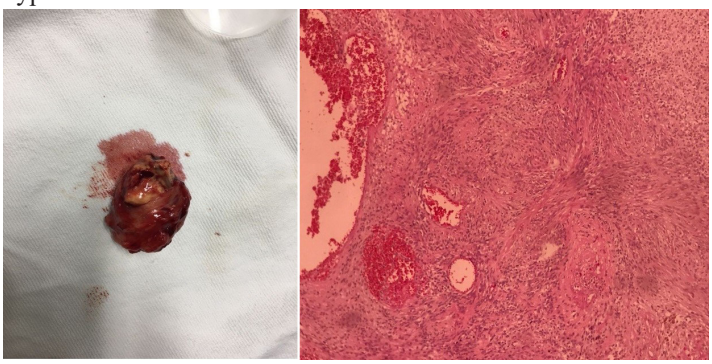

Figure 2A: Appearance of the surgical specimen.
Figure 2B: Histological appearance of the lesion.

Discussion

Schwannomas are benign tumors usually located on the head and neck, on the flexor surfaces of the limbs or the posterior mediastinum. They are rarely found in the vulvar area or other female genital organs; therefore, few such cases have been reported in the literature. Descriptions of vulvar schwannomas in elderly women such as the patient in the present report are even rarer, since these tumors are generally diagnosed in young patients of 20 to 40 years of age, as reported by Park et al. and other investigators [1-4]. Schwannomas usually present as single, slow growing, encapsulated nodular tumors. In the present case, the tumor mass had been noticeable for only a year. However, Tan et al. reported a case involving a 58-year old Chinese woman with a vulvar lesion that had progressed over a 10-year period and was eventually diagnosed as a schwannoma. In most cases, these tumors present as a mass that is painless or almost painless, depending on the site of presentation. The likelihood of malignancy is low [4]. They are often associated with neurofibromatosis; nevertheless, these pathologies are not necessarily present in conjunction [1,2].

When the tumor is in the vulvar region, diagnosis based only on clinical characteristics is difficult due to the broad range of possible differential diagnoses such as Bartholin's gland cyst, lipomas, liposarcomas, cysts of the labia minora/majora, mesenchymal tumors, fibrosarcoma, fibroids, angiosarcoma and epidermoid cysts [3]. Ultrasonography can be used to differentiate cystic tumors from solid tumors and to assess whether the tumor is benign or malignant. Nonetheless, the definitive diagnosis is obtained following histopathology and immunohistochemical determination after complete surgical excision of the lesion [3].

Microscopically, the tumor comprises a well-circumscribed, encapsulated lesion consisting of Schwann cells arranged in two distinct patterns: Antoni A and Antoni B tissue patterns. The Antoni A pattern is characterized by areas containing spindle cells with elongated dark nuclei arranged in multidirectional bundles forming a palisade-type effect referred to as Verocay bodies [4-6]. The Antoni B pattern, on the other hand, is characterized by a less compact area with a more haphazardly arranged pattern of oval cells with abundant cytoplasm. The palisade arrangement is notably absent and there may be foci of microcystic degeneration [4-6] (Figure 3).

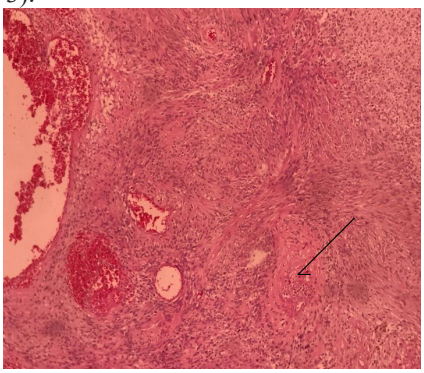

Figure 3: Appearance of the schwannoma at microscopy.

Immunohistochemistry enables tumor cell markers to be identified based on positivity for the S-100 protein, indicating that they originated in the Schwann cells and differentiating this type of lesion from neurofibromatosis [4-6].

Due to the wide range of vulvar pathologies and the similarity between them, identifying the characteristics of schwannomas is of crucial importance when investigating differential diagnoses. Meticulous anamnesis and a thorough physical examination are important steps in managing this pathology, guaranteeing coherent

management and correct diagnosis of the condition.

Acknowledgments

We are grateful to Dr. Paulo Bittencourt de Miranda for his valuable contribution in the pathological analysis and interpretation. We also thank the patient, who promptly agreed with the publication of this case, and the staff of the Department of Obstetrics and Gynecology for their important support during the study.

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