Giant Vulvar Schwannoma: A Case Report

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Abstract- Schwannoma is a solitary, slow growing, benign tumour of the peripheral nerve sheath, and it is most common locations are the head, neck, the flexor surfaces of the extremities, retroperitoneal and posterior mediastinium. External female genital organs are the least common location site of the schwannoma. Only a few cases of vulvar schwannoma have been reported.We report 65 year- old woman presented with a left vulvar swelling, which had been present for several years. The tumor size is 15x12 cm and slowly increased but it prevent patient activities such as walking and sitting. The tumor was resected for treatment and the histological examination confirmed to be a vulvar schwannoma. In our best knowledge, vulvar schwannoma in this case is the largest size in the literature.

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Introduction

Shwannoma is a benign tumor that arises from the Schwann cells of the peripheral , cranial or autonomic nerve sheats. This tumor is usually a solitary and most commonly occur from the head, neck, the flexor surfaces of the extremities, retroperitoneal and posterior mediastinium. It rarely effects the female genitalia. Epithelial tumors such as squamous cell carcinomas are the most common tumor than the mesenchymal tumors such as fibrosarcomas, liposarcomas, angiosarcomas, and schwannomas, located in the vulvar region. According to our best knowledge, only a few cases of vulvar schwannomas have been reported in the literature. In this study, we report a case of benign schwannoma arising in the left vulva and pons pubis.

Case Report

A 65-year-old, gravida 5, para 3, curettage 2, post menopausal woman presented with painless left vulvar swelling which had been present for several years. The tumor size increase slowly but recently it affects daily activities like walking and sitting. 10 years ago, she was a history of umbilical hernia operation. But there was no family history of cancer and hereditary disease.

On gynecologic examination, there was a 12 cm subcutaneous, immobile, non-tender, well-circumscribed mass lesions located on the left vulva and mons pubis (Figure 1). The vulva and vaginal mucosa are atrophic. There was no inguinal lymphadenopathy.

Superficial sonographic examination revealed, 15x12 cm, hypoechoic, oval, solid, encapsulated mass lesion located on the vulva. Doppler sonographic evaluation showed there was no vascularization.

Magnetic Resonance Imaging (MRI) with contrast material showed heterogenous, oval, T1 weighted hypointense, T2 weighted hyperintense, solid lesion located on the subcutaneous tissue of the left vulva with peripheral and internal heterogenous contrast enhancement. Tumor extension being limited to 1/3 outer part of the vagina.

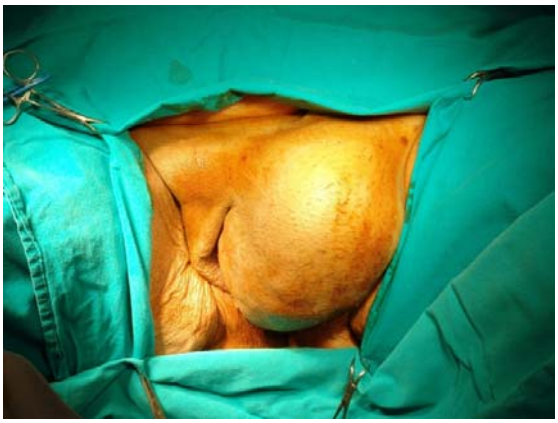

Figure 1. Subcutaneous, immobile, non-tender, well-circumscribed mass lesions located on the left vulva and mons pubis.

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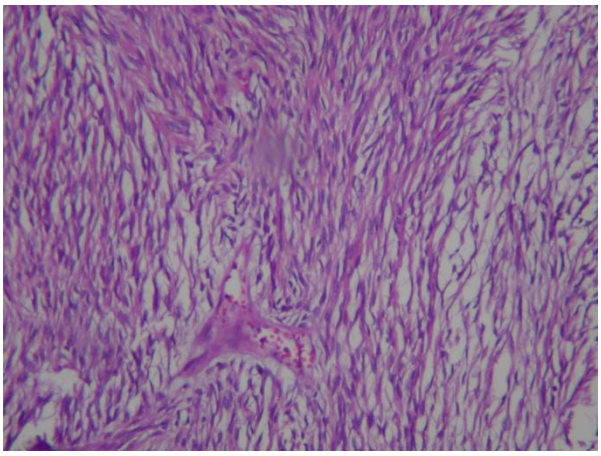

Figure 2. A fibrous capsule, hyaline vessels, cellular (Antoni A) and loose textured (Antoni B) areas and Verocay bodies (opposing rows of spindle nuclei separated by anucleate rows of eosinophilic processes) (Hematoksilen and Eosine).

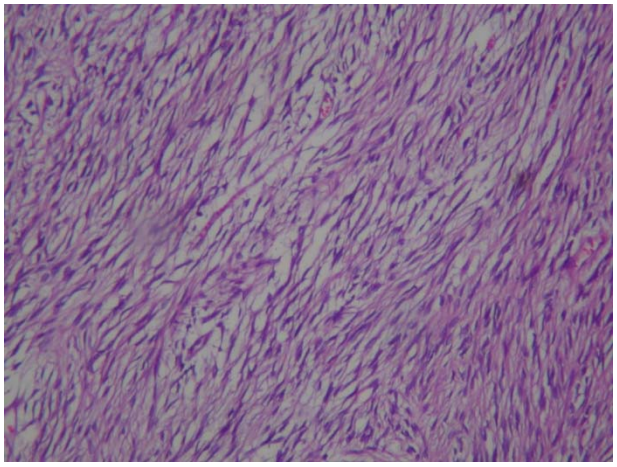

Figure 3. The tumoral cells in these areas present ill-defined cytoplasmic margins, eosinophilic, homogenous cytoplasm and spindled to oval or angulated nuclei.

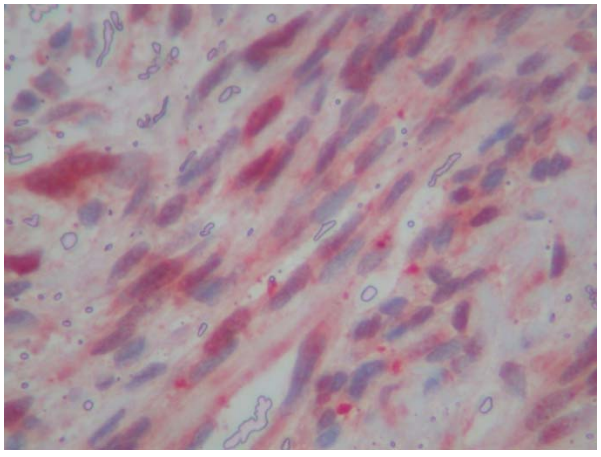

Figure 4. The types of areas being positive for vimentin and being negative for CD-34.

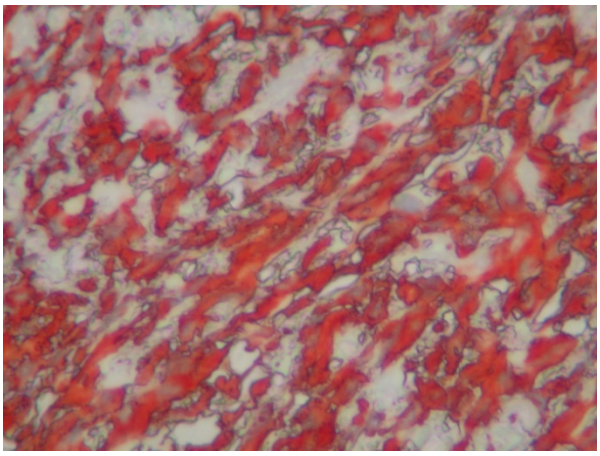

Figure 5. Immunhistochemically, the spindle cells were strongly positive for S100 protein.

There was no extension of the internal genital organs. On abdominal images there was no metastasis and pathologic intraabdominal lymphadenopathy. The results of laboratory tests and tumor markers were normal.

Surgical excision with clear margins was performed. The resected specimen to be a well-circumscribed, encapsulated lobular lesion measured 15x12 cm in size. The cut surface contains red and gray mass with necrotic and hemorrhagic areas.

On microscopic examination, fibrous capsule was identified. Central of the tumor, necrotic and hemorrhagic areas were noted. Nuclear palisating or in an organoid arrangement that formed Verocay bodies were found in the Antoni A area and in the Antoni B areas, the cells were loosely arranged with a more rounded cell morphology There were extensive myxoid and hyalinized areas. The tumor cells were spindle shaped with not prominent nucleoli, mitosis and necrosis was not seen (Figures 2, 3). Immunohistochemically the types of areas in Figure 4 being positive for vimentin and being negative for CD-34. Immunohistochemically, the spindle cells were strongly positive for S100 protein (Figure 5). As a result, diagnosis of the benign schwannoma was made. The woman was discharged after 5. postoperative days and remained asymptomatic 6 months later.

Discussion

Schwannoma is a benign nerve sheath tumor composed of schwann cells which arising from the myelin sheath of the peripheral nerves. Schwannomas are usually benign and less than 1% become malignant,

degenerating into a form of cancer known as neurofibrosarcoma which diagnosis depends on the number of mitotic figures present per tumor slide.

Schwannomas can occur anywhere along peripheral nerve fibers involving the neck, face and skalp. It has been described infrequently in the female lower genital tract (1,2). That arising from the female genital organs, such as the ovary, round ligament, cervix, clitoris and vulva, is seldom reported (3).

Schwannomas age range has varied from 5-84 years (4). But occur most commonly in the age group of 20-40 years in female population (5).

In a population-based study of schwannomas, 90% were observed to be sporadic, 3% occurred in patients with neurofibromatosis (NF-2), 2% occurred in association with schwannomatosis, and 5% in association with multiple meningiomas in patients without NF2. Rarely do schwannomas occur in patients with NF-1 (6). In our patient there was no history of any genetic disorders.

Clinical symptoms of the schwannoma vary appreciably depending on which nerve affected. Mostly painless, immobile, non-tender mass lesions are observed. Size of the tumor is an another important factor for clinical symptoms. In our case the size of the lesion was 15x12 cm and slowly increased but it prevent patient activities such as walking and sitting. Therefore, the quality of the patient's life was deteriorated. In the literature, most of the lesion size relatively small (2,7). In our best knowledge, the size of the lesion in our case is the largest size in the literature. Treatment modality is surgical. Wide local excision is done. In our patient, painless, immobile, non-tender mass was palpated on physical examination. Wide surgical exicision was done and the patient was followed.

Some tumors of the vulva including Bartholin cyst, labial cyst, and mesenchymal tumors such as lipoma and liposarcoma are relatively uncommon and may show nonspecific clinical features (8). The pre-operative diagnosis of the vulvar neoplasm is difficult. Therefore, a biopsy is often needed to exclude a malignant neoplasm and to indicate proper treatment. Schwannoma can rarely occur in the vulva and should be considered in the differential diagnosis. Sometimes schwannomas have been misdiagnosed as malignant tumors such as neurosarcoma, neurofibrosarcoma, and the malignant schwannoma (9).

The characteristic histologic appearance can make the accurate diagnosis. Histologically, conventional variant is the most common type of the vulvar schwannoma also plexiform and ancient variant of the vulvar schwannoma have been reported (4). Immunohistochemically malign schwannomas are thought to be composed of dedifferentiated schwann cells that had somewhat lost their capacity to syntheise S-100 protein (10). In our case, most of the schwann cells were strongly positive for S-100 protein. This method supported that the lesion is a benign nature.

Pre-operative gynecologic and radiologic examinations are so important for characterization and the differentiation of the benign and malign vulvar mass and guiding the surgical treatment. To slow growth of the mass in our patient, and the absence of lymph node metastasis, led us to think of a benign disease. Only surgical excision with clear margins and follow-up would be sufficient. Histopathologic and immunohistochemical examinations give the correct diagnosis. In the differential diagnosis of the benign vulvar tumors, we should be aware that schwannoma can occur in the vulva.

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