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CASE REPORT

A rare case of synovial sarcoma masquerading as a vulval cyst: A case report

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ABSTRACT

After gynecological cancers of the breast, cervix, uterus, and ovaries, vulvar cancer is the sixth most common type. Epidermoid carcinoma is the most typical histological form. However, the vulva, which is histologically defined as a collection of different tissues (cutaneous, glandular, adipocytes, muscular, and mucous), can experience a variety of modifications that can lead to lesions of various histological types, some of which may be uncommon, such as the synovial sarcoma of the vulva. There aren't many cases mentioned in the literature. Despite its name, Synovial Sarcoma (SS) is not of synovial origin; rather, it is produced from multipotent stem cells that have the capacity to differentiate into mesenchymal and epithelial cells. The most typical symptom is a localized pain or discomfort during intercourse, and the median age at diagnosis is 50. Frequently, these lesions are misdiagnosed as cysts or Bartholin abscesses. The surgical removal of the lesion with healthy tissue margins is the mainstay of treatment for synovial sarcoma. The outlook is not good. At two years, there is a 50% chance of locoregional recurrence or metastasis of SS. Local lymph nodes, lungs, bones, and liver are where metastatic lesions most frequently appear. A case of a 28-year-old lady who had vulvar synovial sarcoma is described in the current case report. This tumor management calls for a multidisciplinary approach.

Key words: Synovial Sarcoma, Bartholin abscesses, Epidermoid carcinoma, KI-67

INTRODUCTION

Vulvar cancer is the sixth type of gynecological cancer, following breast cancer, cancer of the cervix, cancer of the uterus, and cancer of the ovaries. Epidermoid carcinoma is the most prevalent subtype that can be seen histologically. Nonetheless, the vulva, which is defined histologically by the set of numerous tissues (cutaneous, glandular, adipocytes, muscular, and mucous), can undergo several alterations that can result in diseases of multiple histological natures, although these lesions might be infrequent [1]. There have been reports of cases of synovial sarcoma occurring in the vulva. Synovial sarcomas are uncommon mesenchymal malignant tumors that develop from multipotent stem cells that are capable of differentiating into mesenchymal and epithelial cells, despite the terminology that suggests a synovial origin. There have only been a few of cases published, leading one to believe that the occurrence of

this lesion is quite low, despite the fact that it has been documented [1,2].

CASE PRESENTATION

PRESENTATION

The current case report is about a 28 year old female subject. She reported to department of gynecology, out patient department, with the chief complaint of painful swelling since a week. She had no complaints of abnormal vaginal discharge, any menstrual disturbances, loss of appetite or weight loss. She had No previous medical / surgical illness. On examination it was observed that a lesion of 3x3cm, that was mobile, fluctuant, tender in upper half of right labia majora, with no palpable inguinal lymph nodes. Pelvic examination, general and other systemic examination showed no abnormality.

PRIMARY TREATMENT

Patient was treated conservatively with oral antibiotics, NSAID's, but with no symptomatic relief. Since there was no decrease in size of swelling after 10 days of treatment, decision for surgical intervention taken. Complete excision of cyst done under GA.

HISTOPATHOLOGY

The histopathology showed that circumscribed mass lesion with sheets of oval to plump spindle cells with vesicular nuclei and prominent eosinophilic nucleoli. Focal areas with frequent mitotic activity 4-5/10 HPF. (Figures 1)

DIAGNOSIS

The impression of poorly differentiated soft tissue neoplasm was arrived at.

DIFFERENTIAL DIAGNOSIS

Malignant melanoma / rhabdomyosarcoma / synovial sarcoma / poorly differentiated carcinoma.

IHC MARKERS

The immunohistochemical study showed CD34 neg, BCL-2 neg, CK5/6 neg, KI-67 positive (40-45%) and anti P-63 occasional cell positive - favouring synovial sarcoma.

MEDICAL TESTS PERFORMED

CT chest and abdomen were showed to be normal.
Patient defaulted for any further treatment as she had no further symptoms.

RECURRENCE

She had local recurrence with the involvement of the right labia majora.
Diagnostic work up for the second time was done and the following tests were performed: MRI pelvis, CT chest done with no abnormalities observed. The management was done by the Surgical oncologist through the wide excision of the lesion in right labia majora done. On the Histopathological examination it was seen that there were undifferentiated tumour with epithelioid morphology. The Margins were free. Mitotic rate was observed at 7/10 HPF. The immunohistochemical study showed positive for ER, PR, patchy Ck, Ki67 - 20% and it was Negetive for SMA, CD34, Melan A. (Figures 2) All this suggested a poorly differentiated high grade synovial sarcoma (FNLCC Grade 3), with a Pathological tumour stage of pT1b.

MANAGEMENT AND FOLLOW-UP

Postop radiation was given. Patient following up biannually (every 6 months) till 7 years. No

local recurrence or the distant metastasis. The subject delivered a child by LSCS during this period and had a uncomplicated pregnancy, CS and PNC period.

Figure 1: synovial sarcoma showing the oval to plump spindle cells with vesicular nuclei and prominent eosinophilic nucleoli.

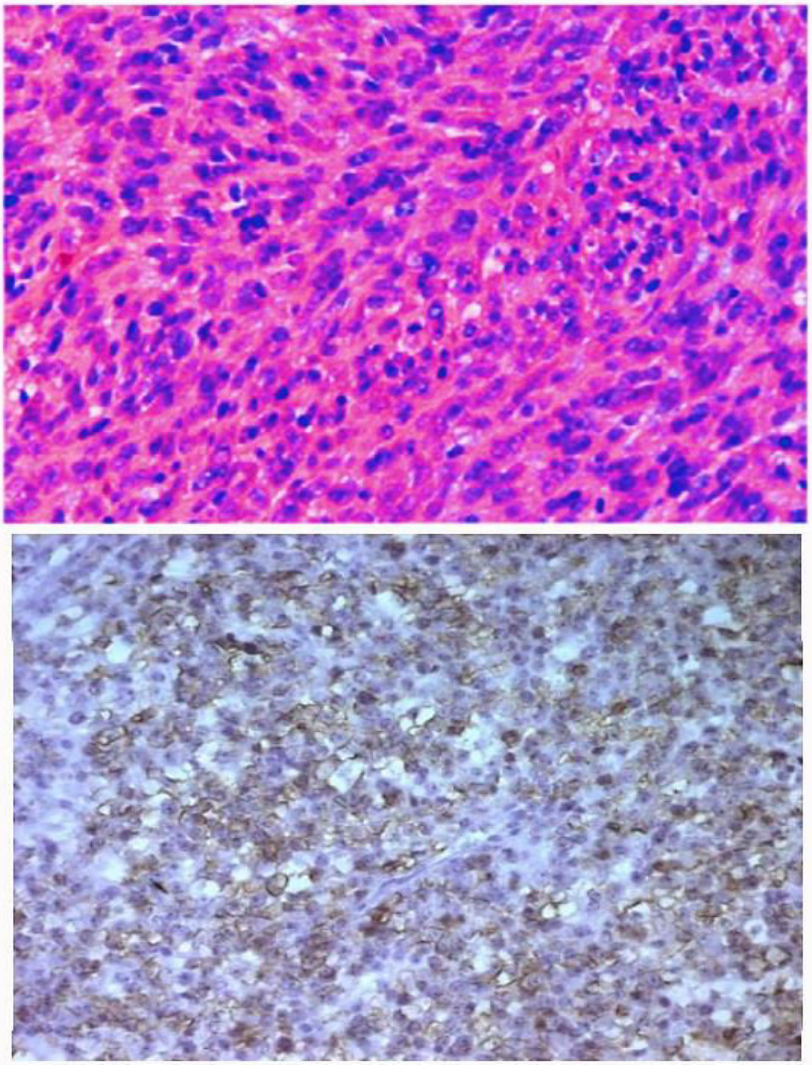

Figure 2: Immunohistochemical appearance after Ki 67 receptor labeling.

DISCUSSION

The term "synovial sarcoma" (abbreviated as "SS") refers to a rare form of malignant mesenchymal tumor that is more prevalent in younger persons and occurs most frequently in the periarticular regions of the limbs. Despite the name, synoviocyte-derived stromal cells (SS) are not formed from synovial fluid but rather from multipotent stem cells that are capable of developing into mesenchymal and epithelial cells [3]. It is an uncommon tumor with an unclear pathophysiology, and it accounts for 7% to 8% of malignant tumors that originate from the mesenchyme [4]. The biopsy is the most important part of making the diagnosis [5]. The anatomopathological descriptions differentiate between three subgroups, which are as follows: Fusiform cells that are monophasic, biphasic with a double cellular contingent consisting of epithelial and fusiform cells, and undifferentiated cells are described in [5]. The use of immunohistochemistry as a diagnostic tool is quite helpful. SS primarily express epithelial markers (cytokeratin and EMA), in 60% of cases the CD99, Ki67 protein, and in 30% of cases the S100 protein. This allows for the exclusion of the most common alternative diagnoses, which include fibrosarcoma, mesothelioma, leiomyosarcoma,

hemangiopericytoma, and a malignant tumor of the peripheral nerve sheaths [3].

In ninety percent of cases, the SS is characterized by a particular chromosomal t translocation (X; 18) (p11; q11), which is the translocation between the SYT gene of the chromosome 18 and one of the two homologous genes to Xp11, which indicates SSX1 or SSX2. It is hypothesized that the SYTSSX1 and SYT-SSX2 fusion proteins will operate as transcriptional regulators, which will either result in the activation of proto-oncogenes or the suppression of tumor suppressor genes [5]. The therapeutic care of synovial sarcoma is mostly surgical, consisting of substantial excision of the lesion with margins of healthy tissue. This is the primary mode of treatment. Local recurrence of tumors is possible (between 28 and 49 percent), as are lymph node metastases and distant metastases, the majority of which occur in the lungs (50 percent) [7]. A lot of the time, the SS will have a faux capsule that makes it possible for the tumor to be dispersed relatively readily. This will provide the patient a phony sense of security [2]. The presence of microscopic positive margins is connected with an increased risk of metastasis and gives a larger possibility of a local recurrence. The likelihood of a local recurrence is decreased by re-excision [2]. As a result, the surgeon must strive to achieve negative margins, despite the fact that there is no universal agreement over the appropriate size of the safety margin [2]. A lymphadenectomy of the inguinal region is not required unless there is evidence of metastases [8]. There is a significant chance of morbidity following radical vulvectomy, both in the short and long terms. However, local broad excision has the potential to produce positive or inadequate margins, which necessitates the use of adjuvant therapy such as radiotherapy, particularly in the case of high-grade tumors [1,9].

In the treatment of early diagnosed patients, particularly youngsters with modest primary tumors, radiation therapy appears to have a role to play [4]. Radiation therapy helps enhance local control of the disease, but it has no impact on the patient's chances of survival overall. In the event that there are metastases or certain pathological poor prognostic criteria, adjuvant radiotherapy may be recommended. There is a correlation between a tumor size that is higher than 5 centimeters, a high histological grade, tumoral surgical margins, and poor histological differentiation [4]. All of these factors are connected with a bad prognostic significance. Even though it is a tumor that responds well to chemotherapy, the necessity of adjuvant chemotherapy is still up for debate. Those who have metastases should seriously consider receiving chemotherapy. Doxorubicin and cyclophosphamide are the chemicals that are used the most, however it is unclear whether or not they actually improve patients' chances of surviving [4,6]. The outlook does not look good. After two years of SS, the typical rate of recurrence, whether it is locoregional or metastatic, is fifty percent. Regional lymph nodes, the lung, the bone, and the liver are the most often seen metastatic locations. A tumoral size of less than 5 centimeters, a reduced mitotic index (less than ten mitoses for ten fields at high magnification), a low proliferation index (Ki-67<10%), the absence of tumor necrosis, the absence of Residual tumor after surgical resection, young age, and female sex are all factors that are considered to be associated with a more favorable prognosis [4]. In the presence or absence of these parameters, respectively, the survival rate after five years ranges anywhere from 76% to 35% and the survival rate after ten years is anywhere from 63% to 10% [4]. Factors that determine prognosis- the function of the fusion transcript as a factor that determines prognosis has not been conclusively proven. Although a number of research have suggested that the presence of SYT-SSX1 is linked to a more dismal prognosis than that of SYTSSX2 [5, subsequent findings do not substantiate this finding [10]. One study revealed that the histological grade of the SS was the most important predictive predictor, and their findings were independent of the histology type and the kind of fusion transcript [10]. This was accomplished by stratifying the SS according to their histological quality. There is no correlation between the histological subtypes of SS and the patient's prognosis [5].

CONCLUSION

The SS of the vulva is an extremely uncommon form of vulvar cancer. Research techniques such as immunohistochemistry and cytogenetic analysis are able to differentiate it from other types of mesenchymal malignancies. Confirmation of the diagnosis is made feasible because of the presence of a SYT-SSX fusion transcript in the subject's genome. The conventional treatment for non-metastatic SS is surgical excision, which may or may not be accompanied by irradiation of the tumor bed.

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