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Synovial Sarcoma of the Vulva: A Rare Yet Existing Entity! Case Report

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Abstract

Vulvar cancer is the fifth gynecological cancer after that of the breast, cervix, uterus, and ovaries. The most common histological type is epidermoid carcinoma. But, the vulva, defined histologically by the set of several tissues (cutaneous, glandular, adipocytes, muscular, mucous) can undergo several modifications resulting in lesions of multiple histological nature that might be rare like the synovial sarcoma of the vulva. Few cases have been reported in the literature. Despite this terminology, the Synovial Sarcoma (SS) is not synovial origin, but is derived from multipotent stem cells capable of differentiating into mesenchymal and epithelial cells. The median age at diagnosis is 50 years old, and the most common symptom is a local discomfort or pain during sex. These lesions are often wrongly diagnosed as cysts or Bartholin abscess. The therapeutic management of synovial sarcoma is primarily surgical with wider excision of the lesion with healthy tissue margins. The prognosis is poor. The average rate of locoregional recurrence or metastasis of SS at two years is 50%. The most frequent metastatic sites are the regional lymph nodes, lung, bone and liver. We report the case of a 40-year-old woman with synovial sarcoma of the vulva. The management of this tumor requires multidisciplinary approach.

Introduction

Vulvar cancer is the fifth gynecological cancer after that of the breast, cervix, uterus, and ovaries. The most common histological type is epidermoid carcinoma. But, the vulva, defined histologically by the set of several tissues (cutaneous, glandular, adipocytes, muscular, mucous) can undergo several modifications resulting in lesions of multiple histological nature that might be rare [1]. Some cases of synovial sarcoma (rare mesenchymal malignant tumors developed from multipotent stem cells capable of differentiating into mesenchymal and epithelial cells, despite the terminology that suggests a synovial origin) have been described in the vulva. Only few cases are reported, which supposes that, the incidence of this lesion is very rare, but still existent [1,2].

Case Presentation

We report the case of a 40-year-old patient, with no medical history, gravida 4, para 4. She had regular menstruation cycles, and used a copper IUD (intrauterine device) as a means of contraception. The physical examination found a patient in good general condition. The vulva was seat of a 4-centimeter-long swelling of the upper half of the left labia majora, polylobed, firm, without inflammatory signs. The rest of the gynecological examination was normal and the ganglionic areas were free. Fine needle biopsy of this swelling concluded to undifferentiated carcinoma. Indeed, it brought 1.5 cc of serohematic liquid. In cytological examination, it was a moderately inflammatory fluid, containing necrotic debris, red blood cells, lymphocytes and numerous tumor cells isolated or arranged in clusters with a basophilic cytoplasm. After incorporation in paraffin, the tumor cells were large, undifferentiated, with large nucleated nuclei that contained many mitoses. The immunohistochemical study showed focal labeling with EMA, vimentin and Ki67. Furthermore, this labeling was negative for keratin, desmin, PS100, CD34, CD20, CD3, CD10, HMB45 and P63. The patient underwent an excision: hemivulvectomy. Macroscopic examination showed a friable, translucent, greyish, mucoid tumor, well limited and encapsulated. Microscopic examination noticed the presence of central rounded cells with regular nuclear contour, which aggregated into solid, dense masses, separated from myxoid substance of variable abundance. The tumor tissue was partially lobulated, traversed by thick eosinophilic fibrous septa predominating at the periphery. There were large areas of necrosis occupying about 60% of the tumor. The excision passed through healthy tissue in contact with the capsule. The tumor cells expressed: CD99, vimentin, CL2, keratin

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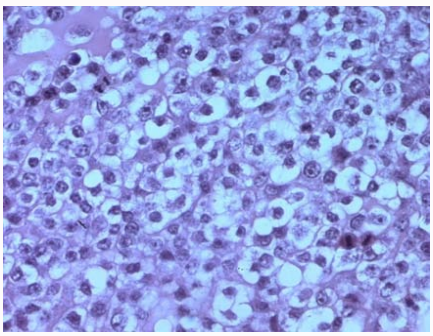

Figure 1: Histological appearance before immunohistochemistry at high magnification: Round cells arranged into solid and dense masses with moderate mitotic activity and a wide range of necrosis which occupies more than 60% of the tumor.

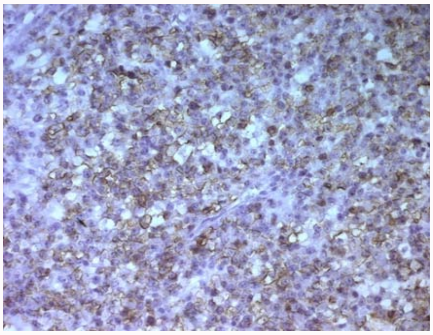

Figure 2: Immunohistochemical appearance after CD99 receptor labeling.

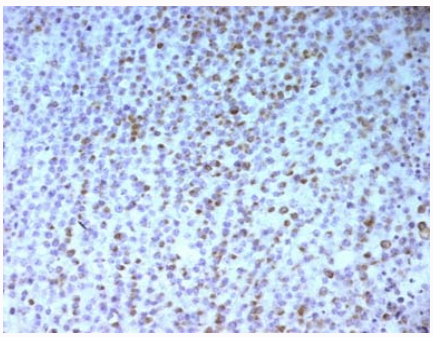

Figure 3: Vimentin receptor labeling. The excision was complete, flush with the fibrous capsule of the tumor at the inner and the bottom edges. The patient had a re-excision without tumor residue. The extension assessment did not find secondary thoracic localization in CT scans.

and EMA. All this suggested a monophasic Synovialosarcoma with 3.5 cm long-axis large cells of grade 3 (differentiation: 3, mitosis: 2, necrosis: 2) (Figures 1-3).

Discussion

Synovial Sarcoma (SS) is a rare malignant mesenchymal tumor, more common in young adults, with prominent occurrences in the periarticular regions of the limbs. Despite this terminology, SS is not of synovial origin, but is derived from multipotent stem cells, capable of differentiating into mesenchymal and epithelial cells [3]. It is a rare tumor with unknown pathogenesis, representing 7% to 8% of malignant tumors of mesenchymal origin [4]. The diagnosis is mainly made by the biopsy [5]. The anatomopathological descriptions define three subtypes: Monophasic fusiform cells, bi-phasic with a double cellular contingent: Epithelial and fusiform and finally undifferentiated [5]. Immunohistochemistry is a useful diagnostic tool. SS mainly express epithelial markers (cytokeratin and EMA), in 60% CD99 and in 30% of cases the S100 protein, allowing to rule out the main differential diagnoses (fibrosarcoma, mesothelioma, leiomyosarcoma, hemangiopericytoma or a malignant tumor of the peripheral nerve sheaths) [3]. In 90% of cases, the SS is characterized by a specific 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 means SSX1 or SSX2. The SYTSSX1 and SYT-SSX2 fusion proteins are expected to function as transcriptional regulators, resulting in either the activation of proto-oncogenes or the inhibition of tumor suppressor genes [5]. The therapeutic management of synovial sarcoma is essentially surgical with extensive excision of the lesion with margins of healthy tissue. Tumors can relapse locally (28% to 49%) or give lymph node metastases or distant metastases mainly in the lungs (50%) [7]. Often, the SS has a pseudo capsule that allows the tumor to be disbursed quite easily, giving a false feeling of security [2]. The presence of microscopic positive margins gives a greater probability of local recurrence and is associated with an increased risk of metastasis. Re-excision reduces the risk of local recurrence [2]. Therefore, the surgeon must aim to obtain negative margins, although there is no consensus on the necessary safety margin [2]. Inguinal lymphadenectomy is not necessary unless there are metastases [8]. Radical vulvectomy has a high risk of short and long-term morbidity. However, local wide excision may result in positive or insufficient margins requiring the use of adjuvant therapy such as radiotherapy, especially for high-grade lesions [1,9]. Radiation therapy seems to have a role in the treatment of early diagnosed patients, particularly for children with minimal primary tumors [4]. Radiation therapy improves local control of the disease, but does not affect overall survival. Adjuvant radiotherapy is considered in case of metastases or pathological poor prognosis criteria: A tumor size greater than 5 cm, high histological grade, tumoral surgical margins and poor histological differentiation are all associated with an adverse prognostic significance [4]. Although it is a chemosensitive tumor, the role of adjuvant chemotherapy remains controversial. Chemotherapy should be considered in patients with metastases. The most widely used molecules are doxorubicin and cyclophosphamide, but their real influence on survival remains contested [4,6]. The prognosis is bad. The average rate of locoregional or metastatic recurrence after 2 years of SS is 50%. The most common metastatic sites are regional lymph nodes, lung, bone and liver. A tumoral size of less than 5 cm, 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, female sex, are considered as factors of better prognosis [4]. The five-year survival rate varies from 76% to 35% and the ten-year survival from 63% to 10%, in the presence or absence of these criteria respectively [4]. Prognostic factors; the role of the fusion transcript as a prognostic factor is not definitively established. Although many studies have reported that the presence of SYT-SSX1 was associated with a more pejorative prognosis than that of SYT-SSX2 [5], more recent results do not confirm this [10]. By stratifying the SS according to their histological grade, one team demonstrated that it was the most important prognostic factor, independently of the histological type and the type of fusion transcript [10]. The histological subtypes of SS are not prognostic factors [5].

Conclusion

The SS of the vulva is a rare malignant tumor. Immunohistochemical study and cytogenetic analysis distinguish it from other mesenchymal tumors. The presence of a SYT-SSX fusion transcript makes it possible to affirm the diagnosis. Surgery associated or not with irradiation of the tumor bed is the standard treatment for non-metastatic SS.

References

1. Behranwala KA, Latifaj B, Blake P, Barton DP, Shepherd JH, Thomas JM. Vulvar soft tissue tumors. Int J Gynecol Cancer. 2004;14(1):94-9.
2. Asher V, van Schalkwyk G, Bali A. Synovial sarcoma of the vulva: A case report. J Med Case Rep. 2011;5:95.
3. Dos Santos NR, de Bruijn DR, van Kessel AG. Molecular mechanisms underlying human synovial sarcoma development. Genes Chromosomes Cancer. 2001;30:1-14.
4. Eilber FC, Dry SM. Diagnosis and management of synovial sarcoma. J Surg Oncol. 2008;97:314-20.
5. Ladanyi M, Antonescu CR, Leung DH, Woodruff JM, Kawai A, Heale JH, et al. Impact of SYT- SSX fusion type on the clinical behavior of synovial sarcoma: A multi-institutional retrospective study of 243 patients. Cancer Res. 2002;62:135-40.
6. Holloway CL, Russell AH, Muto M, Albert M, Viswanathan AN. Synovial cell sarcoma of the vulva: Multimodality treatment incorporating preoperative external-beam radiation, hemivulvectomy, flap reconstruction, interstitial brachytherapy, and chemotherapy. Gynecol Oncol. 2007;104(1):253-6.
7. Ambani DS, White B, Kaplan AL, Alberto A. A case of monophasic synovial sarcoma presenting as a vulvar mass. Gynecol Oncol. 2006;100(2):433-6.
8. Spurrell EL, Fisher C, Thomas JM, Judson IR. Prognostic factors in advanced synovial sarcoma: An analysis of 104 patients treated at the Royal Marsden Hospital. Ann Oncol. 2005;16(3):437-44.
9. Ulutin HC, Zellars RC, Frassica D. Soft tissue sarcoma of the vulva: A clinical study. Int J Gynecol Cancer. 2003;13(4):528-31.
10. Guillou L, Benhattar J, Bonichon F, Gallagher G, Terrier P, Stauffer E, et al. Histologic grade, but not SYT-SSX fusion type, is an important prognostic factor in patients with synovial sarcoma: A multicenter, retrospective analysis. J Clin Oncol. 2004;22:4040-50.