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Case Report

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Periductal Stromal Sarcoma of the Breast: A Case Report

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ABSTRACT

Introduction: Periductal Stromal Sarcoma (PSS), especially spindle and epithelioid types, is a rare subtype of the malignant fibroepithelial tumor with benign ductal elements and a sarcomatous stroma composed of spindle cells. The therapeutic management of PSS is based on wide surgery with free margins, and adjuvant therapies are not required.

Case Presentation: This report describes a 37-year-old woman who presented to Shariati hospital with a right breast mass for review and a second opinion pathology report. The patient had undergone a radical mastectomy in another hospital three months previously. Histological and immunohistochemical examinations revealed PSS and all dissected lymph nodes were free of tumor. Based on the diagnosis, the patient received no adjuvant treatment (such as chemotherapy or radiotherapy). After nine months of close follow-up examinations, no recurrence was observed.

Conclusion: PSS is an extremely rare disease with low-grade sarcomatous behavior, which may evolve into a phyllodes tumor or an entity of breast cancer. Therefore, frequent follow-up examinations are required.

Introduction

Primary sarcomas of the breast are extremely rare, with less than 0.1% of all malignant tumors of the breast [1]. Breast cancer manifests itself in the mammary epithelium, yet there is a growing recognition that mammary stromal cells also play an important role in tumorigenesis [2]. Breast neoplasms of connective tissue origin have been categorized morphologically as cystosarcoma (fibroepithelial neoplasms), pure connective tissue tumors (without epithelial component, resembling their soft tissue counterparts), and Periductal Stromal Sarcoma (PSS) with intermediate features [3]. Biphasic breast tumors with benign ductal elements

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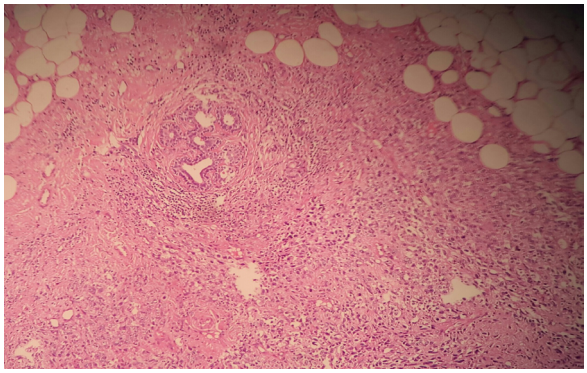

Figure 1. Microscopic examination of the breast tumoral tissue revealed spindle cell proliferation with atypia surrounding intact ducts (H&E stain, x40)

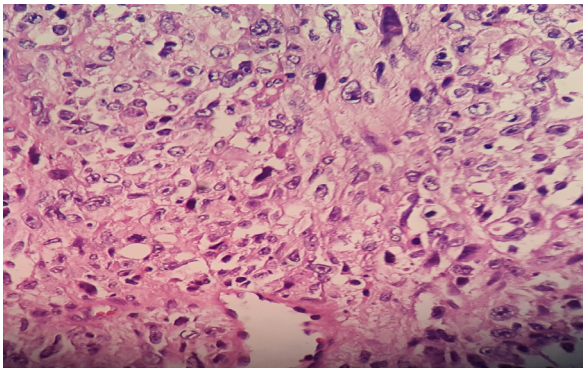

Figure 2. Microscopic examination of the breast tumoral tissue revealed spindle cell proliferation with marked atypia (H&E stain, x400).

and a sarcomatous stroma lacking a phyllodes architecture are a source of diagnostic problems, particularly because of the lack of an appropriate designation [4]. Histologically, they are composed of spindle cells with highly pleomorphic nuclei and abundant mitoses [5]. The epithelial component in the current tumor demonstrated usual epithelial hyperplasia and there was no evidence of ductal carcinoma in situ or invasive carcinoma [6]. Patients with PSS ranged in age from 37 to 89 years (mean 55.3 years) and the tumors are 0.2–6.0 cm (mean 2.97 cm) [4]. Its therapeutic management is based on wide surgery with free margins and adjuvant therapies are not needed [7]. The role of radiation therapy and chemotherapy remains unclear [8]. All attempts should be made to achieve a negative margin as this appears to be the only factor influencing survival and local recurrence in these patients [9]. Tumors larger than 5 cm are associated with an elevated risk of systemic failure and a poor prognosis [10].

Case Presentation

A 37-year-old woman was referred to the department of pathology of Shariati hospital affiliated to Tehran University of Medical Science and provided the pathology report from another city for right breast mass for review and second opinion. The patient had undergone a radical mastectomy in another hospital three months previously and was diagnosed with stromal sarcoma after the operation. The patient's family history and past medical history were unremarkable. The patient had a solid tan-whitish lesion with firm incontinency 2.5cm in maximum diameter. Histological and Immunohistochemical (IHC) examinations revealed a periductal stromal tumor and all dissected lymph nodes were free of tumor. Histology examination showed plump spindle cell proliferation with atypia and marked mitosis surrounding ductal epithelium with no significant pathologic changes. Also, lymphocyte infiltrations were observed (Figure 1 & 2). For confirmation of the diagnosis, the IHC study was done and CD34 and vimentin were positive in spindle cells. S100 and cytokeratin staining were negative in tumoral cells. PSS, especially spindle and epithelioid types, is a rare subtype of the malignant fibroepithelial tumor with benign ductal elements and a sarcomatous stroma composed of spindle cells with positive vimentin and CD34, which is compatible with our case. Based on the diagnosis, the patient received no adjuvant treatment (such as chemotherapy or radiotherapy). After nine months of close follow-up examinations, no recurrence was observed. Thus, PSS is a useful descriptive designation for generally low-grade biphasic tumors with sarcomatous stroma with no features of a phyllodes tumor.

Discussion

PSS should not be confused with other spindle cell breast tumors because they require different treatments [11]. For most mammary sarcomas, simple mastectomy is an appropriate therapy [12]. Excision of the axillary lymphatics and adjuvant radiotherapy are unlikely to be beneficial [13]. PSS is an extremely rare disease with low-grade sarcomatous behavior, which may progress into a phyllodes tumor or an entity of breast cancer. Therefore, frequent follow-up examinations are required [14]. While primary non-phylloides breast sarcomas are rare tumors, poor treatment and prognosis have been reported [15]. The degree of mitotic activity is not highly correlated with the behavior of the sarcoma but it is apparent that any tumor with five or more mitotic figures per 10 high-power fields is capable of

metastasis [16]. Tumor size >5 cm is the only significant prognostic indicator of overall survival [13]. Overgrowth of the sarcomatous stroma is likely a requirement for metastasis [17].

Conclusion

PSS is treated with excision with clear margins. Axillary lymph node dissection is not required. Thus, a definite diagnosis is necessary before receiving extra and unnecessary treatments on patients. There is a need for further studies on the histogenesis and biological behavior of mammary sarcomas.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

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Conflict of interest

The authors declared no conflict of interest.

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