American Journal of Cancer Prevention, 2015, Vol. 3, No. 3, 62-64
Available online at http://pubs.sciepub.com/ajcp/3/3/4

DOI:10.12691/ajcp-3-3-4

Malignant Phyllodes Tumor in Patient with Breast Cancer: A Case Report

Mehrdad Payandeh[1], Hooshang Usefi[2], Edris Sadeghi[3,4,*], Masoud Sadeghi[3,4]

[1]Department of Hematology and Medical Oncology, Kermanshah University of Medical Sciences, Kermanshah, Iran
[2]Blood Transfusion Research Center, High Institute for Research and Education in Transfusion Medicine, Kermanshah, Iran
[3]Students Research Committee, Kermanshah University of Medical Sciences, Kermanshah, Iran
[4]Medical Biology Research Center, Kermanshah University of Medical Sciences, Kermanshah, Iran
*Corresponding author: Sadeghi_mkn@yahoo.com

Received April 15, 2015; Revised May 10, 2015; Accepted May 31, 2015

Abstract **Background:** Phyllodes tumors (PT) have an incidence of 1 per 100,000 women and account for only 0.5% of all breast neoplasms. Herein, we present a case of malignant PT characterized by stromal overgrowth of a high-grade spindle cell sarcoma with marked stromal pleomorphism. Which this disease is rare and the lack of standard care options for it. **Case Report:** A 36-year-old woman with a metastatic malignant PT presented to our clinic to discuss treatment options. Immunohistochemical analysis showed that the lesion was negative for estrogen and progesterone receptor and HER-2/neu, ki67 > 80% positive and vimentin positive. During a futher evaluation in computed tomography (CT) of the chest revealed multiple lung masses was seen. Pathology review of the mastectomy product at our institution confirmed the diagnosis of a malignant PT characterized by stromal overgrowth of a high-grade spindle cell sarcoma with marked stromal pleomorphism, a high mitotic rate, and areas of necrosis. She treated with chemotherapy regimen for soft tissues sarcoma consist of vincristine, cyclophosphamide, doxorubicine. After three courses of this regimen despite of decease in severity of dyspnea the breast mass lesion progressed and the therapy change to ifosfamide and etoposide combined with Nexavar (Sorafenib). At now, the patient is treating with this policy. **Conclusions:** We reported a patient with malignant PT should receive adjuvant chemotherapy to reduce the risk of recurrence. We suggest chemotherapy with ifosfamide and etoposide combined with Nexavar (Sorafenib) because it is a good policy for close treatment.

Keywords: *chemotherapy, CT scan, Phyllodes tumors, Sorafenib*

Cite This Article: Mehrdad Payandeh, Hooshang Usefi, Edris Sadeghi, and Masoud Sadeghi, "Malignant Phyllodes Tumor in Patient with Breast Cancer: A Case Report." *American Journal of Cancer Prevention*, vol. 3, no. 3 (2015): 62-64. doi: 10.12691/ajcp-3-3-4.

1. Introduction

Breast cancer is the most common cancer (27% of all cancers) and common cause of death [1] that can be a leading cause of death through middle-aged women [2]. Phyllodes tumors (PT) have an incidence of 1 per 100,000 women and account for only 0.5% of all breast neoplasms [3]. WHO in 1981 classified PT into three types: benign, borderline and malignant in 2003 [4]. Patients with PT tumors with distant metastasis are incurable and have a poor prognosis; most will die within 3 years following the diagnosis of metastasis [5]. PT is predominantly benign, asymptomatic tumors that manifest as large masses. The median size of a PT is 4–7 cm, and it may cause fatigue, dyspnea, and bone pain in metastatic disease [6]. The tumor usually occurs in 35- to 55-year-old women [3]. Herein, we present a case of malignant PT characterized by stromal overgrowth of a high-grade spindle cell sarcoma with marked stromal pleomorphism. Which this disease is rare and the lack of standard care options for it.

2. Case Report

A 36-year-old woman with a metastatic PT presented to our clinic to discuss treatment options. Eight months ago, a progressive mass had been detected in her left breast for which last two months earlier a biopsy was consistent with breast sarcoma (PT). The breast mass not completely resectable and only biopsy was done, and the diagnosis high- grade sarcoma arising in a PT with no positive lymph nodes (0/8). Immunohistochemical analysis showed that the lesion was negative for estrogen and progesterone receptor and HER-2/neu, ki67 > 80% positive and vimentin positive. During a future evaluation in computed tomography (CT) of the chest revealed multiple lung masses was seen (Figure 1). No other sites of metastatic disease were detected. The largest mass measured 10.2 × 7.6 × 8.1 cm. Pathology review of the mastectomy product at our institution confirmed the diagnosis of a malignant PT characterized by stromal overgrowth of a high-grade spindle cell sarcoma with marked stromal pleomorphism, a high mitotic rate, and

areas of necrosis (Figure 2). She treated with chemotherapy regimen for soft tissues sarcoma consist of vincristine, cyclophosphamide, doxorubicine (Figure 3). After three courses of this regimen despite of decease in severity of dyspnea the breast mass lesion progressed and the therapy change to ifosfamide and etoposide combined with Nexavar (Sorafenib) (Figure 4). At now, the patient is treating with this policy.

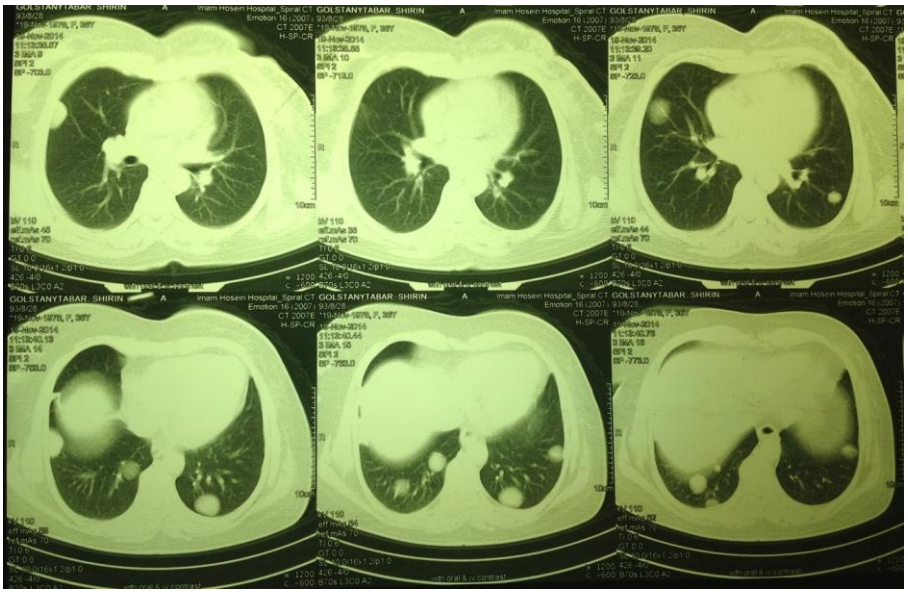

Figure 1. There is multiple round shape lesions in both filed of the lungs (before the first therapy).

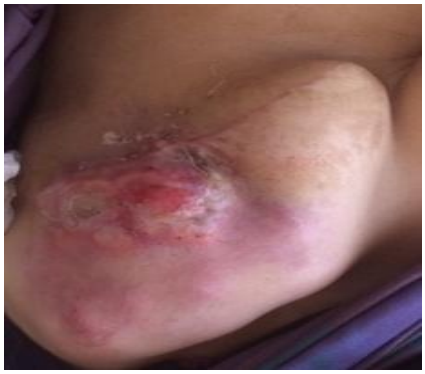

Figure 2. Malignant phyllodes tumor characterized by stromal overgrowth of a high-grade spindle cell sarcoma with marked stromal pleomorphism

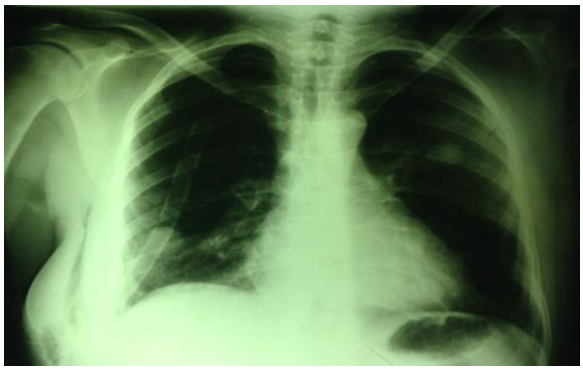

Figure 3. In chest X-ray, there is multiple shrinkage defined lesions with right side chest tube in the lung (after 3 courses of chemotherapy).

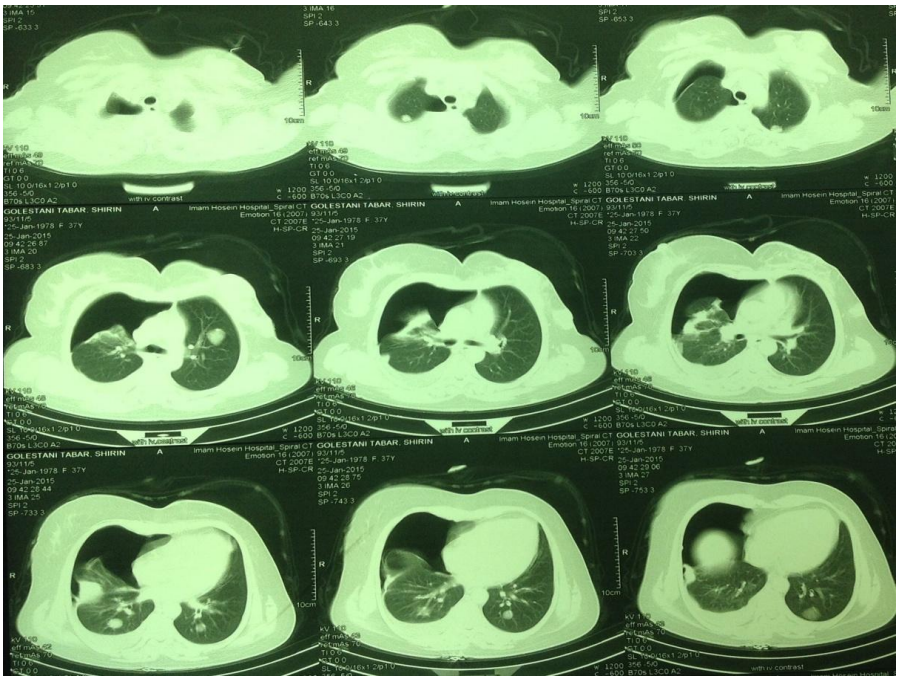

Figure 4. Right side pneumothorax was induced after shrinkage of right side lesion due to chemotherapy

3. Discussion

PTs are rare lesions with an incidence of less than 1% of all breast tumors [7]. The 2003 WHO tumor classification proposed the classification of PTs into three categories (benign, borderline and malignant) according to the degree of cellular atypia, mitotic activity, characteristics of the tumor margins and the presence of stromal growth [8]. That is benign PT is more likely to choose excision, however, for the malignant, mastectomy is favored [9]. The PT of the breast is a rare disease usually presents as a large lump. In few cases, it is bilateral or multifocal [10]. The benefit of adjuvant chemotherapy is controversial. There have been no published randomized studies of adjuvant therapy specifically in PTs. In an observational study, 28 patients with malignant PTs of the breast received four cycles of adjuvant chemotherapy (doxorubicin 65 mg/m^2 and dacarbazine 960 mg/m^2) [6]. Adjuvant chemotherapy with doxorubicin and dacarbazine was found be not effective in patient survival. The patient who developed lung metastasis, the institution of doxorubicin-based chemotherapy did not change the outcome [11]. In our study the patient treated with chemotherapy regimen for soft tissues sarcoma consist of vincristine, cyclophosphamide, doxorubicine and then decease in severity of dyspnea the breast mass lesion progressed. The difficulty in distinguishing between phyllodes tumors and benign fibroadenoma may lead to misdiagnosis. In fact, there are no characteristic features that clinically distinguish PTs from other breast tumors [3]. Malignant PTs are the most aggressive, and are characterized by marked stromal cellularity and atypia, positive infiltrative margins, more than ten mitoses per ten high power fields, and stromal overgrowth [12]. In this case, our diagnosis was a PT characterized by stromal overgrowth of a high-grade spindle cell sarcoma with marked stromal pleomorphism, a high mitotic rate, and areas of necrosis. And Jara-Lazaro et al. showed that moderate-to-severe stromal cellularity, stromal overgrowth, moderate nuclear atypia, stromal mitoses of $\geq$ 2/ 10 hpf and ill-defined lesional borders on core needle biopsy were exclusive to the diagnosis of PT on excision [13].

4. Conclusions

We reported a patient with malignant PT should receive adjuvant chemotherapy to reduce the risk of recurrence. We suggest chemotherapy with ifosfamide and etoposide combined with Nexavar (Sorafenib) because it is a good policy for close treatment.

Acknowledgement

There is no acknowledgement.

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