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OLGU SUNUMU

UNDIFFERENTIATED SARCOMA OF THE BREAST

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ABSTRACT

The mesenchymal tumors of the breast are rare and they generally consist less than 1% of all breast tumors. Wide surgical excision with clear surgical margins is the most effective treatment. Adjuvant radiotherapy and chemotherapy may be considered in selected patients.

A fourteen year old girl admitted to our breast clinic with a recurrent big mass in her right breast. She underwent an excisional biopsy two times before in different hospitals. And, histopathologic studies showed indifferentiated sarcoma of the breast without surgical margin examination. At physical examination, she had a 13 cm in diameter mass lesion in her right breast, invading to whole breast, pectoralis major muscle and palpable lymph nodes at ipsilateral axillary region. Right radical mastectomy was performed with reconstruction of chest wall with free skin graft. Pathologic studies were same as previous results of undifferentiated sarcoma. Following the initial surgical procedure she had 3 cycles vincristine, adriamycine, cyclophosphamide and 3 cycles iphosphomide and etoposide and radiotherapy [total 5040 cGy (thoracic and internal mammarian) irradiation]. She had pulmonary metastases at 12th month and died at 14th month due to systemic metastases to the lungs and the liver.

With this case presentation, we want to remind the possibility of mesenchymal malignant tumors in especially young females. These lesions should be resected with negative surgical margins.

Key words: breast, mesenchymal tumor, sarcoma

MEMENİN İNDİFERANSİYE SARKOMU

ÖZET

Memenin mezenkimal tümörleri tüm meme tümörlerinin %1'inden azını oluşturur. Tedavisinde genişçe, cerrahi sınırlar negatif olacak şekilde eksizyonu önerilir. Seçilmiş hastalarda radyoterapi ve kemoterapi tedavi seçenekleri arasındadır.

14 yaşında genç bayan sağ memesinde tekrarlayan büyük kitle ile kliniğimize başvurdu. Hastaya değişik hastanelerde aynı memeden iki kez eksizyonel biyopsi yapılmıştı. Patoloji sonucu indiferensiye sarkom olarak gelen hastanın ikinci eksizyonel biyopsi sonrası memesindeki kitlenin hızla büyümesi üzerine yapılan tetkiklerinde sağ memede 13 cm boyutunda tüm memeyi kaplayan, pektoralis major kasını invaze eden kitle saptandı. Yapılan fizik muayenesinde sağ memede tüm memeyi kaplayan kitle ve aksillada palpabl lenf adenopatiler saptandı. Hastaya radikal mastektomi ve toraks duvarı onarımı yapıldı. Ameliyat sonrası 3 kür vinkristin, adriyamisin, siklofosfamid ve 3 kür ifosfamid, etoposid ve radyoterapi aldı. 12. ayda akciğer metastazı gelişen hasta 14. ayda kaybedildi.

Anahtar sözcükler: meme, mezenkimal tümör, sarkom

Breast sarcomas are very rare breast tumors. They originate from various cell types of mesencymal tissue. They constitute less than 1% of all breast malignancies and most commonly ocur at fifth to sixth decades (1-3). The most frequently reported breast sarcomas are cystosarcoma phylloides. The other sarcomas seen in the breast are angiosarcomas, liposarcomas, fibrosarcomas, osteosarecomas, leiomyosarcomas, rhabdomyosarcomas, dermatofibrosarcoma protuberans and malignant fibrous histiocytomas (4). Wide tumor excision with clear surgical margins is the most effective treatment, and mastectomy may be necessary for large tumors. Lymph node dissection is required in patients with positive lymph node(s) in physical examination and radiologic studies. Adjuvant chemotherapy and/or radiation therapy may be necessary in selected patients.

Case Report

A 14-year-old female patient admitted to our clinic with a large mass lesion in her right breast. She recognized this lesion 9 months ago and underwent excisional biopsy in a hospital. Her lesion recurred and excisional biopsied two months ago. The pathology results of these two biopsies revealed undifferentiated sarcoma. She stated that the tumor was getting much bigger after last excisional biopsy. At physical examination in our clinic, there was a hard, solid, irregular mass with a 12x13 cm in diameter, invading the whole right breast and pectoralis major muscle with a palpable right axillary lymph nodes (Figure 1). She had no family history. Breast magnetic rezonans imaging (MRI) and thorax computed tomography (CT) revealed a 12x13 cm in diameter lesion in right breast including internal septations with sternum and pectoralis

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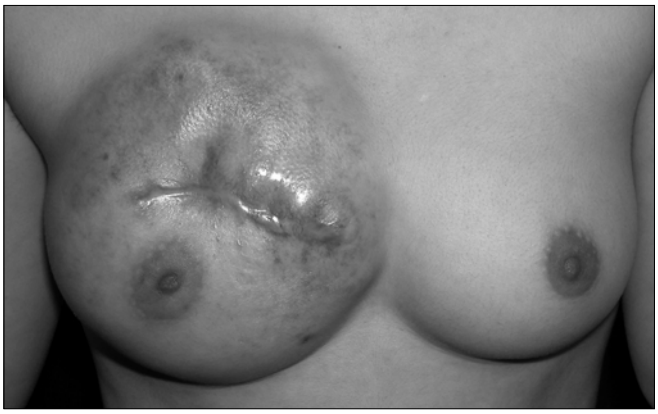

Figure 1. The appearance of the ecchymotic, hard, irregular mass invading the entire right breast and the scar of the previous incision.

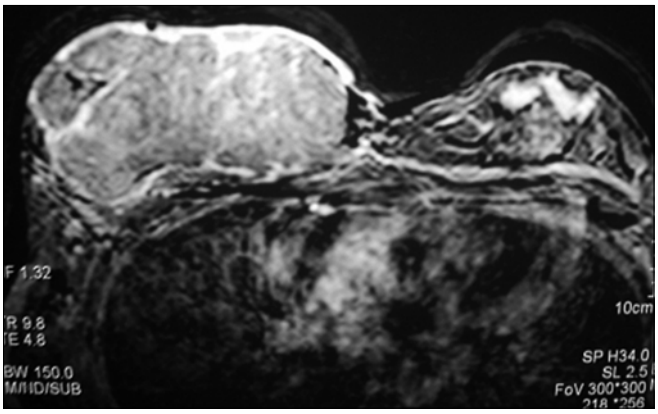

Figure 2. The mass with internal septations invading the entire right breast tissue and neighboring the pectoral muscles and the sternum.

major muscle invasion (Figure 2 and 3). Abdominal CT examination and whole body bone sintigraphy showed normal results.

Right radical mastectomy with excision of invaded sternum and reconstruction of chest wall with latissimus dorsi muscle flap and skin graft were performed. The histopathologic results showed an undifferentiated sarcoma with dermal invasion, pectoralis muscle invasion and 10 mm negative surgical margins. The tumour had mesenchymal component but not appropriate with cystosarcoma phylloides and no epithelial component and also high mitotic index, necrosis and with only vimentine positivity (Pancytoceratine, SK 7, 19 and 20, Desmine, Myo D1, CD 31, CD 34, MSA, NSE, Myc-2, Synaptophysine and FVIII were negative) (Figure 4). The retrieved 18 axillary lymph nodes were negative.

She discharged from the hospital at postoperative day 12 and she had chemotherapy of 3 cycles vincristine, adriamycine, cyclophosphamide and 3 cycles iphosphomide and etoposide and radiotherapy [total 5040 cGy (thoracic and internal mammarian) irradiation] as an adjuvant therapy. She was followed-up at office visits in a regular fashion. During her follow-up period, she had lung metastasis at 12th month and she died at 14th month with extensive systemic dissemination of the disease.

Discussion

Despite the low incidence of malignant mesenchymal tumors of the breast, they may proceed aggressively leading to mortality with local recurrences and systemic metastases. Because of the small number of case reports in the literature, there is still no agreed consensus about the treatment options of these tumors.

Surgical resection of the mesenchymal tumors is the first choice. The most important factor effecting the local control and survival rates is the achievement of a complete microscopic resection of the tumor (R0 resection) (2-5). Confavreux et al. reported that the survival rates of the patients that underwent successful R0 resection was calculated to be 72% while of those in whom this vital necessity couldn't be achieved was as low as 38% (6). Blanchard et al. also reported that any patient who underwent resections with positive surgical margins developed local recurrences. On the other hand, they also stated that only 42% of the patients those were treated with successful R0 resections developed local recurrences (4). However, the extent of the surgical procedure does not effect the recurrence or survival rates. To achieve the R0 resection, the extent of surgery can vary from local excisions to radical mastectomies depending on each individual patient and their lesions features (2,4-6).

Adding the axillary disection to surgical procedure depends on presence of palpable axillary lymph nodes. Because, these tumors usualy spread hematologically, lymphatic spread is very rare (7-9). Barrow et al. and Shabahang et al, reported that they performed axillary dissections to 35 and 10 patients in their series and they detected no evidences of any axillary metastases (2, 10). McGowan et al. stated that there was only 3 patients (8%) possessing axillary lymph node involvement in their 36 cases series as well (3). Also, in our case, despite of extensive invasion to the deep structures and palpable lymph nodes in the axillary region, no axillary metastases was detected.

Controversies exist concerning the effects of adjuvant chemotherapy and radiotherapy on local recurrence and survival rates. Pandey et al. reported that adjuvant radiotherapy played a beneficial role on malignant phylloides tumor patients in terms of local recurrence and survival rates (11). However, this favourable effect couldn't be proven in large numbers of different studies carried out for the research of the mesenchymal tumors of the breast (1,3,6,12).

Blanchard et al (4). reported that the survival rate in their study group consisting of 34 patients who did not take any chemoradiotherapy was calculated to be 62% while this ratio was found to be 37% in their other group of 16 patients those underwent combination chemoradiotherapy. As a result of these findings, they stated that adjuvant therapy brought no beneficial effects

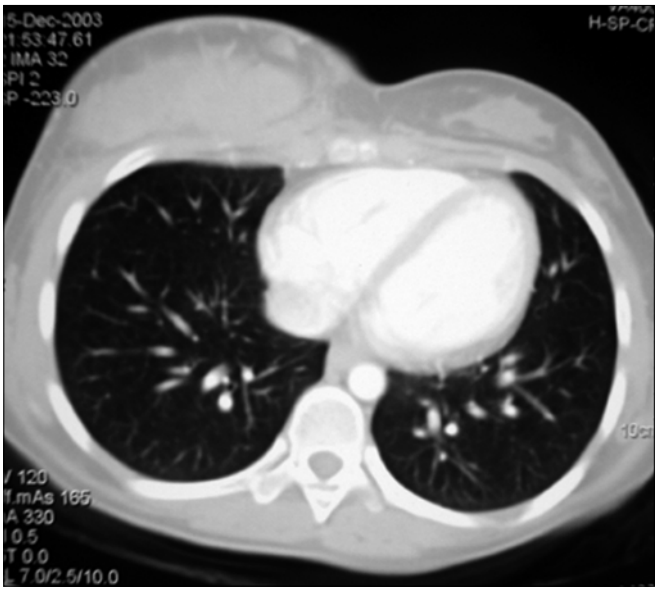

Figure 3. No evidence of any intrathoracic spread present.

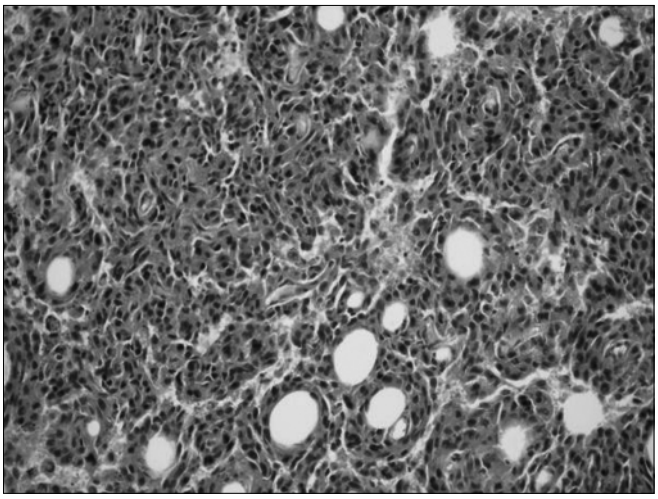

Figure 4. The microscopic appearance of malignant cellular structures originating from the mesenchymal tissue (H&E × 10).

in terms of survival rates and that this completion therapy should only be reserved for the patients having suspicious or positive surgical margins. Barrow et al.(2) reported that adjuvant radiotherapy did not play any positive roles on survival rates and that it might be reserved for elected patients for its probable beneficial role on local recurrences. In the same study, it has been stated that the most important factors effecting the local recurrence rates were the positivity of the surgical margins and the diameter of the tumor being greater than 5 cm. Also in this study the study groups of patients that underwent only mastectomy, mastectomy followed by radiotherapy, only segmental mastectomy and segmental mastectomy followed by radiotherapy revealed local recurrence rates of 34%, 13%, 25% and 0% respectively emphasizing that adjuvan radiotherapy could be indicated only for a number of selected cases. It can be considered as a conclusion statement that the effectivity of adjuvant therapeutic techniques are still obscure and further controlled prospective studies are needed to be carried out. In our patient, there was not local recurrence in 14 months of follow up, but adjuvant chemotherapy could not prevent systemic recurrence.

Factors effecting the local recurrence rates are determined variably in different studies. According to Blanchard et al. (4) R0 resection is the only favourable prognostic factor by itself while this parameter stands to be R0 resection and tumor diameter (< 2 cm) according to Barrow et al. (2) and R0 resection and histopathological findings (except angiosarcoma) according to Confavreux et al (6). The common conclusion of these studies is that the most important prognostic parameter is the complete microscopic resection of the tumor. In our case, the greater diameter of the lesion was found that13 cm, its histopathological diagnosis was indifferentiated malignant tumor and the nearest negative surgical margin is 1mm. She did not have local recurrences, but she had pulmonary metastasis and died at 14th month.

With the occasion of this case, we want to remind that, the possibility of mesenchymal malignant tumor in palpable breast lesions should be remembered, however thay are very rarely seen lesions.

Because of rarity of these tumors, prospective large studies are difficult in order to obtain the exact information of how the adequate therapeutic methods should be for the treatment of these malignant tumors. For this reason, it is of great importance that all diagnosed patients must be referred to breast centers in order to increase the experience for the management of these rare cases.

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