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Radical Surgery with a Multidisciplinary Approach in a Case of Radiation Induced Rhabdomyosarcoma of Breast Invading the Chest Wall, Lung and Heart

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Abstract

Sarcoma of the breast is usually seen following radiation therapy. Radiation-induced sarcoma treatment modalities include chemotherapy, radiotherapy and surgery. Even conservative treatment can achieve treatment response, locally advanced tumors should be removed with negative surgical margins. Unfortunately, the literature lacks of prospective data regarding the disease rarity. Clinicians should seek for individualised treatment options considered in multidisciplinary tumor boards. Here we present a radiation-induced chemotherapy resistant breast sarcoma patient successfully managed with en-bloc radical surgical removal and reconstruction under the provision multidisciplinary team.

Keywords: Breast Cancer, Breast Sarcoma, Radiation-Induced Sarcoma, Surgery.

Göğüs Duvarı, Akciğer ve Kalbi İnvaze Eden Radyasyona Bağlı Meme Rabdomiyosarkomu Olgusunda Multidisipliner Yaklaşımla Radikal Cerrahi

Öz

Meme sarkomu sıklıkla radyasyon tedavisini takiben görülür. Radyasyona bağlı sarkom tedavisi kemoterapi, radyoterapi ve cerrahiyi içerir. Konservatif tedavi ile tedavi cevabı elde edilse dahi, lokal ileri tümörler sağlam cerrahi sınırlarla çıkarılmalıdır. Ne yazık ki, hastalığın nadir görülmesine bağlı olarak literatür ileriye dönük veriden yoksundur. Klinisyenler multidisipliner tümör konseylerinin önerdiği bireyselleştirilmiş tedavi seçeneklerini araştırmalıdırlar. Bu olguda, multidisipliner bir ekibin kontrolünde radikal cerrahi ile en-blok çıkarım ve rekonstrüksiyon ile başarılı bir şekilde yönetilen kemoterapiye dirençli radyasyona bağlı meme sarkomu hastasını sunuyoruz.

Anahtar Kelimeler: Meme Kanseri, Meme Sarkomu, Radyasyona Bağlı Sarkom, Cerrahi.

INTRODUCTION

Breast sarcomas cover less than 1% of all breast cancers and less than 5% of soft tissue sarcomas (Russell et al., 1977). Radiation-induced rhabdomyosarcoma (RiRMS) is a rare clinical entity with unfavourable outcomes. The surgery is the mainstay of the definitive treatment. In this case report, we present the R0 resection approach to a chemotherapy resistance RiRMS patient with chest wall invasion under the provision of a multidisciplinary team including a breast surgeon, thoracic surgeon, plastic surgeon, and medical oncologist.

CASE REPORT

A 62-year-old female patient underwent a left lumpectomy and sentinel lymph node dissection for left breast cancer in 2011. Postoperative six cycles of FEC (5-fluoro-uracil; epirubicin; cyclophosphamide) chemotherapy and 45 Gy radiotherapy in 25 fractions were given as the adjuvant treatment. She admitted to the hospital with a complaint of a mass in the left breast in April 2021. A comprehensive breast examination and radiological evaluation revealed a 5.8 x 8.7 x 5.1 cm mass invading the chest wall and skin (Picture 1 and 2).



Picture 1: Physical examination image of the tumoral mass



Picture 2: Contrast-enhanced thoracic tomography examination image showing that the mass has invaded the chest wall and pleura

Tru-cut biopsy report was consistent with a malignant mesenchymal tumor. A whole-body scan presented no distant metastases. Neoadjuvant chemotherapy, including VAC (vincristine, adriamycin, cyclophosphamide) and IMA (Ifosfamide/Mesna/Adriamycin) regimens, were planned immediately. However, the tumor was progressive with no clinical signs of treatment response. The multidisciplinary tumor board considered surgery, and the patient was informed. A multidisciplinary team performed the operation, including a breast, thoracic and plastic surgeon. The mass and surrounding skin, anterior chest wall with left radical mastectomy, sternum including the left 3-4-5-6. ribs were resected en-bloc from the level of the costochondral joint on the right and the midclavicular line on the left (Picture 3 and 4).



Picture 3: En-bloc resection of left breast with radical mastectomy including pectoral muscles, ribs, pleura and pericardium (One clip represents Superior, two clip represents lateral margin)



Picture 4: The defect on the right hemithorax following the en-bloc resection

The pericardium and the parietal fascia of the lung were closed with dual propylene and polyglactin mesh. The defect on the chest wall was reconstructed with titanium plates (Picture 5). A latissimus dorsi (LD) flap with an ipsilateral skin islet was transposed over the reconstruction area. Split-thickness skin grafts harvested from the right thigh were replaced over the LD flap and surrounding area (Picture 6). Pathology reported an en-bloc tumoral mass with intact surgical margins. The tumor was 11.5x8.2x5.2 cm and invaded the breast skin, nipple, pectoralis major and minor muscles, ribs, parietal pleura, and pericardium. The final diagnosis was pleomorphic rhabdomyosarcoma associated with radiation toxicity (Picture 7 and 8).

The patient was taken to the ward with three Jackson-Pratt and one thoracic drain postoperatively. Postoperative follow-up was uneventful. She was discharged on the 14th day.



Picture 5: Anterior chest wall reconstruction with dual-mesh and titanium plates after en-bloc resection of the mass



Picture 6: Closure of the reconstructed area with composite LD muscle-skin flap and STSG flaps



Picture 7: Hematoxylin and Eosin (H&Ex200) staining of the specimen presents Bizarre cells with extensive eosinophilic cytoplasm, atypical appearance, prominent nucleoli regarding radiation associated breast rhabdomyosarcoma



Picture 8: Immune staining of the specimen with MyoD1 (x400)

CONCLUSION

Adjuvant radiotherapy-induced breast sarcomas may occur in long-term breast cancer follow-up. Although it's difficult to distinguish between radiotherapy-induced or primary breast sarcomas clinically, the appearance of dense cellular fibrosis, atypical fibroblasts, and sarcoma with impaired vascular structures in the pathological examination presents a differential diagnosis. RiRMS is identified as ionizing radiation exposure to the breast cancer area, related symptoms in at least four-year follow-up, and histopathological diagnosis (Sheth et al., 2012). Locoregional extended disease, including tumor invasion to surrounding structures with one or more foci, and lymph node involvement play an essential role both during the staging of the disease and the management of local treatment options in the chest wall located RiRMS. In addition, all patients considered for RiRMS should be evaluated for distant metastases. Surgical treatment is the only curative option in locoregional disease, and positive surgical margins increase the risk of local recurrence and death. Expected 5-year overall and disease-free survival rates in the advanced disease decrease to 49% and 44%, respectively.

Although radiation-associated sarcomas are rare tumors, their incidence tends to increase due to conservative treatment approaches. Breast sarcomas are the most frequent soft tissue tumors associated with radiation therapy (Brady et al., 1992). Chemotherapy and radiation therapy have limited beneficial effects over a specific subgroup of patients with sarcoma (Riad et al., 2012).

The surgical removal with clear margins, R0 resection, is the definitive and curative treatment of soft tissue sarcomas regarding lower recurrence rates (Lagrange et al., 2000). Locoregional disease without distant metastasis should be considered for complete resection of the tumor when necessary, under the provision of a multidisciplinary team to prolong survival for curative intent (Cha et al., 2004).

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