

Primary Chondroblastic Osteosarcoma of the Breast

Memenin Primer Kondroblastik Osteosarkomu

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ABSTRACT

Pure sarcomas of the breast are uncommon, accounting for less than 1% of primary breast malignant tumors. Mammary osteogenic sarcomas are very rare and less than 100 cases have been reported in literature. They mainly affect older and middle aged women and are highly aggressive. We report an additional case in a 56-year-old woman. Histological and immunohistological characteristics were similar to those described in other localizations. Differential diagnosis involves phyllodes sarcoma, breast metaplastic carcinoma with chondroid and osteoid differentiation, osteosarcoma of the ribs, and metastatic osteosarcoma. The prognosis is poor.

Key Words: Breast, Osteosarcoma, Sarcoma

ÖZ

Memede primer sarkomlar nadirdir ve primer malign meme tümörlerinin %1'inden azını oluşturur. Primer meme osteosarkomu olarak literatürde 100'den az sayıda olgu bildirilmiştir. Meme osteosarkomu orta-ileri yaş kadınlarda görülen oldukça agresif tümördür. Burada 56 yaşında kadın hastada saptanan primer meme osteosarkomu sunulmaktadır. Histolojik ve immünhistokimyasal özellikleri diğer lokalizasyonda saptanan osteosarkomlarla benzerdir. Ayırıcı tanıda filloides sarkom, kondroid ve osteoid farklılaşma gösteren metaplastik karsinom, kosta yerleşimli osteosarkom ve metastatik osteosarkom yer almaktadır. Prognoz kötüdür.

Anahtar Sözcükler: Meme, Osteosarkom, Sarkom

INTRODUCTION

Extraskeletal osteosarcomas are uncommon neoplasms representing less than 1% of soft tissue sarcomas. Mammary sarcomas are also rare, accounting for less than 1% of all primary breast malignant tumors. In this group, primary osteosarcomas are one of the least common sarcomas of the breast (1,2). Most cases have been reported in the literature as case reports except Silver et al who reported a retrospective analysis of 50 cases (3). With less than a hundred cases of primary mammary osteosarcoma reported in the literature (4) we report an additional case with this unusual tumor.

CASE REPORT

A 56 year-old postmenopausal woman, without clinical antecedent, presented with a breast lump that had rapidly grown for 7 months. On examination, breast palpation revealed a painless, mobile, hard, relatively regular shaped and large mass of 11x8 cm, involving all four quadrants of the right breast. There was no axillary lymphadenopathy. Ultrasound showed a heterogenous hypoechoic solid mass. Mammography showed a large hyperdense and round mass with calcifications (Figure 1). Biopsy was performed and showed a high-grade sarcoma with chondromatous

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differentiation. Preoperatively, the staging did not reveal primary or metastatic extramammarian sarcomatous tumor.

A mastectomy was performed. Grossly, a right mastectomy specimen measuring 18x12x5cm was received. The tumor was grey white, hard, with regular margins, measured 11x8.5x6 cm, and involved all four quadrants of the breast with small areas of haemorrhage, cysts and necrosis.

Histological examination revealed chondrosarcomatous proliferation, with foci of chondroid background containing atypical neoplastic cells. Furthermore, there were solid and compact sheets of spindle cells (Figure 2) producing an osteoid and bony matrix (Figure 3). No areas of epithelial component were identified despite achieving complete section slices of the tumor. Nipple areolar areas, skin and lymph nodes were free of tumor. On immunohistochemistry, tumor cells displayed immunoreactivity for vimentin. Positivity for S-100 and EMA was noted in chondroid areas (Figure 4). The spindle cell component was positive for smooth muscle actin but negative for desmin. Cytokeratin, p63 and CD34 were all negative. Thus, a diagnosis of primary chondroblastic osteosarcoma was made.

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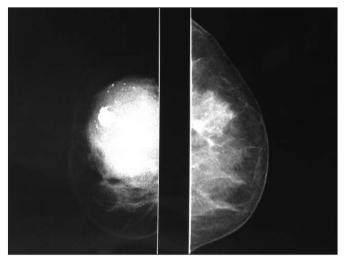


Figure1: Mammogram showing dense calcified lesion.

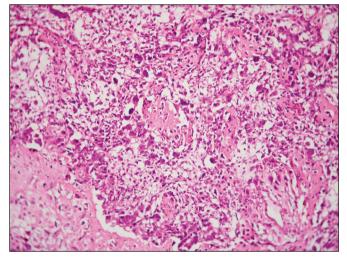


Figure 3: Atypical osteosarcomatous area with osteoid (H&E; x20).

The search for a primary bone tumor, especially in the rib cage, was negative. The patient presented with lung metastasis 2 months after mastectomy. She died 3 months later.

DISCUSSION

Pure sarcomas of the breast are uncommon, accounting for less than 1% of primary breast malignant tumors (5,6). More common breast sarcomas include fibrosarcomas, malignant fibrous histiocytoma, angiosarcoma, and liposarcoma (3,7). Primary mammary osteosarcoma of the breast represents 12.5% of breast sarcomas (5,8). Tumors with the following criteria are considered 'pure osteosarcoma': absence of bone origin, presence of osteoid or bony matrix, absence of an epithelial differentiation, and absence of a benign tumor

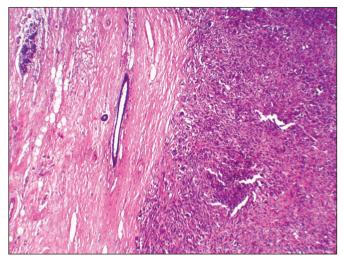


Figure 2: Tumor area showing regular margins (H&E; x10).

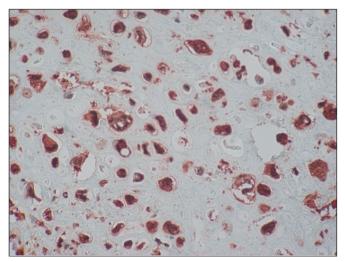


Figure 4: On immunohistochemistry cells of chondromatous component were positive for S-100 protein (S100; x40).

(9). The largest series, from the Armed Forces Institute of Pathology, Washington in the U.S.A., reported 50 cases, (3).

Mammary osteosarcoma predominates in middle-aged and older women (3,9). Most often, they arise as de novo conditions without predisposing factors especially after radiation therapy (3). The association between osteosarcoma and fibroadenoma was previously reported (4). The duration of symptoms is variable but most present within few months (3). All cases reported in literature had unilateral disease (5).

Clinical features, mammographic and macroscopic findings are not specific (10). Mammographically, these tumors are usually dense and well-circumscribed with focal or extensive coarse calcifications (5, 11). They may radiologically simulate a benign tumor (5).

Microscopic findings of primary mammary osteosarcoma are similar to skeletal and other extraskeletal osteosarcomas. Considerable diversity in morphological appearance has been reported with variants like fibroblastic, osteoblastic, osteoclastic, and chondroblastic; a variable amount of osteoid tissue and bone are present in all (5). Chondrosarcomatous differentiation is unusual (3,10) but was extensive in our case.

The histogenesis of this entity is unclear. It probably develops from totipotent stromal cells or a transformation from pre-existing fibroadenoma or phyllodes tumor (3,5). The main differential diagnosis is malignant phyllodes tumor with osteosarcomatous component but also metaplastic carcinoma, osteogenic sarcoma arising from the underlying ribs or sternum, and metastatic osteosarcoma (3,9).

In localized forms, the treatment is based on surgical excision with clear margins. Lymph node metastases are exceptional so that lymphadenectomy is not indicated (12,13). There is controversy regarding the use of chemotherapy. For some authors, it is not recommended especially in localized and well-resected low-grade tumors (13). Regarding metastasizing tumors, treatment is based mainly on chemotherapy using conventional drugs for osteosarcoma (doxorubicine, ifosfamide, cisplatinium, methotrexate) (12,13).

The prognosis is poor and the five-year survival is 38%. Recurrences are less frequent in patients treated with mastectomy than those treated with local excision. Metastases occur mainly in the lung; there is no axillary node involvement in almost all cases (9). Cases of death occurring a few months after the diagnosis have been reported (1,4,6).

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