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Case Report

A rare case report of breast sarcoma ☆

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ABSTRACT

Malignant tumors that originate from the mesenchymal tissue of the mammary gland, known as breast sarcomas, are very rare and can be divided into 2 types: primary and secondary (therapy-related development). Breast sarcomas are aggressive tumors associated with a poor prognosis. Treatment options include the coordination of surgery, chemotherapy, and radiotherapy. We present a case of a 51-year-old female who presented to the hospital after noticing a palpable mass in the left breast and bloody nipple discharge. These symptoms lasted for more than 4 months. Postoperative histopathology revealed an undifferentiated pleomorphic breast sarcoma. After 8 months of treatment, the patient experienced metastasis to the brain and lungs.

Introduction

Primary breast sarcoma is extremely rare, accounting for less than 1% of all breast cancer cases [1]. Histologically, breast sarcoma can be classified into several subtypes, including fibrosarcoma, pleomorphic sarcoma, leiomyosarcoma, rhabdomyosarcoma, and angiosarcoma [2]. Undifferentiated pleomorphic sarcoma is a high-grade malignancy and represents fewer than 5% of all adult sarcomas [3]. Breast sarcoma typically affects patients aged 55-59 years [4]. Breast sarcoma is typically difficult to differentiate from other types of breast

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cancer based on the clinical and imaging results [1]. In this article, we highlight the prognosis of a rare type of primary breast sarcoma.

Case report

A 51-year-old female was admitted to the hospital due to bloody discharge from the left nipple. A physical examination revealed nipple retraction associated with a large, firm, and fixed lump in her left breast. A breast ultrasound revealed a large, heterogeneous echotexture located in the left breast. The mass featured indistinct margins and hypervascularity. Breast magnetic resonance imaging (MRI) revealed that the mass (measuring 8 × 4 × 9 cm) was hypointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 1A and B). The mass was characterized by restricted diffusion (Fig. 1C and D) and the marked enhancement of solid components, although central necrosis was not enhanced (Fig. 1E). The tumor invaded the chest wall and skin (Fig. 1E). The patient underwent a breast biopsy.

Histological examination revealed a diffuse, patternless arrangement of highly atypical spindle cells with multiple perivascular aggregates. Coagulative necrosis was abundant, and mitoses were numerous (Fig. 2). Immunohistochemical staining showed that the tumor cells were negative for epithelial (cytokeratin), mesenchymal (CD34, S100, smooth muscle actin, Myogenin), lymphoid (CD3 and CD20), and melanocytic (human melanoma black 45 [HMB45]) differentiation markers (Fig. 3). Negative cytokeratin and HMB-45 immunoreactivity excluded carcinoma and melanoma, respectively. Negative CD3, CD20, and CD34 immunoreactivity excluded T-cell lymphoma, B-cell lymphoma, and angiosarcoma, respectively. Negative myogenin, S-100 protein, and smooth muscle actin immunoreactivity excluded rhabdomyosarcoma, liposarcoma, and leiomyosarcoma, respectively. The Ki-67 index was as high as 70% (Fig. 3). Based on the histological features and the immunohistochemical study, a diagnosis of undifferentiated pleomorphic sarcoma was made. The patient was treated with radical mastectomy combined with both neoadjuvant and adjuvant chemoradiotherapy. She developed brain and bilateral lung metastasis after 8 months.

Discussion

Breast sarcoma is rare and aggressive. Undifferentiated pleomorphic sarcoma accounted for 10.5%-24% of all primary breast sarcomas [5]. Clinically, patients often present with a unilateral, rapidly growing breast mass [1]. The tumors may be very large, up to 40 cm [6]. Some patients may present with nipple discharge or thin breast skin.

Imaging techniques, such as mammography, ultrasound, and MRI, are incapable of distinguishing breast sarcoma from other tumors [6]. On mammography, the tumors often present a single oval of hyperdense tissue that can be either well- or ill-circumscribed, and calcification is rare [7]. On ultrasound, the tumors have indistinct margins and are hypoechoic and heterogeneous with internal vascularization [7]. On MRI, the

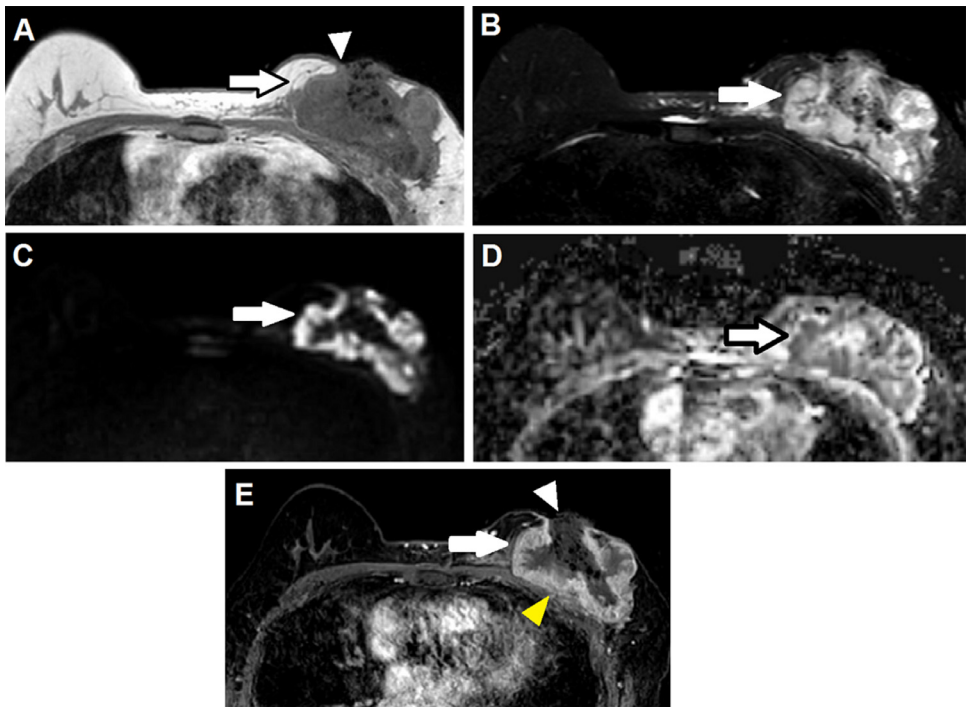

Fig. 1 – The mass was hypointense on T1-weighted images (A, arrow) and hyperintense on T2-weighted images (B, arrow), with restricted diffusion (C and D, arrows), and was markedly enhanced (D, arrow). The mass invaded the skin (A and E, arrowheads) and chest wall (E, yellow arrow).

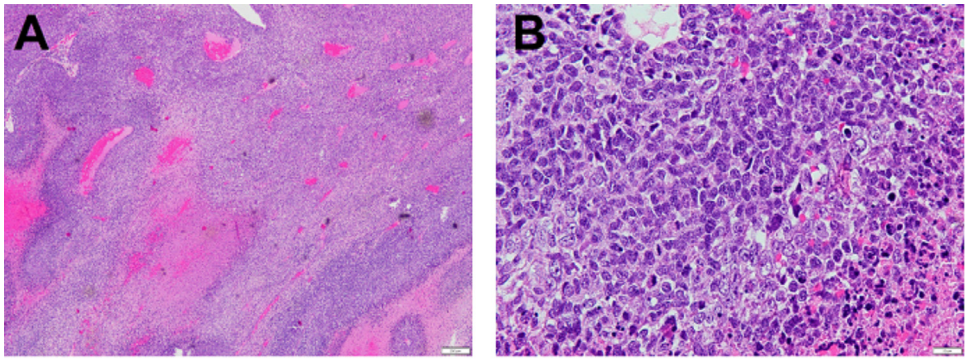

Fig. 2 – Hematoxylin and eosin staining showed a diffuse, patternless arrangement of highly atypical spindles and several necrotic regions (A). Mitoses were numerous (B).

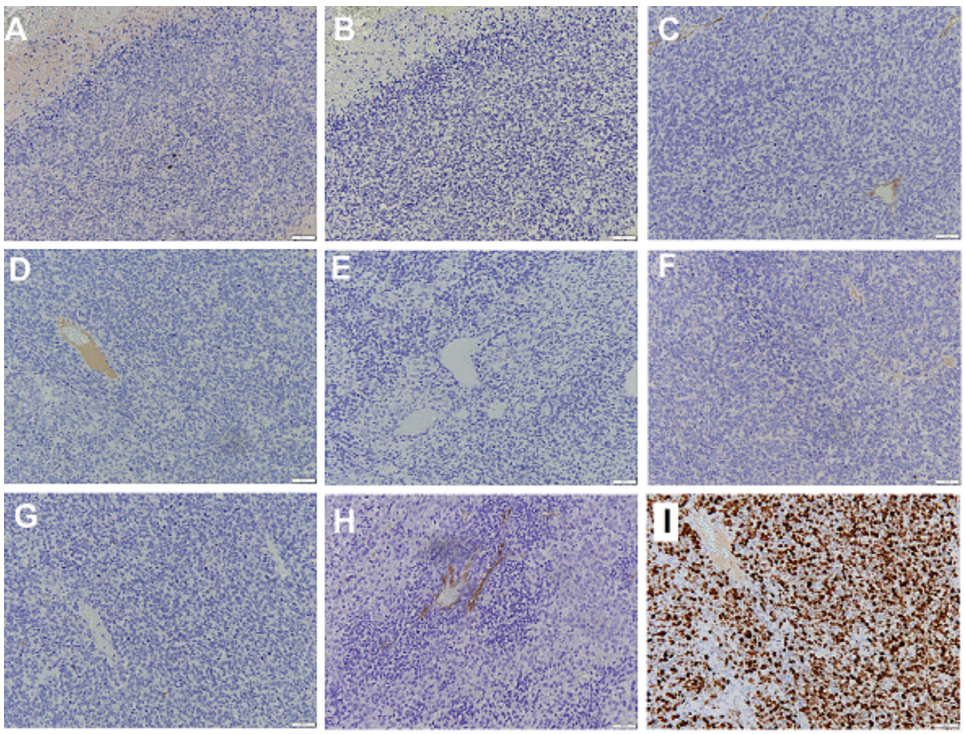

Fig. 3 – Immunohistochemical staining showed that the tumor cells were negative for the markers CD3 (A), CD20 (B), CD34 (C), CK (D), HMB45 (E), myogenin (F), S100 (G), and SMA (H). The Ki-67 index was 70% (I)

masses commonly present as hyperintense on T2-weighted images, with irregular margins and inhomogeneous enhancement [8]. In the present case, the tumor was very large, 10 cm in diameter at the largest point, and was hyperintense on T2-weighted images and hypointense on T1-weighted, with central necrosis. The solid components were characterized by restricted diffusion and were markedly enhanced. Imaging techniques may provide information regarding local invasiveness, current lymph node status, and distant metastasis.

Microscopically, the tumor cells showing marked pleomorphism, admixed with bizarre giant cells, spindle cells, and variable foamy cells [9]. A storiform growth or diffuse pattern and necrotic areas may be detected [5]. Immunohistochemistry may be useful for distinguishing breast sarcomas from nonmesenchymal malignant tumors and excluding breast sarcoma subtypes, such as leiomyosarcoma, rhabdomyosarcoma, and angiosarcoma [10]. In the present case, the final diagnosis was made by excluding other breast cancer types, including carcinoma, lymphoma, melanoma rhabdomyosarcoma, liposarcoma, and leiomyosarcoma.

According to the National Comprehensive Cancer Network (NCCN) clinical practice guidelines, preoperative chemother-

apy, radiotherapy, and chemoradiation, associated with surgery and adjuvant chemotherapy, are recommended for soft-tissue sarcoma treatment, depending on the tumor stage [11]. At the time of diagnosis, this patient was stage III. Treatment consisted of preoperative radiotherapy and chemotherapy, radical mastectomy, and adjuvant postoperative chemotherapy and radiotherapy. However, 8 months later, the tumor cells metastasized to the brain and lungs.

Tumors typically spread through local invasion or hematogenous spread, and the lungs, bone marrow, and liver are common metastasis sites [12]. Breast sarcomas have a high recurrence rate and poor prognosis [13]. Tumor size, histopathological type, histopathologic grading, the presence of positive margins, local recurrence, and margins status appear to be prognostic factors [14]. Tumors larger than 5 cm are associated with worse outcomes [8]. The median overall survival for breast sarcoma was 108 months, and the 5-year survival rate varies, ranging from 14% to 90% [4]. The overall 5-year survival rate is approximately 50% in patients with undifferentiated pleomorphic sarcoma [10]. The tumor in this patient was very large and had invaded the chest wall at the time of diagnosis. Multiple distant metastasis sites were detected, leading to a poor prognosis.

Conclusion

Breast sarcomas are rare and associated with a poor prognosis. Imaging modalities, including ultrasound and MRI, can assess the tumor stage, facilitating treatment decisions. This patient was diagnosed when the tumor was large and presented signs of invasion, resulting in a low survival rate. The histopathological tumor type and grading were also associated with poor prognosis.

Informed consent

Informed consent for patient information to be published in this article was obtained.

Ethical statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

Author contributions

Nguyen VS and Nguyen MD contributed to this article as co-first authors. All authors have read the manuscript and agree to the contents.

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