Human Pathology: Case Reports 7 (2017) 57–61

Contents lists available at ScienceDirect

Human Pathology: Case Reports

journal homepage: http://www.humanpathologycasereports.com

Malignant phyllodes tumor of the breast with heterologous high-grade angiosarcoma

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ARTICLE INFO

Article history:
Received 16 May 2016
Received in revised form 28 June 2016
Accepted 11 July 2016

Keywords:
Angiosarcoma
Breast
High grade angiosarcoma
Phyllodes tumor

ABSTRACT

Phyllodes tumors (PTs) account for < 3% of fibroepithelial breast lesions and for 0.3% to 1.0% of primary breast tumors. They occur predominantly in middle-aged women (mean age range, 40–50 years). PTs can be categorized into benign, borderline, and malignant; the first 2 categories are distinguished only by degree of cellular atypia and mitotic activity. Malignant PTs are more frequent among persons of Hispanic ethnicity, especially those born in Central America or South America. Heterologous sarcomatous elements may be present in malignant PTs, predominantly liposarcoma and rarely fibrosarcoma, rhabdomyosarcoma, leiomyosarcoma, osteosarcoma, and chondrosarcoma. Breast angiosarcoma (BA) is a rare heterologous, sarcomatous element that may arise secondary to malignant PT. We report a 47-year-old woman with no history of previous surgery or radiation therapy who presented to the emergency department with a painful right breast mass. She admittedly noticed the right breast mass for many years; however, recently it increased in size. Mammography and ultrasonography identified a partially cystic mass. Core needle biopsy showed dense hyalinized fibrous tissue with old blood clots, suggestive of infarcted fibroadenoma. The patient received antibiotics and analgesics; however, she reported intractable pain and a worsening skin rash of her right breast. Chest computed tomography and magnetic resonance imaging showed a doubling in mass size, with pectoralis major muscle involvement. Incisional biopsy showed malignant PT with heterologous high-grade angiosarcoma. The diagnosis of angiosarcoma was confirmed through immunoreactivity for CD31, FLI1, and ERG immunostains.

1. Introduction

Phyllodes tumors (PTs) are rare fibroepithelial lesions and are considered true neoplasms [1,2]. They historically have been given many names that are misleading and unhelpful. Thus, the term *cystosarcoma phyllodes* is no longer used [2]. The name *phyllodes tumor* is derived from the Greek word *phyllos*, meaning *leaf*. Gross examination shows that PTs can have variable sizes ranging from a few centimeters to 20 cm (mean, 4–5 cm) [2]. Benign PTs are well defined and often lobulated, whereas borderline and malignant PTs are ill defined [2,3]. Microscopic examination shows cellular stroma lined by an epithelial and myoepithelial bilayer, forming a leaflike structure. A focally infiltrative border can be seen in borderline PT [2,4]. Malignant PTs show a prominent infiltrative border, unequivocal sarcomatous areas, and stromal overgrowth [2,5]. Heterologous sarcomatous elements occasionally may be present in malignant tumors [2]. By comparison, angiosarcoma is a rare heterologous sarcomatous element that could arise secondary to malignant PT [2,6].

Breast angiosarcomas (BAs) are rare malignant tumors arising from endothelial cells or their precursors [7]. They comprise about 0.05% of all breast cancers, but they are the second most common mesenchymal malignancy of the breast after malignant PT [8]. Primary or idiopathic BAs are more common in young premenopausal women and are present as a deep breast mass [7,8]. Secondary BAs develop in older postmenopausal patients, frequently 5 to 6 years after radiotherapy, and they are more superficial and usually present as a rash or bruising [9,10]. Radiation treatment of breast cancer has increased the risk of BA of the skin by 9 times [10]; however, radiation treatment for Hodgkin disease increased the risk of breast cancer but not of BA [4]. After breast and axillary surgery, the arm with complicated lymphedema at the surgery site has increased risk of BA of the skin, known as *Stewart-Treves syndrome* [8–11].

We describe a case of high-grade angiosarcoma secondary to malignant PT in a middle-aged woman.

2. Case presentation

A 47-year-old woman with no history of alcohol or tobacco use or radiation exposure presented to the emergency department with a painful right breast mass. She admitted noticing a right breast mass for many years; however, recently it increased in size. The mass invaded

Abbreviations: BA, breast angiosarcoma; MRI, magnetic resonance imaging; PT, phyllodes tumor.

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http://dx.doi.org/10.1016/j.ehpc.2016.07.002

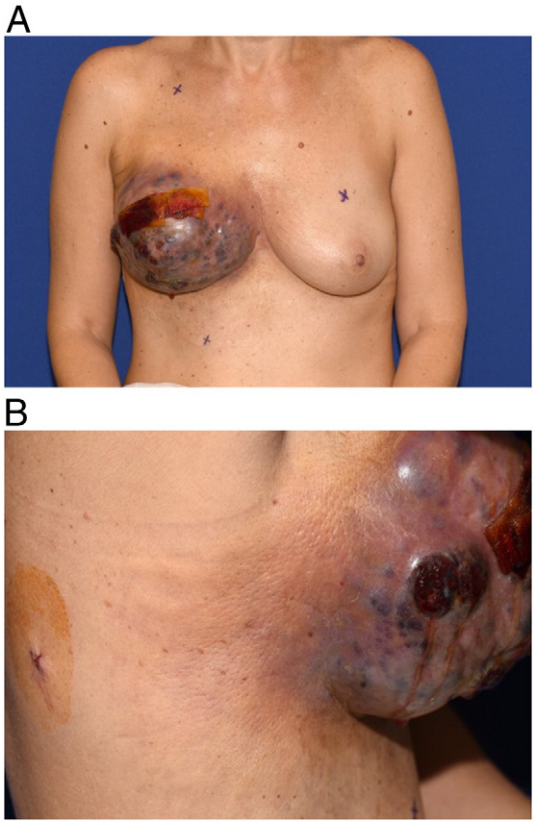

Fig. 1. The right breast with skin changes. A, Skin changes with ecchymosis. B, Erythematous plaques and surface ulcerations.

the overlying skin, deformed the right nipple (Fig. 1A and B), and invaded the right pectoralis muscle.

2.1. Radiologic evaluation

Mammography and ultrasonography identified a partially cystic mass measuring 4.8 × 6.6 × 5.9 cm (Fig. 2A). Ultrasound-guided aspiration revealed 35 mL of old blood, presumed as a hematoma. Core needle biopsy showed dense, hyalinized fibrous tissue with old blood clots, suggestive of an infarcted fibroadenoma. The patient was treated with antibiotics and analgesics. However, the skin ecchymosis and erythema were increasing and covering almost the entire right breast, and the breast pain was intractable. The patient had chest computed tomography and magnetic resonance imaging (MRI) at 2 months after her initial presentation. The imaging showed multicystic complex, heterogeneously enhancing mass in the right breast measuring 11.3 × 14 × 10 cm—double the size seen in the initial ultrasonography examination 2 months previously (Fig. 2B and C). The patient underwent incisional biopsy, which showed malignant PT with heterologous angiosarcoma.

2.2. Gross examination

The specimen was a 6.5 × 3.7 × 3.5-cm, 115-g right breast lumpectomy with a ragged cut surface. It consisted mostly of hemorrhagic tissue with ill-defined cystic lesion that had hemorrhagic contents. No skin or nipple was identified.

2.3. Microscopic examination

Multiple sections demonstrated a fibroepithelial lesion with extensive infarction and an associated malignant epithelioid and spindle cell

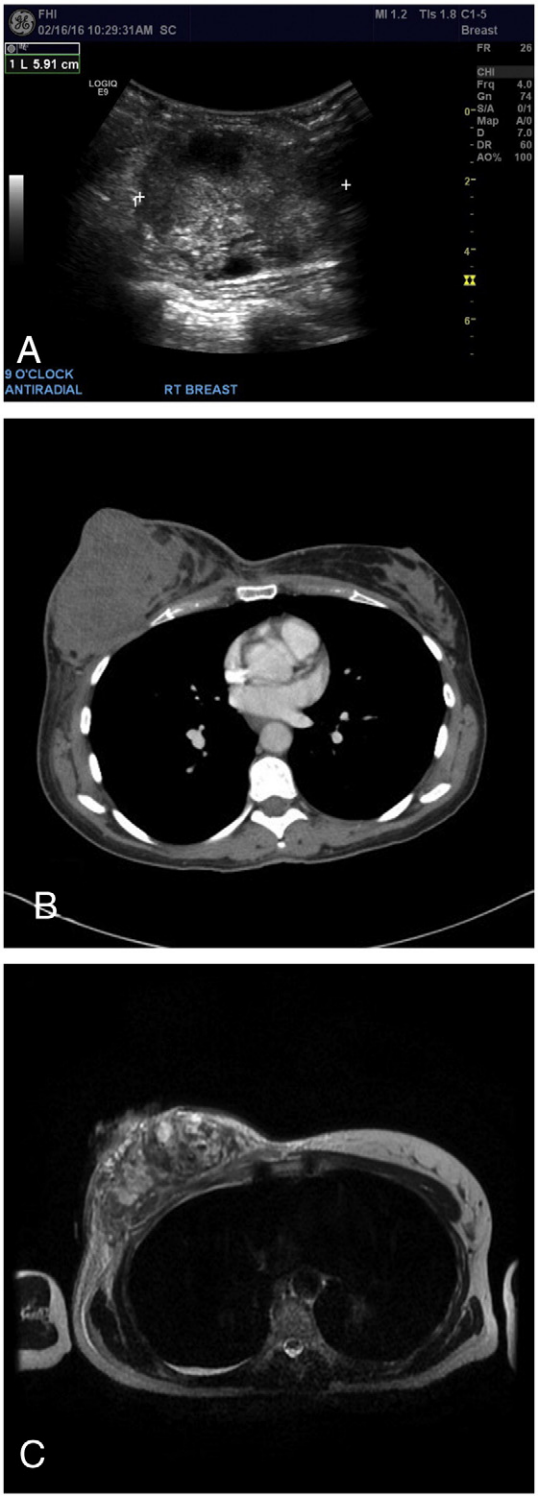

Fig. 2. Imaging of the right breast. A, Ultrasonography showing heterogeneous, partially cystic mass. B, Chest computed tomography scan showing the heterogeneous mass. C, Magnetic resonance imaging of the chest showing the mass with skin and pectoralis major muscle involvement.

proliferation involving the stromal component. The latter proliferation showed high-grade cytologic atypia with numerous mitoses and areas of extensive necrosis. Vague areas of vasoformative growth were seen, as well as prominent hemorrhage (Fig. 3A–D).

The differential diagnosis was between angiosarcoma secondarily involving a PT and malignant PT with heterologous angiosarcoma. Given the intimate association of angiosarcoma with the stromal