A Case Report of Breast Fibromatosis Mimicking Breast Cancer

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INTRODUCTION

Fibromatosis, also known as a desmoid tumor is a slow-growing, benign rare stromal tumor that accounts for approximately 0.3% of all soft tissue tumors [1]. A desmoid tumor commonly occurs in the superficial musculoaponeurotic tissue of the limbs and the abdominal wall [2]. The breast is a rare target site, comprising only 0.2% incidence of all breast tumors [3]. To our knowledge, the incidence of desmoid tumors affecting breasts has not been documented in the context of Indonesia.

On the physical examination, a firm mass with the inclusion of nipple retraction and skin dimpling is often found in a breast fibromatosis. While, in mammography examination, a spiculated mass that resemble malignancy is common [4].

Due to its rarity, by the time we submitted this case report, it is counted as the first case report of a breast fibromatosis in Indonesia. Taking into account the rarity with the clinical presentation and diagnostic imaging that often imitate a malignant tumor, we presented a case report of a breast fibromatosis in Onkologi Surabaya Hospital to emphasize further consideration to establish a diagnosis and adequate treatment.

CASE PRESENTATION

A 22-year-old unmarried woman with a palpable mass in the upper inner of her left breast one month before her visit, with no history of tenderness, nipple discharge, and skin dimpling was present and examined. She denied any history of trauma on her breast. The patient had no remarkable past medical history. Any abnormalities features including dental abnormalities, benign bone growth, skin abnormalities, polyps of the gastrointestinal tracts, and another history of cancer were also denied. Her menstruation period is regular, and her first menarche was at 12 years old. None of her family members had known health problems or any familial history of breast and ovarian cancer.

Due to its rareness, by the time we submitted this case report, it is counted as the first case report of a breast fibromatosis in Indonesia. Taking into account the rarity with the clinical presentation and diagnostic imaging that often imitate a malignant tumor, we presented a case report of a breast fibromatosis in Onkologi Surabaya Hospital to emphasize further consideration to establish a diagnosis and adequate treatment.
described an irregular echopoor solid mass sized 37 x 20 x 25 mm with an echoic part inside (Figure 1). The Color Doppler Ultrasound (CDUS) showed minimal flow with muscle infiltration, no evidence of edema, and no infiltration in the skin and nipple. Additional bilateral mammography with Cranial Caudal (CC) and Mediolateral Oblique (MLO) projections showed an irregular spiculate-like high-density mass in the upper inner of the left breast with pectoral muscle attachment (Figure 2). The conclusion was accorded Breast Imaging Reporting & Data System (BIRADS) 4C with high suspicion of malignancy. A core biopsy procedure was performed, and the result stated no sign of breast carcinoma, with a different diagnosis of fibroepithelial tumor favor to phyllodes tumor, and breast fibromatosis.

A further imaging examination was performed to determine the following treatment for the patient. The breast MRI with contrast suggested for malignant mass in the deep layer of the parenchyma at the inner upper quadrant, size 4.72 x 2.02 x 2.89 cm (mixed type II & III Time Intensity Curve (TIC) pattern – BIRADS MRI4 (Figure 3). Invasion of the mass to the adjacent pectoralis major muscle was noticed. MSCT thorax with contrast was added to evaluate the intrathoracic extension, and the result showed no intrathoracic involvement (Figure 4). She underwent an incisional biopsy, accompanied by a frozen section procedure. The result showed a benign spindle cell mesenchymal tumor, consistent with fibromatosis. The procedure was continued with a wide excision of the tumor and removal of the affected tissue to the muscle.
The histopathology examination revealed a poorly circumscribed tumor, infiltrating within breast stroma, fat, and muscle tissue. The tumor was composed of cells displaying oval to spindle-shaped nuclei and fine chromatin, arranged in long fascicles within a collagenous matrix. The tumor was lack of mitosis and necrosis when excision margins were uninvolved. Immunohistochemistry examination showed the tumor cells were negative for pancytokeratin, diffusely positive for vimentin, Ki67 proliferation index less than 1%, negative for CD34, and positive for β-catenin (Figure 5).

One week after the operation, the patient came for an evaluation. The operation site was in a good condition. Six months after the surgery, a physical examination of the surgical site was unremarkable.

**DISCUSSION**

Breast fibromatosis accounts for 0.2% of all breast tumors and 0.3% of all solid tumors [1]. It can occur in any age from 14 years to 80 years with the mean age at diagnosis ranges vary from 30 to 40 years old [2,5]. The etiology remains unclear, but it was associated with Gardner’s syndrome, silicone breast implants and surgical trauma have been reported in some studies [1,5,6].

Fibromatosis, also referred to as a desmoid tumor commonly presents as a painless, firm, and mobile mass. The average size of fibromatosis ranges from 2.5 cm to 3.0 cm. Several studies also mentioned that skin dimpling, retraction, or depression are often found during a physical examination [2,5,7]. WHO designated breast fibromatosis as an intermediate tumor, according to its locally aggressive and nonmetastatic characteristics. Due to their infiltrative appearance and lack of a defined tumor margin, macroscopically breast fibromatosis can readily be mistaken for invasive malignant tumors [8].

Imaging findings are characterized by irregular shapes, high density, and spiculate appearance that often-resembling malignant features. In ultrasound, fibromatosis presents as a poorly defined, hypoechoic mass with posterior acoustic shadow and echogenic rim [9–11]. Meanwhile, the MRI with contrast-enhanced of breast fibromatosis are varied and unspecific, including spiculated borders, linear-branching enhancement, and
rim enhancement. Ebrahim et al. [12] stated that the use of MRI in clinical practice is to look for possible chest wall involvement.

Cytology examination for diagnosing breast epithelial lesions is suggested and has high sensitivity and specificity [13]. However, the cytology evaluation in this present case which showed spindle cells is complicated because of the rareness and the heterogeneous conditions.

The morphological features are essential to establish breast fibromatosis. In addition to fibromatosis, Phyllodes tumor and metaplastic carcinoma should be considered in the differential diagnosis of spindle cell lesions of the breast. The lack of leaf-like patterns in microscopic features can eliminate the diagnosis of benign Phyllodes tumor for this case. The lack of nuclear pleomorphism and mitosis, in this case, can eliminate the diagnosis of a malignant Phyllodes tumor. The nuclear features of tumor cells that display bland spindle cells with a lack of pleomorphism and mitosis, favor a diagnosis of fibromatosis. However, Kuba et al. [14] stated that immunohistochemical examination could support the diagnosis of breast fibromatosis. Immunohistochemistry with pancytokeratin and vimentin can differentiate metaplastic carcinoma from fibromatosis. This case showed negative for pancytokeratin and diffusely positive for vimentin. The low index of Ki-67, negative for CD34, and immunoreactivity for beta-catenin in this case did not support for malignant Phyllodes tumor. The nuclear expression of β-catenin could be a helpful marker and was found in approximately 80% of cases of breast fibromatosis. However, this finding is not specific, because Phyllodes tumors can be positive for β-catenin in 60 to 95% of cases. The combination of β-catenin and the absence of CD34 could support the diagnosis of breast fibromatosis. Almost all normal breast stromal cells and stromal cell lesions express CD34, except fibromatosis and nodular fasciitis [15]. This present case showed negative CD34 and positive β-catenin. Therefore a diagnosis of breast fibromatosis can be established.

As described in this present case, malignancy was highly suspected based on imaging examination but was ruled out by pathology results of core biopsy tissue before the operation and frozen section procedure during the operation. Pathology diagnosis is important for giving appropriate treatment to the patient.

The treatment of choice for breast fibromatosis is wide local excision with clear margins and the removal of the skin, muscle, or fascia invasion. No predictive factor can be a reliable feature to determine the likelihood of recurrence. Local recurrence rates range from 21 to 27%. Recurrence rates can be significant, especially in cases when the margins are involved. If a wide excision is conducted and disease-free resection margins are ensured, recurrence is less likely. Therefore, close monitoring in the first three years after the operation is highly recommended [16].

CONCLUSIONS

Despite the low incidence and the indistinguishable imaging features from malignant tumors, further consideration and awareness of breast fibromatosis should be attained. A definitive tissue diagnostic before surgery is mandatory to prevent radical treatment for a non-malignant case.

DECLARATIONS

Competing of Interest
The authors declare no competing interest in this study.

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REFERENCES