

Synchronous Breast and Abdominal Fibromatosis: A Rare Case

Senkron Meme ve Abdominal Fibromatozis: Nadir Bir Olgu Sunumu

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ABSTRACT

Fibromatosis, also known as desmoid tumor, is a rare disease. Breast fibromatosis is extremely rare. Herein, we present a case of breast and abdominal aggressive fibromatosis in a woman to discuss optimal treatment management.

Key Words: Aggressive fibromatosis, breast, intraabdominal

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ÖZET

Fibromatozis; diğer adıyla desmoid tümör; nadir görülmektedir. Memede ise daha da az rastlanır. Burada memede ve karın içinde senkron olarak saptanan agresif fibromatozis uygun tedavi yaklaşımlarının tartışılması amacıyla sunulmuştur.

Anahtar Sözcükler: Agresif fibromatozis, meme, karıniçi

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INTRODUCTION

Fibromatosis, also known as desmoid tumor, is a rare disease. Breast fibromatosis is extremely rare and represents 4% of extra-abdominal desmoid tumors and 0,2% of all breast tumors. Aggressive fibromatosis (AF) arises from fibroblasts and it can be located at any part of the body. Although it is benign, it is locally aggressive. The cause of the AF is uncertain and can be related to trauma, hormonal factors or genetic association (1). The AF of the breast generally occurs in implant-associated breast or a history of trauma (often surgical) (2-8). Herein, we present a case of breast and abdominal AF in a woman to discuss optimal treatment management.

CASE REPORT

A 32-year-old woman presented with a palpable mass on right breast with a history of two weeks. On physical examination there was a painless, soft, palpable lesion, 2 cm in diameter with unclear borders. Ultrasonography revealed an irregular shaped, hypoechoic mass of size 18x15 mm at 1 o'clock direction of right breast, 10 cm away from the nipple. The lesion was categorized as BIRADS 5 and tru-cut biopsy was performed. Histopathological examination indicated a possibility of a low grade mesenchymal tumor (Fig 1).

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During staging work-up thorax tomography noted the involvement of pectoralis muscle and abdominal tomography revealed a similar-looking another lesion located on abdominal wall, right paraumbilical region and trunct biopsy was the same as breast pathology (Fig 2). Therefore the patient underwent wide local excision for both breast and abdominal wall and cavity. The breast lesion was excised with tumor free margins. Also, thin layer of pectoralis muscle was excised with the breast specimen. Macroscopically the tumor was 1.8 cm in diameter and immunohistochemically SMA, Vimentin, and Desmin were positive, and CD34, AE1/AE3 were negative.

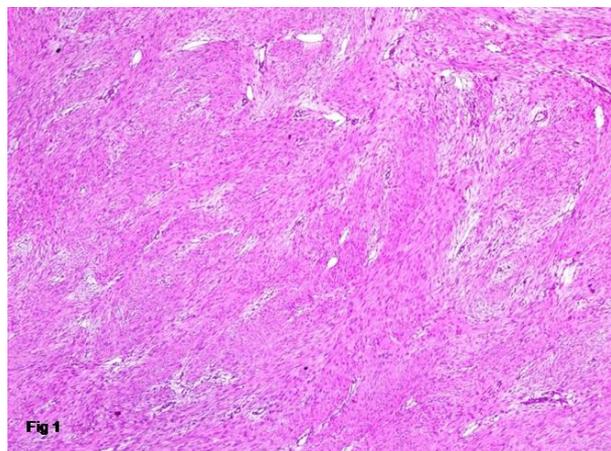


Figure 1: Fibromatosis: Tumoral lesion composed of long sweeping fascicles.HEx50, Breast.

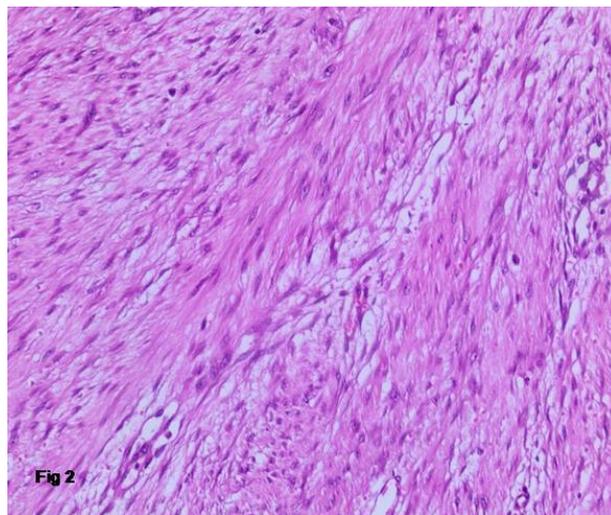


Figure 2: Fibromatosis: Uniform, bland spindle cells with pale, eosinophilic cytoplasm, poorly defined cell borders, oval to elongated nuclei.HEx200, Breast.

During abdominal surgery small intestine was explored to be involved in tumor from 120th cm of Treitz ligament. So, 70 cm small intestine was resected with tumor. The final histopathological findings were consistent with fibromatosis. The patient is under follow-up for 54 months without any treatment.

DISCUSSION

Aggressive fibromatosis is a rare fibroblastic proliferation arising in musculoaponeurotic structure. In women, most commonly occur after childbirth. The prevalence of AF is higher in patients with Gardner syndrome and familial adenomatous polyposis. Although histologically benign, desmoids are often locally invasive and associated with a high local recurrence rate after resection (1).

Aggressive fibromatosis can be seen at any body site but especially trunk/extremity, abdominal wall and intraabdominal (bowel and mesentery). In patients with familial adenomatous polyposis, intraabdominal desmoids predominate. Extra-abdominal AFs are generally located scapular girdle, abdominal wall, extremities, pelvic girdle, chest wall, trunk, head and neck region. Breast involvement is rare and its prevalence is 4% in the patients with extra-abdominal (10). The etiology of the breast involvement is uncertain. However, the patients with breast AF have the histories of breast cancer or breast surgery and familial adenomatous polyposis (2). The major presenting symptom is a palpable mass.

In our patient, the presenting symptom was breast mass. Ultrasound revealed irregular shaped hypoechoic mass with ill-defined borders which attached to the pectoralis muscle sonographically.

In the treatment of the AFs, if the patients have asymptomatic and stable disease, observation is suggested. The other patients are treated. The treatment choices are surgery (if possible, with negative margins), radiotherapy or systemic therapy.

In conclusion, breast AF is extremely rare, also to our knowledge this is the first case reported with concomitantly breast and abdominal AF in a woman. Although it is a benign entity pathologically, it can grow aggressively. More over this unusual lesion can display atypical radiologic features and mimic carcinomas. Optimal management of fibromatosis is radical surgical excision. Chemotherapy, radiotherapy and hormonal therapy have also been suggested.

Conflict of interest

No conflict of interest was declared by the authors.

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