



## Case report

## Desmoid fibromatosis of the breast; a rare case report

Zuhair D. Hammood<sup>a</sup>, Abdulwahid M. Salih<sup>a,b</sup>, Fahmi H. Kakamad<sup>a,b,c,\*</sup>, Ari M. Abdullah<sup>a,d</sup>,  
Bakhan Sharif Ali<sup>a</sup>, Lana R.A. Pshtiwan<sup>a</sup>

<sup>a</sup> Smart Health Tower, Madam Mitterrand Str, Sulaimani, Kurdistan, Iraq

<sup>b</sup> School of Medicine, University of Sulaimani, Sulaimani, Kurdistan, Iraq

<sup>c</sup> Kscien Organization, Hamid Str, Azadi Mall, Sulaimani, Iraq

<sup>d</sup> Sulaimani Teaching Hospital, Sulaimani, Kurdistan, Iraq

## ARTICLE INFO

## Keywords:

Desmoid tumor  
Case report  
Fibromatosis  
Breast cancer  
Local recurrence

## ABSTRACT

**Introduction:** Desmoid tumor is an uncommon tumor with variable spectrum ranged from being a locally lesion to an aggressive and destructive one. The current case aims to report a rare condition of desmoid type fibromatosis of the breast.

**Presentation of case:** A 59-year-old female presented with a right breast mass for 9-months. Mammography showed a small speculated iso-hyper dense mass, just anterior to the pectoralis muscle measuring about 15 mm (M5) in longest axis. Ultrasound examination revealed an irregular mass with internal vascularity and posterior shadowing in the right breast with a single borderline lymph node (25 \* 14 mm of 4 mm cortex). Wide local excision with sentinel axillary lymph nodes biopsy was performed. Histopathological examination of the specimen confirmed the diagnosis of desmoid type fibromatosis of the breast.

**Discussion:** The etiology of this tumor is unknown, however, physical, hormonal and genetic factors have a significant role in the development of desmoid tumor.

**Conclusion:** Desmoid type fibromatosis of the breast is an uncommon, benign, locally aggressive fibroblastic tumor with lack of metastatic potential, it may present with features of malignancy.

## 1. Introduction

Desmoid tumor is an uncommon tumor with a variable spectrum of activities ranging from a local lesion to an aggressive and destructive one. The term is derived from Greek terms "desmos" which means band or tendon like structure [1]. The incidence is approximately 0.2% of all neoplasms of the breast and 0.3% of all solid neoplasms [2]. Desmoid type fibromatosis is an aggressive sporadic condition having typical characteristics of a benign lesion that doesn't have metastatic potential, and characterized by a locally aggressive growth pattern with high possibility of local recurrence [3–5]. It is reported that this tumor may arise from within the parenchyma of the breast [6]. However others suggested that it arises from the musculo-aponeurotic structures that overlying pectoralis major muscle [7].

The current report aims to report a rare case of desmoid type fibromatosis of the breast. The report has been arranged in line with SCARE guidelines with a brief literature review [8].

## 2. Case presentation

## 2.1. Patient Information

A 59-year-old house wife patient presented with right breast painless mass for 9 months. She was gravida 7, para 7 and abortion 0. With three year history of lactation. Her past medical history was significant for hypertension and diabetes mellitus. She underwent lumbosacral spinal surgery for degenerative disc disease. She had used oral contraceptive pills in the past for several years (the type and dosages were not recalled). Family history was unremarkable.

## 2.2. Clinical Examination

The patient had right breast hard, mobile, irregular mass measuring 2 cm in longest diameter, associated with skin tethering and a palpable right axillary lymph nodes. She had normal weight (BMI = 23 kg/m<sup>2</sup>).

\* Corresponding author at: Doctor City, Building 11, Apartment 50, Sulaimani 0064, Iraq.

E-mail address: [fahmi.hussein@univsul.edu.iq](mailto:fahmi.hussein@univsul.edu.iq) (F.H. Kakamad).

<https://doi.org/10.1016/j.ijscr.2021.106363>

Received 22 May 2021; Received in revised form 30 August 2021; Accepted 31 August 2021

Available online 6 September 2021

2210-2612/© 2021 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

### 2.3. Diagnostic assessment

Mammography revealed a small speculated iso-hyper dense mass, anterior to the pectoralis muscle measuring about 15 mm (M5) suspecting fibromatosis (a pathological finding of myofibroblastic tumor). Ultrasound examination depicted a speculated mass (15 \* 9 mm) with speculated outline in the right upper outer quadrant near the axillary tail extended to the subdermal layer, associated with skin dimpling (subdermal attachment) (Fig. 1). The mass showed an irregular shape and margin with internal vascularity and posterior shadowing (U5). There was a single borderline lymph node (25 \* 14 mm of 4 mm cortex). Fine needle aspiration cytology (FNAC) from the right axillary lymph node showed a benign lymphoid cell.

### 2.4. Therapeutic intervention

Wide local excision (purely breast tissue without surrounding muscles) with sentinel axillary lymph nodes biopsy was performed. Histopathological examination of the specimen revealed Low-grade spindle cell neoplasm, suggestive of fibromatosis (Fig. 2). Immunohistochemistry tests showed positive control: P63: Negative, CK7: Negative, Trichrome: Blue stain for fibrous tissue. It was diagnosed as desmoid type fibromatosis of the breast.

### 2.5. Follow up

The patient developed post-operative pus collection in the operative bed, ultrasound guided aspiration was done. The patient was covered with antibiotics. The condition improved. The patient was healthy six months after the intervention.

## 3. Discussion

World Health Organization (WHO) defined desmoid type fibromatosis as an intermediate soft tissue tumor with the characteristic of clonal fibroblastic proliferation derived in the deep soft tissue with the ability of local infiltration [9]. It is composed of a bland appearing proliferation of spindle cells [10]. Desmoid tumors are divided into intraabdominal and extra abdominal desmoids. The most common site of extra abdominal desmoid tumors are the extremities [11]. Desmoid tumor of the breast is an extremely rare condition [12]. The age of the current case is 59 years which is much older than the peak age [20-40 years]. The etiology of this tumor is unknown, however, physical,

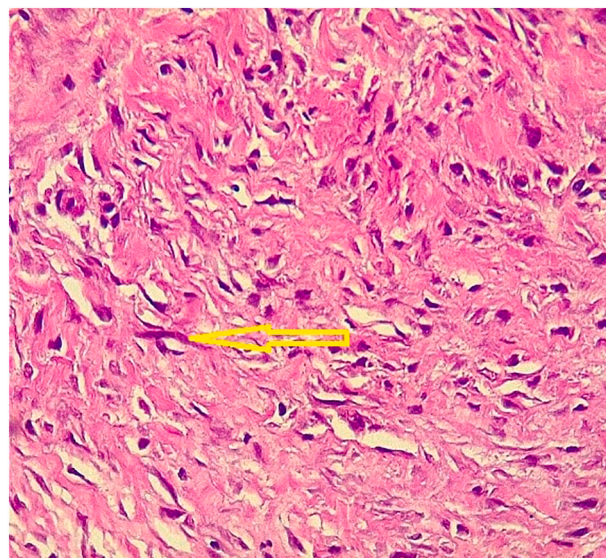


Fig. 2. Histopathological examination of the specimen showing band looking spindle cells (yellow arrow) in a fibrous stroma.

hormonal and genetic factors have a significant role in the development of desmoid tumor [14]. Desmoid tumor fibromatosis most often affects women during their childbearing age, often occurs after a trauma or different surgical procedures of the breast [15]. Augmentation mammoplasty with saline and silicone has been reported as a cause for the development of breast desmoid tumor [11]. Oral contraceptive pills and chemotherapy have been reported as the risk factors [11]. However, Benej et al. reported that there is no predilection of desmoids for age, family history or exposure factors [4]. The current case had no identifiable risk factor apart from oral contraceptive pills.

Desmoids have a clinical presentation that is similar to the breast carcinoma, which makes difficulty in differentiating this tumor with the carcinoma of the breast [16]. There is a variable degree in the presentations of the lesions, the most common one is skin tethering or retraction, hard lump, usually painless unless compressing the underlying nerve or tissue. Loss of function may occur if the tumor is fixed to the musculature [7]. The tumor may be mobile or fixed, skin dimpling may be present, it may lead to nipple retraction if it is close to it [2]. Rarely, it may present with bone erosion and invasion [7]. Current case

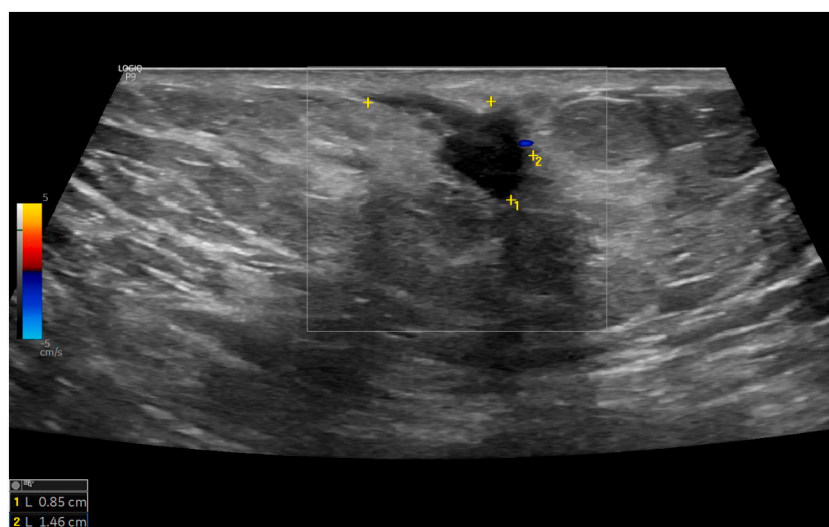


Fig. 1. Ultrasound examination showing a speculated mass with speculated outline in the right upper outer quadrant near the axillary tail extended to the subdermal layer.

presented with hard, mobile, irregular mass with skin tethering. Association with familial adenomatous polyposis has been reported as, regardless to the anatomical position, it is about one hundred times more common in patients with familial adenomatous polyposis than the general population [17]. Grossly the tumors might be mistaken as an invasive malignant neoplasm due to the characteristics of infiltrative appearance and absence of clear tumor margin [18,19].

Breast imaging techniques aren't specific for desmoid type fibromatosis, it is difficult to distinguish fibromatosis with breast malignancy through imaging [4]. However imaging and histopathological examination have significant role for planning of treatment and follow up [20]. On mammography, desmoids appear as an irregularly star shaped tumor, noncalcified, high density masses with speculated margins [13,17]. Computed tomography (CT) scan and magnetic resonance imaging (MRI) aid in defining the infiltration of the tumor to the adjacent tissue, particularly for patients with chest wall involvement [4]. Microscopically the tumor is characterized by fibroblastic proliferation, presence of abundant collagen fibrils and positive beta-catenin staining on immunofluorescence [18]. The hallmark of histological diagnosis of fibromatosis is the presence of proliferation of the bland looking spindle cells which is non-encapsulated and organized into long sweeping and intersecting fascicles with fingerlike extensions at the lesion's periphery into the adjacent of the parenchyma and adipose tissue of the breast [13].

Early diagnosis of desmoids and proper complete wide local excision is recommended with at least 3 cm negative margin [12]. However, complete excision is sometimes because of undesired negative cosmetic outcome [14]. There are guidelines recommending nonoperative management such as administration of non-steroidal anti-inflammatory drugs, hormonal therapy, radiotherapy, chemotherapy and tyrosine kinase inhibitors [18]. Although there is a controversy regarding the effect of radiotherapy, its efficacy is dependent on the dose; 60 to 80% control rate is expected [12]. Local recurrences have been reported in 24 to 77% of patients over a period of 10 years [16].

In conclusion, desmoid type fibromatosis of the breast is an uncommon, benign, locally aggressive fibroblastic tumor with lacking of metastatic potential, it may present with features of malignancy on physical examination and imaging. Have a high tendency to recur, early diagnosis and local excision is the treatment of choice.

#### Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Provenance and peer review

Not commissioned, externally peer-reviewed.

#### Ethical approval

Approval is not necessary for case report in our locality.

#### Funding

None is found.

#### Guarantor

Fahmi Hussein Kakamad is the guarantor of submission.

#### Research registration number

According to the previous recommendation, registration is not required for case report.

#### CRediT authorship contribution statement

Zuhair D. Hammood, Abdulwahid M. Salih: Doctors managing the case, follow up the patient, and final approval of the manuscript.

Fahmi H. Kakamad: literature review, writing the manuscript, final approval of the manuscript.

Ari M. Abdullah, Bakhan Sharif Ali, Lana R.A.Pshtiwan: major contribution to the idea, revision and final revision of the manuscript.

#### Declaration of competing interest

None to be declared.

#### References

- [1] S. Chummun, N.R. McLean, S. Abraham, M. Youseff, Desmoid tumour of the breast, *J. Plast. Reconstr. Aesthet. Surg.* 63 (2) (2010) 339–345.
- [2] B. Erguvan-Dogan, P.J. Dempsey, G. Ayyar, M.Z. Gilcrease, Primary desmoid tumor (extraabdominal fibromatosis) of the breast, *Am. J. Roentgenol.* 185 (2) (2005) 488–489.
- [3] S. Roussin, C. Mazouni, F. Rimareix, C. Honore, P. Terrier, O. Mir, et al., Toward a new strategy in desmoid of the breast? *Eur. J. Surg. Oncol.* 41 (4) (2015) 571–576.
- [4] R. Benej, I. Meciarová, K. Pohlodek, Desmoid-type fibromatosis of the breast: a report of 2 cases, *Oncol. Lett.* 14 (2) (2017) 1433–1438.
- [5] S.P. Povoski, W.L. Marsh, D.G. Spigos, A.E. Abbas, B.A. Buchele, Management of a patient with multiple recurrences of fibromatosis (desmoid tumor) of the breast involving the chest wall musculature, *World J. Surg. Oncol.* 4 (1) (2006) 1–8.
- [6] S.P. Povoski, R.E. Jimenez, Fibromatosis (desmoid tumor) of the breast mimicking a case of ipsilateral metachronous breast cancer, *World J. Surg. Oncol.* 4 (1) (2006) 1–5.
- [7] Y. Godwin, T.A. McCulloch, L. Sully, Extra-abdominal desmoid tumour of the breast: review of the primary management and the implications for breast reconstruction, *Br. J. Plast. Surg.* 54 (3) (2001) 268–271.
- [8] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, A. Thoma, et al., The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (1) (2020) 226–230.
- [9] N. Eastley, T. McCulloch, C. Esler, I. Hennig, J. Fairbairn, A. Gronchi, et al., Extra-abdominal desmoid fibromatosis: a review of management, current guidance and unanswered questions, *Eur. J. Surg. Oncol.* 42 (7) (2016) 1071–1083.
- [10] G. Magro, Differential diagnosis of benign spindle cell lesions, *World J. Surg. Oncol.* 11 (1) (2017) 1–8, <https://doi.org/10.1016/j.path.2017.09.005>.
- [11] C. Ioannidis, in: Extra Abdominal Desmoid Tumor (fibromatosis) of the Breast 17 (2), 2018, pp. 41–46.
- [12] S. Amourak, F.F. Alaoui, S. Jayi, H. Chaara, M.A. Melhouf, Fibromatose desmoide du sein: à propos d'un cas et Une revue de la littérature, *Pan Afr. Med. J.* 21 (88) (2015) 333.
- [13] W.L. Ng, S.Y. Teoh, M.H. See, K. Rahmat, P. Jayalakshmi, M.T. Ramli, et al., Desmoid type fibromatosis of the breast masquerading as breast carcinoma: value of dynamic magnetic resonance imaging and its correlation, *Eur. J. Breast Health* 17 (2) (2021) 197.
- [14] L. Pankratjevaite, I. Cesleviciene, L. Poškiene, A. Boguševicius, A case report of desmoid type fibromatosis of the breast, *Lietuvos Chirurgija* 18 (2) (2019) 107–111.
- [15] L. Wuys, A. De Schepper, Desmoid-type fibromatosis of the breast mimicking carcinoma, *J. Belg. Soc. Radiol.* 103 (1) (2019) 13.
- [16] A.R. Park, J.Y. Kim, H.J. Kim, J.H. Kim, M.J. Jeong, S.H. Kim, et al., Slow growing desmoid-type fibromatosis of the breast: a case report, *J. Korean Soc. Radiol.* 76 (4) (2017) 259–263.
- [17] J.S. Reis-Filho, F. Milanezi, L.Z. Pope, J. Fillus-Neto, F.C. Schmitt, Primary fibromatosis of the breast in a patient with multiple desmoid tumors: report of a case with evaluation of estrogen and progesterone receptors, *Pathol. Res. Pract.* 197 (11) (2001) 775–779.
- [18] N.C. Eastley, J. Krupa, R.U. Ashford, Desmoid (aggressive) fibromatosis of the breast, *Breast Cancer Manag. Surg.* 1 (1) (2018) 559–563.
- [19] K.Y. Ha, P. DeLeon, R. Hamilton, Breast fibromatosis mimicking breast carcinoma, *Baylor Univ. Med. Cent. Proc.* 26 (1) (2013) 22–24.
- [20] J. Lorenzen, M. Cramer, N. Buck, K. Friedrichs, K. Graubner, C.S. Lüher, et al., Desmoid type fibromatosis of the breast: ten-year institutional results of imaging, histopathology, and surgery, *Breast Care.* 16 (1) (2021) 77–84.