Case Report

Bilateral breast desmoid-type fibromatosis, case report and literature review

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Background: Breast desmoid-type fibromatosis (BDF) is a rare mesenchymal tumor accounting for only 0.2% of solid breast tumors. It is classified as an intermediate tumor because it is locally aggressive but has no metastatic potential. Its diagnosis is often difficult because it shares many clinical and radiologic aspects with breast carcinomas and therefore relies on anatomopathological analysis which may be supplemented by genetic analysis. The treatment of BDF has considerably evolved in the past years. While surgery was the cornerstone of the management prior to the 2000s, recent data have shown the value of active surveillance (AS) from the time of diagnosis. Indeed, after 2 years of AS, the progression-free survival (PFS) of the disease is identical or superior to surgery. Moreover, spontaneous regression has been observed in 30% of patients undergoing AS. In case of disease progression, surgery can be considered on a case-by-case basis, as well as systemic treatments.

Case Description: We present a case of bilateral BDF affecting a 20-year-old woman for whom the first suggested treatment was bilateral mastectomy with reconstruction. After a second opinion, the decision was revised and AS was initiated. Almost 3 years after the onset of AS, tumors have shown a continuous regression.

Conclusions: This case demonstrates the need for experience in the management of mesenchymal tumors to avoid overtreatment by mutilating surgeries which promote recurrence. Moreover, to our knowledge, very few cases of bilateral BDF have been published to date. It thus seemed relevant for us to report this rare case which supports the interest of AS for DF, as recently advised by the Desmoid Tumor Working Group guidelines.

Keywords: Desmoid-type fibromatosis (DF); breast; beta-catenin; active surveillance (AS); case report

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Introduction

Desmoid-type fibromatosis (DF) is a locally aggressive and non-metastasizing mesenchymal tumor arising from musculoaponeurotic structures accounting for 0.03% to 0.1% of solid tumors and 3% of mesenchymal tumors (1,2). It typically occurs in young adults from 20 to 40 years old and is about twice as common in women (1,3-6). There

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are intra-abdominal forms, mainly associated with familial adenomatous polyposis (FAP) and extra-abdominal forms, of which breast desmoid-type fibromatosis (BDF) accounts for about 4% (5,7,8). Bilateral cases are rare and account for around 1% of BDF (9). In 80% to 85% of cases, DF is sporadic and results from a mutation in the *CTNNB1* (Catenin Beta 1) gene encoding β-catenin. Whereas 5% to 15% of cases are familial forms associated with Gardner syndrome, a form of FAP linked to a mutation in the *APC* (Adenomatous Polyposis Coli) gene. As the development of DF seems more frequent in women, particularly during pregnancy, a potential role for estrogen in the pathogenesis of DF has been suggested (8,10). The main risk factors for DF are trauma and surgery and it is estimated in literature that 30% of DF cases occur in such context (7). Breast implants have also been identified as another important risk factor (11).

There are only 5 to 6 cases of DF per million inhabitants per year. Therefore, this rare tumor presents an important diagnostic challenge. In addition, despite its inability to metastasize, DF tends to locally infiltrate tissues and to recur (1,3-5,7,8,10,12).

The diagnosis is not always straightforward because of clinical and radiologic similarities with breast carcinomas, making pathological and immunohistochemical analyses crucial. Furthermore, the results must be read and interpreted by an expert in mesenchymal tumors (5,8,12). In addition to the diagnostic issues, we also face a therapeutic challenge resulting from the unpredictable behavior of this tumor in terms of evolution and recurrence (10).

Here is reported a case of bilateral BDF in a 20-year-old woman, which is rare as only few cases of bilateral BDF have been published so far (9,13-18). After reporting clinical findings in this patient, we will discuss DF features, as well as diagnostic elements and therapeutic recommendations, which have been updated in 2020, drastically modifying the former management of DF in the 90s. We present the following case in accordance with the CARE reporting checklist (available at https://gs.amegroups.com/article/view/10.21037/gs-22-271/rc).

Case presentation

A 20-year-old woman presented bilateral breast indurations discovered through self-examination. Three years earlier she underwent a surgical excision of a juvenile fibroadenoma in the right breast. Her family history is not known as she was adopted as a child. Her only medication is a contraceptive pill.

The breast examination revealed a mass estimated at 20 mm in the right upper outer quadrant and a large induration estimated at 50 mm in the left upper outer quadrant as well as skin dimples and nipple umbilication in the left breast. No adenopathy was palpated.

Mammographic examination (*Figure 1*) showed Breast Imaging-Reporting And Data System (BI-RADS) C breast density and bilateral areas of hyperdensity in the upper outer quadrants measuring over 15 mm in the right breast and 50 mm in the left breast. The ultrasound identified highly absorbing breast tissue and an irregular attenuating 35 mm mass in the left upper outer quadrant and a 14-mm mass in the right upper outer quadrant (*Figure 2*). This led to a BI-RADS 4C category in the left breast and a BI-RADS 4A category in the right breast thus requiring a histological sample. The anatomopathological analysis argues for a bilateral DF with negative hormone receptors, weak Ki67 expression (<1%), weak actin positivity and strong nuclear β-catenin positivity (*Figure 3*). The tumor is negative for desmin, Smooth Muscle Myosin Heavy Chain (SMMHC) and p63. An additional magnetic resonance imaging (MRI) with contrast was performed, but turned out uninterpretable due to background parenchymal enhancement. This phenomenon results from fibroglandular tissue enhancement on breast MRI and is influenced by exogenous and endogenous hormone levels (19) (*Figure 4*). However, MRI remains the most efficient exam to assess muscular and thoracic structures infiltration, which is absent in this case. It led to repeating the ultrasound which showed bilateral upper outer architectural distortions extending over 35 mm on the left and 45 mm on the right, demonstrating a stationary evolution on the left and an increasing evolution on the right within about 1 month.

The case was then discussed during multidisciplinary tumor board (MTB) and a bilateral mastectomy with reconstruction by prostheses was proposed to the patient. Almost six months after the MRI, a chest tomodensitometry was performed and showed a bilateral hypodense breast tumor, infiltrating the underlying deep fatty tissues and coming in contact with the pectoral muscles, without infiltrating them.

Due to the rarity of this tumor, a second opinion from a mesenchymal tumor expert was requested as well as a review of the histopathological analyses, which confirmed the diagnosis of bilateral BDF. One week before the surgery date, the therapeutic decision was revised and active surveillance (AS) was implemented. It consisted of clinical and radiological

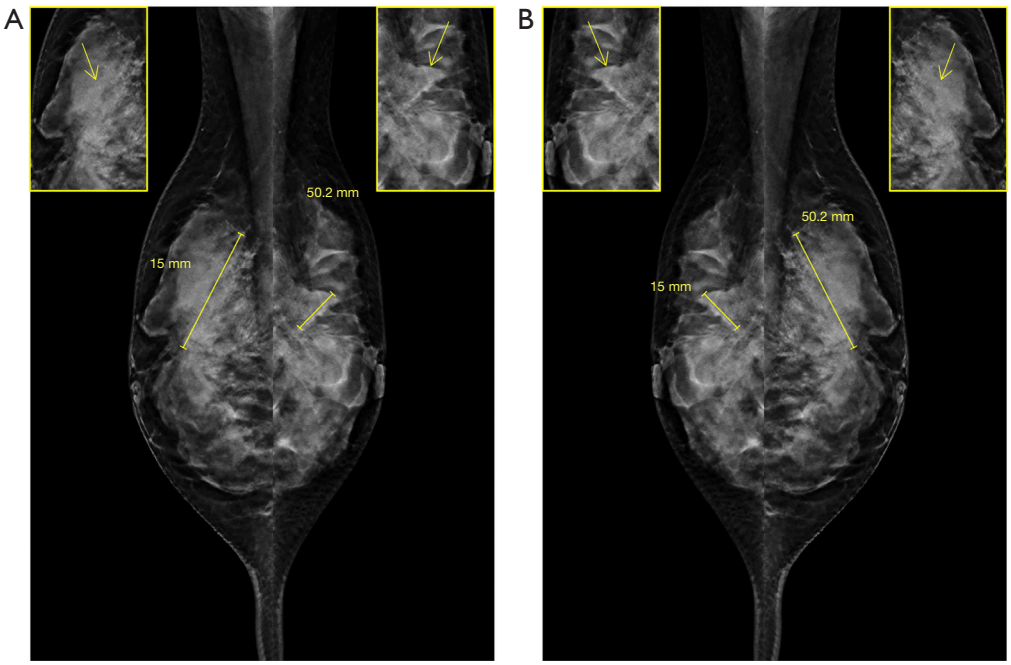

Figure 1 Mammogram at initial diagnosis. MLO views display BI-RADS C breast density. Craniocaudal views were not performed to minimize radiation due to the young age of the patient. (A) 2D s-views mammogram, right breast MLO images show a 15-mm hyperdense profound tumor in the right upper outer quadrant, left breast MLO images show a 50-mm hyperdense tumor in the left upper outer quadrant with a distortion image at its lower pole. (B) Digital breast tomosynthesis with a close-up view of each tumor. MLO, mediolateral oblique; BI-RADS, Breast Imaging-Reporting And Data System.

follow-up every 3 to 6 months mainly by ultrasound, as MRI is not an optimal technique in this patient. In order to increase the possibility of spontaneous regression, the patient was advised to stop her oral contraceptive pill. She was also referred for a genetic consultation and somatic molecular analysis did not reveal any somatic mutation of *APC* or *MUTYH* genes, which does not argue for FAP or *MUTYH*-associated polyposis (MAP). Furthermore, the screening colonoscopy did not show any polyp.

Almost 3 years after the initial diagnosis, the follow-up breast examinations showed a continuous regression of DF that reaches 73% and 21% in the left and right breast, respectively. Tumors are currently impalpable on clinical examination (*Table 1*).

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Differential diagnosis

DF accounts for less than 0.2% of breast tumors and therefore is not the first diagnosis considered on discovery of a breast mass (4). In addition, it shares many clinical, radiological and pathological features with breast tumors and tumor-like lesions, expanding the differential diagnosis (5,6,7,20,21). Indeed, DF can be confused with malignant lesions such as fibrosarcoma, breast adenocarcinoma and particularly spindle cell metaplastic carcinoma (12,21). The differential diagnosis also includes benign breast tumors such as myofibroblastoma, phyllodes tumor, locally aggressive tumors such as Darier-Ferrand dermatofibrosarcoma and reactive lesions such as nodular fasciitis or hypertrophic scarring (1,12,21,22).

How to discriminate a BDF from other breast lesions

BDF clinically presents as a deeply located, painless, firm and mobile mass. Skin and nipple retractions are

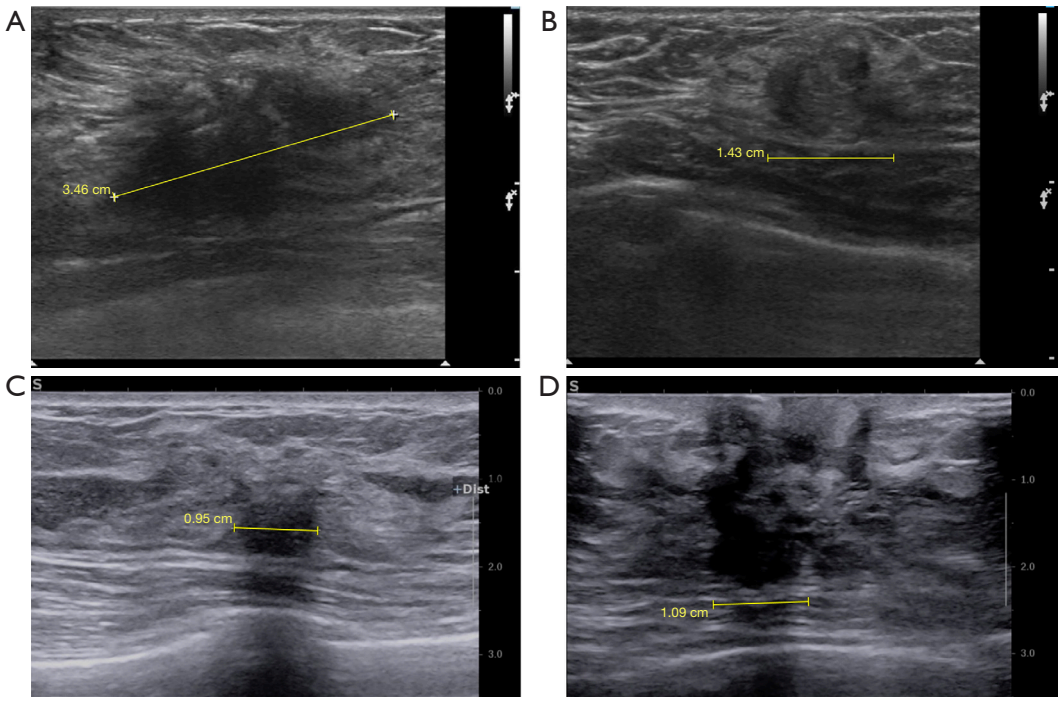

Figure 2 Transverse breast ultrasonography. (A) At initial diagnosis, ultrasound images of the left upper outer quadrant show a 35-mm irregularly shaped and attenuating mass. No adenopathy was found in the left axilla. (B) At initial diagnosis, ultrasound images of the right upper outer quadrant reveal a 14-mm mass. No adenopathy was found in the right axilla. (C) 2.7 years after diagnosis, ultrasound images of the left upper outer quadrant show a 9.5-mm hypoechoic mass. (D) 2.7 years after diagnosis, ultrasound images of the right upper outer quadrant reveal a hypoechoic mass measuring 11 mm.

possible (5,8). Its radiological features are non-specific and the mammographic and ultrasonographic appearance is often confused with a malignant process (6,8,21). Indeed, mammography usually shows a mass with spiculated, irregular borders and no calcification (6,23). On ultrasound, BDF appears as an irregular, hypoechoic mass with no associated adenopathy (5,6). The gold standard to assess BDF extension and to exclude chest wall infiltration is MRI (6,8,23). However, diagnosis based on imaging alone is impossible because of the infiltrating nature of BDF often leading to misdiagnosis of BI-RADS 4 or 5 (8,24). In this case, the clinical and ultrasound findings were crucial to establish 4A and 4C BI-RADS scores and to decide to perform biopsies. Indeed, due to the young age of the patient, a limited number of mammographic images were taken. It is why other techniques were preferred for the diagnosis of DF.

Histologically, DF develops from a monoclonal fibroblastic or myofibroblastic proliferation organised in cellular bundles surrounded by a profuse fibrous stroma (7,8,22). There is no sign of malignancy in the nucleus and cytoplasm as well as a low number of mitoses and no cell necrosis (12,25). To differentiate DF from other spindle cell lesions of the breast, immunohistochemistry (IHC) is a crucial step, particularly intranuclear β-catenin immunostaining for which approximately 80% of DF cases are positive (1,7,12,26). Unfortunately, this test is not very specific as other mesenchymal tumors show an accumulation of intranuclear β-catenin such as phyllodes tumor or metaplastic breast carcinoma (5,12). IHC uses other markers to clarify the diagnosis such as CD34, cytokeratins, and p63 for which DF is negative and actin for which it is positive (5,22).

The intracellular Wnt signaling pathway is central to the pathogenesis of DF, both in sporadic and familial forms (*Figure 5*). Activation of this pathway in mesenchymal cells causes translocation of β-catenin into the nucleus and initiates the transcription of proto-oncogenes responsible for cell proliferation (10,27). The *APC* gene is responsible for the phosphorylation of β-catenin, preventing its translocation into the nucleus and leading to its destruction

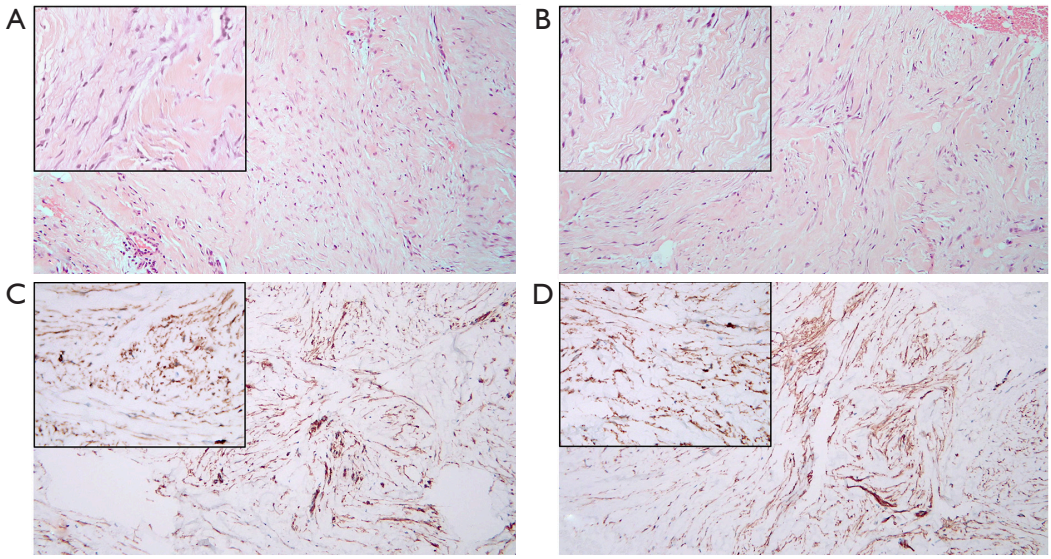

Figure 3 Pathological specimens of bilateral breast tumors after H-E staining and β-catenin immunostaining. Tissue samples were obtained by core needle biopsy. (A) Section of the right breast tumor after H-E staining at a 250-fold magnification shows bland spindle cells with regular nuclei arranged in broad, short fascicles with large collagen deposits intervening, all pushing aside the adjacent glandular tissue. (B) Section of the left breast after H-E staining at a 250-fold magnification. A close-up of this slide shows the profuse extra-cellular collagen deposition. (C) Section of the right breast tumor after beta-catenin immunostaining at a 400-fold magnification shows a strong nuclear β-catenin expression. (D) Section of the left breast tumor after beta-catenin immunostaining at a 250-fold magnification also shows a strong nuclear β-catenin expression. Close-ups at 400-fold magnification are displayed for each slide. H-E, hematoxylin-eosin.

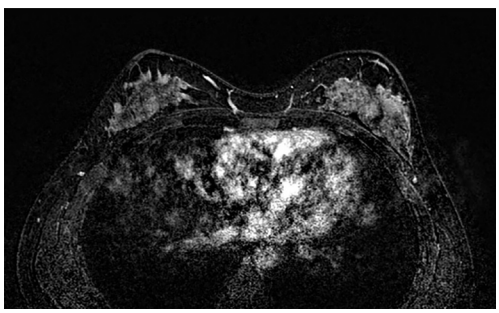

Figure 4 Breast MRI. T1-weighted dynamic sequence with subtraction technique. Subtraction images obtained 3 minutes after contrast (Dotarem®) injection demonstrate background parenchymal enhancement, which impairs desmoid-type fibromatosis identification. No infiltration of adjacent thoracic structures and muscles is displayed on subtraction images. MRI, magnetic resonance imaging.

in the proteasome (27). This explains why *APC* gene mutations, as well as activating mutations in exon 3 of the *CTNNB1* gene lead to intranuclear accumulation of β-catenin and ultimately to cell proliferation (1,7). Those mutations are mutually exclusive, meaning that detection of a mutation in exon 3 of CTNNB1 excludes APC mutation and vice versa (7,10,28).

If *CTNNB1* gene sequencing is not available or in case of *CTNNB1* wild-type status, an extended work-up by colonoscopy is justified. Especially in patients with risk factors such as a young age, abdominal DF or multifocal DF (7,10,28). In our case, the patient was screened by colonoscopy, justified by the lack of knowledge of her family history. Let us remind that the lack of specificity of β-catenin immunostaining is sometimes source of diagnostic uncertainty; therefore, searching for a mutation in the *CTNNB1* gene, which is more specific, is particularly useful (1). However, *CTNNB1* exon 3 mutation is less common (65–77%) in BDF than in other tumor sites (88%) (8,9). Hence, it is possible to diagnose sporadic BDF with no evidence of *CTNNB1* mutation (8). Finally, identifying mutations might be helpful in case of diagnostic doubt but mutation status does not predict systemic therapy responses (29).

Table 1 Timeline of the diagnosis and management process

Timeline	Diagnosis and management process
End of 2018	Bilateral breast indurations discovered through self-examination
Feb 18th 2019	Mammography and breast ultrasound
Feb 21st 2019	Core needle biopsy
Mar 3rd 2019	Anatomopathological results leading to breast desmoid fibromatosis diagnosis and MRI
Apr 5th 2019	First MTB leading to a bilateral mastectomy proposition
July 25th 2019	Second opinion from a mesenchymal tumor expert, histopathological results review and chest tomodensitometry
Aug 2nd 2019	Specialized MTB leading to active surveillance
Mar 3rd 2020	Genetic consultation
Nov 8th 2021	Latest ultrasound follow-up showing bilateral tumor regression

MRI, magnetic resonance imaging; MTB, multidisciplinary tumor board.

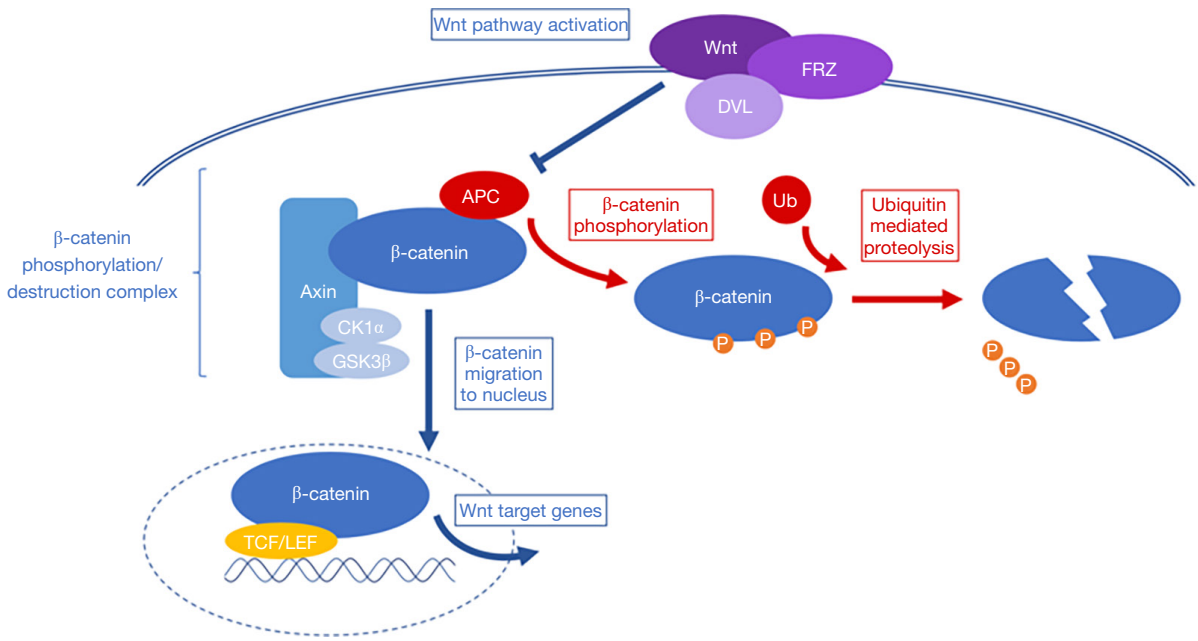

Figure 5 Intranuclear β-catenin accumulation can result from an inactivating mutation of the *APC* gene, which is responsible for β-catenin phosphorylation and degradation, as well as a mutation of *CTNNB1* exon 3, which encodes β-catenin phosphorylation domains. Adapted with permission from Desmoid Tumor Working Group (10). The management of desmoid tumours: A joint global consensus-based guideline approach for adult and paediatric patients. Eur J Cancer 2020;127:96-107. DVL, Dishevelled; FRZ, Frizzled; LEF, lymphoid enhancer factor; TCF, T-cell factor; Ub, ubiquitin; Wnt, int/Wingless.

Recent guidelines for the management of BDF

DF management was previously mainly based on surgery. But in recent years, literature has shown the effectiveness of an initial conservative approach, which has been largely confirmed by the Desmoid Tumor Working Group guidelines published in 2020 (10).

According to current recommendations, BDF management at the time of diagnosis should start with a period of AS. Indeed, this strategy has shown a 2-year progression-free survival (PFS) of 60%, which is equal or superior to surgical management (1,3-5,7,8,10,12,23,30). AS consists of patient follow-up preferably by MRI, within 4 to 6 weeks of diagnosis, at 3 to 6 months intervals for the first 3 years and then annually (8,10). Tomodensitometry, although less accurate in assessing BDF, is acceptable in cases of MRI unavailability or if the technique is inadequate, as in our case (10). Currently, initial medical treatment has not shown any advantage in terms of PFS compared to AS. Moreover, AS has a greater benefit in terms of clinical outcomes: less toxicity and morbidity, better quality of life. Initial medical treatment is therefore not recommended in the first-line setting (10). Besides its safety, this method also selects a minority of patients who will require medical or surgical treatment, avoiding many unnecessary mutilating surgeries (8). In addition to AS, an estrogen-free contraceptive method should be recommended to patients because of the suspected tumorigenic role of estrogen in BDF. Moreover, pregnancy is not recommended in the first year of diagnosis but is not contraindicated (8,10).

If radiological disease progression occurs or if symptoms' intensity increases during AS, evaluated by at least two further assessments and possibly not before one year from diagnosis in the absence of fulfilled RECIST progression criteria, medical or surgical management should be initiated (8,10,31). This switch to active treatment affects about 30% of patients and the risk factors are young age (<37 years old), large tumor diameter (>7 cm) and a painful tumor site, especially in the first 2 years after diagnosis (4,7,10). Medical management includes several treatment options such as tamoxifen, a selective estrogen receptor modulator used alone or along with non-steroidal anti-inflammatory drugs (NSAIDs), although the effectiveness of this treatment is controversial (5,7,8,10,26). For rapidly growing tumors, low-doses of chemotherapy can be used as methotrexate plus vinblastine or oral vinorelbine alone, which has shown a PFS of 80% at 1 year (7,10,26). For more aggressive diseases, chemotherapy with anthracycline-based regiments can be considered (7,10). The most significant data for systemic therapy in DF so far is a randomized controlled trial (RCT) published in 2018 assessing sorafenib, a tyrosine kinase inhibitor (TKI) which showed 81% PFS versus 36% in the placebo group with a follow-up of 27 months (32). This RCT has largely contributed to demonstrating the potential for spontaneous regression in DF, which is estimated at 30% (7,10,12,32). Sorafenib offers real clinical benefit in patients with progression (according to RECIST criteria) but unfortunately, TKIs are not yet included in standards of care for DF treatment (10,32,33). Some prospective studies have also assessed imatinib in progressive DF cases, which seems to have 65% progression arrest rate after 6 months with low response rates (6% to 19%) (12,32,33). However, as imatinib has not yet been assessed in a RCT, its actual efficacy is hard to determine and it should not be a first-line treatment for DF (10). The randomized, open-label, phase 2 DESMOPAZ trial evaluated Pazopanib (800 mg per day) versus intravenous chemotherapy (vinblastine-methotrexate) in progressive DF setting. The authors showed that the proportion of patients who had not progressed at 6 months was 83.7% and 45% with pazopanib and chemotherapy, respectively (10,34). In all cases, toxicity profile should always be considered and medical treatments have to be used gradually, starting first with the least toxic (10).

Despite DF high recurrence rate (20% to 40%) after excision, surgical management remains a treatment option in case of progression (5,7,8,10,22). The aim is to achieve complete resection with negative margins (R0). Positive margins (R1) are acceptable if the tumor is deeply infiltrative and the resection cannot ensure aesthetics or organ function (8,10). The relationship between positive margins and recurrence is controversial in the literature, but there is a consensus avoiding radical surgery technics (7,8). Adjuvant radiotherapy shows no benefit over surgery alone and is not recommended, especially in young patients because of the risk of radiation-induced sarcoma (7,8,10). On the other hand, radiotherapy alone is an appropriate option for symptomatic patients that are not eligible for surgery (7).

Lately, the management of BDF has made progress but could be further improved, in particular by conducting more RCTs and prospective studies and by developing precise criteria for AS setup (10). In addition, better identification of patients at risk of progression can be expected. Recently, detection of the *CTNNB1* gene in circulating cell DNA predicted DF evolution in 65% of cases and was able to

anticipate patients at risk of progression who could benefit from medical or surgical treatement (8,10). A key point to improve DF management is patient centralization to reference centers (10,35,36). Indeed, referral to a network of mesenchymal tumors specialists in France has shown to increase the number of diagnosed cases, mainly through second opinions, systematic second pathological reviews and molecular tests. It reduces the delay to diagnosis obtention and to the first oncological consultation. It also results in fewer invasive and costly procedures (35,36).

Conclusions

BDF is a very rare, locally aggressive mesenchymal tumor. Despite its inability to metastasize, it tends to locally recur and is readily confused clinically and radiologically with malignant tumors. Hence, histopathological analyses play an important role in the diagnosis and should always be performed by a pathologist with expertise in mesenchymal tumors. Histological diagnosis can also be supplemented by genetic analyses searching for *CTNNB1* or FAP mutations in case of diagnostic doubt. In the last years, management has been extensively reviewed and it is now recommended to initiate AS at diagnosis time in case of a primary, asymptomatic and stable tumor. BDF evolution is quite unpredictable, PFS rate can reach 60% with 30% of spontaneous regression. This further motivates the implementation of a conservative attitude. In case of disease progression, systemic therapy or surgical management should be considered while avoiding any functional or aesthetic sacrifice. The management of these rare tumors must be discussed during MTB specialized in mesenchymal tumors with network collaboration.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://gs.amegroups.com/article/view/10.21037/gs-22-271/rc

Peer Review File: Available at https://gs.amegroups.com/article/view/10.21037/gs-22-271/prf

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://gs.amegroups.com/article/view/10.21037/gs-22-271/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the editorial office of this journal.

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