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Case Report
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Dermatofibrosarcoma Protuberans in the Breast: Case Report

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Abstract

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon type of skin cancer that originating in the dermis. While it typically occurs on the trunk, DFSP in the breast is extremely rare and can appear similar to other breast tumors on imaging tests. This case study focuses on a middle-aged woman who had no previous health issues and visited the clinic due to an abnormal growth in her right breast that had been slowly growing for over the course of about 17 years. The medical team examined the clinical and physical features of the tumor, as well as considered different diagnostic tools and treatment options.

Keywords: Skin malignancy; Soft tissue sarcoma; Dermatofibrosarcoma protuberans; Breast surgery; Breast tumor

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a fairly infrequent soft tissue malignancy that originates from the dermis, it accounts for <1% of soft tissue malignancies and only <0.1% of all malignant tumors [1]. In 1924, Darier and Ferrand referred to DFSP as progressive recurrent dermatofibroma and the term Dermatofibrosarcoma Protuberans (DFSP) was first coined by Hoffman in 1925 [2]. It is classified as a low to intermediate grade malignancy that grows slowly rendering an early diagnosis very challenging until it enters a rapid growth phase [3]. DFSP is locally aggressive, this highly infiltrative nature often results in cosmetic and functional morbidity as well as a considerably high local recurrence rate. Despite the tendency of untreated DFSP to extend to local subcutaneous fat, fascia, muscles and even bones, it fortunately rarely metastasizes to distant structures [4].

The annual incidence of DFSP is less than 5 cases per million in the USA and the 5-year survival rate is up to 88.9%. It most commonly affects the trunk, proximal extremities, head and neck and the breast respectfully. People between ages 20 to 50 years old are mostly susceptible and it very rarely occurs in children [4,5]. The incidence among African Americans is almost twice as other races and the difference in incidence between males and females is still controversial. Initially DFSP appears as painless slow growing superficial nodules that often go unnoticed or ignored by patients [6].

DFSP of the breast is an extremely uncommon condition, with most cases occurring on the trunk. However, diagnosing it solely through imaging can be challenging due to its non-specific appearance [7]. It is important for clinicians to be aware of this rare lesion in breast skin to prevent misdiagnosing it as a benign breast condition.

This case study focuses on a middle-aged woman who had no previous health issues and visited the clinic due to an abnormal growth in her right breast that had been slowly growing for over the course of about 17 years.

Case Presentation

A 53-year-old woman came to the surgery clinic at Al-Amal Hospital in Khartoum, Sudan, with concerns about a mass in her right breast that had been present for 17 years. Recently, the mass had started to bleed two weeks before her visit. Initially, she noticed a small red area with itching, and she was given topical steroids which provided minimal improvement. She visited dermatological clinics multiple times but eventually ignored her symptoms. The lesion continued to grow and at the time of her visit to our clinic, there were multiple nodules located 2 cm below the nipple of her right breast. These nodules varied in size and were hard and oval-shaped with ulceration (Figure 1).

A breast ultrasonography revealed the masses originated from the skin, were partially vascular

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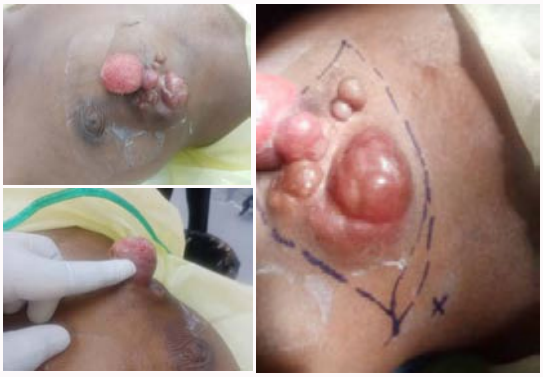

Figure 1: Physical examination findings at the time of initial presentation.

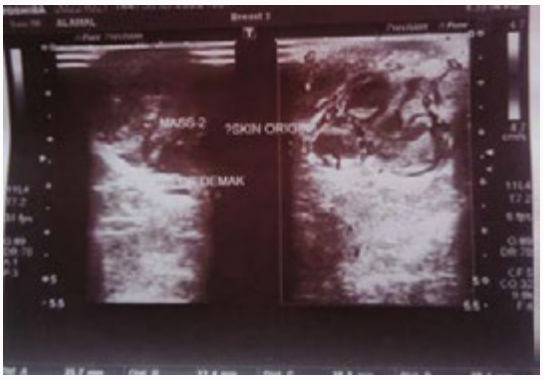

Figure 2: Ultrasound findings in the right breast.

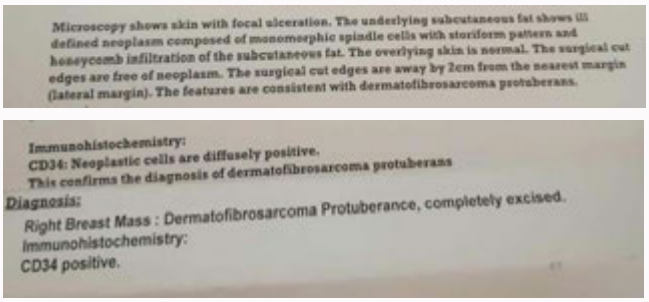

Figure 3: Histopathology report showed the microscopically findings and the surgical margin. And immunohistochemistry report.

and showed a heterogenic echo pattern, with the largest one measuring 4 cm × 3 cm × 3 cm, Figure 2. CT scan of the chest showed the extent of the mass was confined to the soft tissue, there was no distant metastasis.

Due to limited resources, the presence of ulcers in this specific area, and the patient's desires, a decision was reached after consulting with the oncologist to perform a surgical removal with an adequate safety margin in order to determine a diagnosis.

Wide Local Excision (WLE) was performed one week after presentation with a safety margin of 2 cm, no residual tissue was left and the specimen was sent to pathologist after marking carefully.

The diagnosis of DFSP was made after histopathological analysis of the excised tissue, which concluded a honeycomb like infiltration of the subcutaneous tissue by an ill-defined neoplasm, morphologically

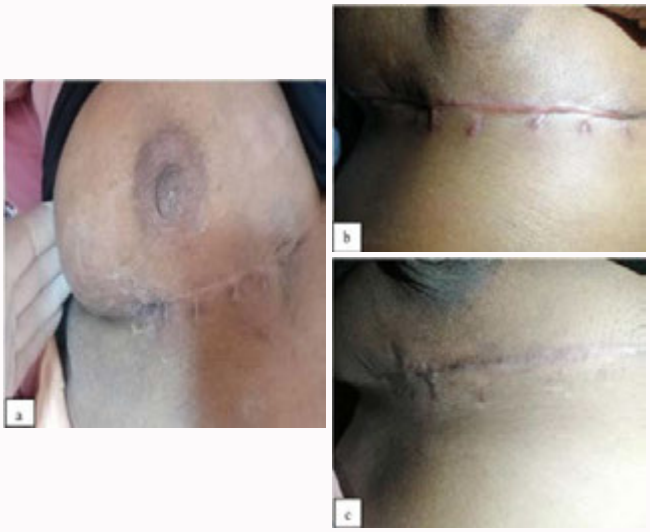

Figure 4: Postoperative wound healing. a) Two weeks post-surgery. b) Three months post-surgery. c) Nine months post-surgery.

consistent with dermatofibrosarcoma protuberans, with 2 cm free margins, and immunohistochemical staining of the tumor cells presented a diffuse, positive reaction for CD34 protein.

Unfortunately, due to the war in Sudan, we were unable to locate a digital version of the slides to include in this study. However, we have attached the reports for histopathology and immunohistochemistry (Figure 3).

The patient was discharged in a stable condition and referred to an oncologist once her wound had fully healed, then She underwent adjuvant radiotherapy. After 6 months, the patient returned for a follow-up appointment and no signs of local recurrence were found (Figure 4).

Discussion

Dermatofibrosarcoma Protuberans (DFSP) is a slow-growing, low-grade malignant tumor that typically starts in the dermis and spreads to deeper tissues. It is most commonly found on the trunk and extremities, but it is rare for it to occur on the breast [3]. DFSP of the breast can be mistaken for benign lesions like dermatofibromas and keloids, so it is important to differentiate it from fibroadenomas and phyllodes tumors [7]. The average age of presentation is around 38.5 years, with an almost equal distribution between males and females [8].

DFSP initially appears as a small, firm, painless plaque that gradually grows into raised nodules. Patients often ignore the symptoms for months or even years before seeking medical attention [9]. In this particular case, she had an enlarging skin lesion for years before seeking care when it became nodular and started to ulcerate.

The etiology of DFSP is still not fully understood. However, some studies have suggested a potential link between DFSP and various types of traumas. These traumas include surgical scars, direct physical trauma, burns, radiation exposure, vaccination punctures, venous central lines, dermatitis, and even insect bites [1].

Although imaging modalities can be misleading and unreliable in diagnosing DFSP, they can still be useful in assessing the extent of the tumor before surgery. This preoperative assessment helps in

determining the size and boundaries of the tumor, which can aid in planning the surgical procedure [7].

Wide Local Excision (WLE) is a widely used and reasonable approach for treating breast DFSP worldwide. It is crucial to determine the optimal width of the surgical margin around the primary tumor. Both the National Comprehensive Cancer Network guidelines [10] and the S1 guidelines [11] recommend a minimum surgical margin width of 2 cm to ensure histological margin control. Several studies have demonstrated that DFSP is responsive to radiotherapy, making it a recommended option for recurrent tumors or unresectable residual lesions to prevent disease progression [12]. A meta-analysis involving 167 DFSP patients treated with adjuvant radiotherapy showed that it could be considered for all patients undergoing surgical excision, regardless of the surgical margin. Although some cases of breast DFSP with negative margins have reported using postoperative radiotherapy, there is a lack of long-term follow-up data. Additionally, most DFSP cases have genetic connections, which supports the use of tyrosine kinase inhibitors like Imatinib as a treatment option [13].

Conclusion

DFSP in the breast is a rare occurrence, and imaging studies are crucial for determining the tumor's location and size before surgery. However, there is a lack of established criteria to differentiate DFSP from other tumors based on imaging findings. Histological examination is useful in identifying the unique features of DFSP. Therefore, surgical excision with sufficient margin at least 2 cm should be the primary treatment approach for DFSP, radiotherapy or Imatinib drug potentially used as adjuvant therapy. Long term follow-up is necessary, Due to its tendency for local recurrence.

Acknowledgment

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