Case Report

Recurrent dermatofibrosarcoma protuberans of the breast: A case report and review of literature

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare and low-grade sarcoma of fibroblast origin with a tendency to invade and recur locally. The most common sites of origin of DFSP are the head, neck, and extremities. However, DFSP breast has also been reported. It infiltrates surrounding subcutaneous tissue and fascia, with an incidence of <2% for lung metastases. Surgery being the main modality of the treatment, literature has shown that radiation therapy also plays an important role to improve local control in case of recurrent tumors. In this article, we will be discussing one such rare case of DFSP in a recurrent setting and the role of radiation therapy.

Key words: Breast, Dermatofibrosarcoma protuberans, Radiotherapy, Recurrent

Dermatofibrosarcoma protuberans (DFSP) is a rare and low-grade sarcoma of fibroblast origin with a tendency to invade and recur locally. It was first described by Darier and Ferrand in 1924 and named later by Hoffmann in 1925 [1,2]. The most common sites of origin of DFSP are the head, neck, and extremities. However, DFSP of the breast and vulva has been reported [3,4]. DFSP breast is a rare neoplasm that usually arises as a blue-reddish small subcutaneous mass that tends to grow slowly, infiltrating the surrounding subcutaneous tissue and fascia. It is generally an indolent lesion characterized by translocation of the COL1A1 and PDGFRB genes. It is usually seen in early or mid-adults, presenting clinically as a painless nodular cutaneous mass anywhere in the body that grows slowly. Although this tumor commonly recurs, it rarely metastasizes. The pathological diagnosis of DFSP depends on histopathological examination and immunohistochemistry (IHC) of the post-operative specimen. It is usually characterized by spindle cells arranged in a storiform pattern with little nuclear pleomorphism and mitosis. It is usually CD34 positive in 90% of the cases and negative to other markers such as S100, Actin, and Desmin [5,6]. The gold standard aim of the treatment in DFSP is surgery with the role of radiation therapy in certain situations like large or recurrent tumors or inoperable tumors [7].

Here, we report a rare case of recurrent DFSP of the breast in a 34-year-old female.

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CASE PRESENTATION

A 34-year-old female presented to our hospital with complaints of a lump in the left breast for 1 month with a history of similar complaints 3 years back which was evaluated and diagnosed as a case of DFSP and treated by surgery (wide local excision).

On examination, the previous surgical scar in the left breast was healed and healthy with 1×1 cm nodularity felt over the scar which was non-tender and had no palpable axillary lymph nodes.

She was evaluated with a sono-mammogram of the bilateral breast which showed an irregular heterogeneous hypoechoic lesion measuring 12×3 mm noted at cutaneous and subcutaneous planes at the previous post-operative scar with no vascularity and calcification. Multiple axillary lymph nodes were noted, the largest measuring 24×7 mm with maintained fatty hilum-suggestive of breast imaging reporting and data system II [8,9].

She underwent wide local re-excision of the previous surgical scar with underlying breast tissues down to the level of pectoral fascia with axillary lymph node dissection. Post-operative histopathological findings showed tumor involves the dermis and subcutaneous tissue, with mitosis 2–3/10HPF, no necrosis was noted and all margins were free from the tumor. All 15 Axillary lymph nodes were reactive and free from the tumor. Spindle cells were arranged in a storiform pattern in the subcutaneous plane, interspersed with blood vessels suggestive of spindle cell neoplasm of the left breast lump. IHC showed that the neoplastic cells were positive for CD34, focally MIC2 and BCL2 positive

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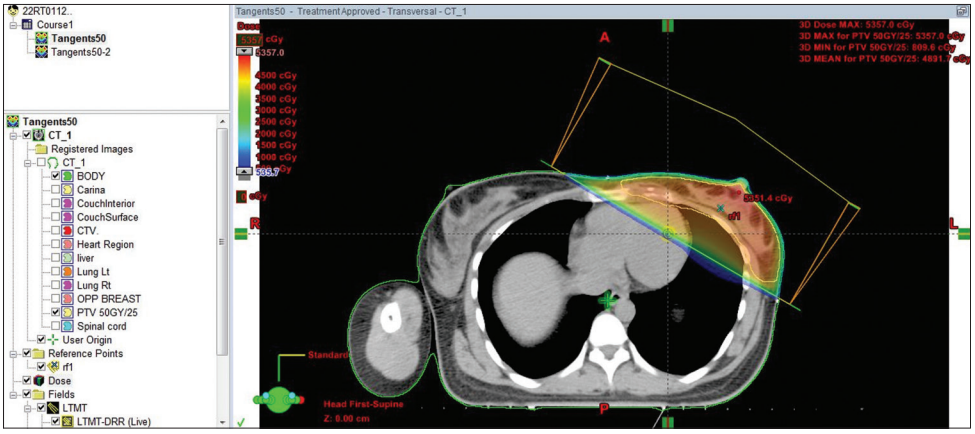

Figure 1: Treatment planning by 3DCRT technique using tangents

and negative for S100, EMA, SMA, DESMIN, and STAT6 with a Ki-67 index of 2%. Morphology and IHC features are suggestive of DFSP.

The case was discussed in the tumor board and in view of the recurrent setting and it was decided to plan for adjuvant radiation. She received external beam radiation therapy with 50 Gy in 25 fractions (2 Gy/fraction) to the left breast by 3DCRT technique, using medial and lateral tangents (Fig. 1). During the course of the treatment, she developed Grade 1 skin reactions which was managed conservatively. She has successfully completed the treatment and is on regular follow-up.

DISCUSSION

DFSP was first described by Darier and Ferrand [1] in 1924 and was termed by Hoffmann [2] in 1925. DFSP accounts for <0.1% of all malignancies and about 1% of all soft-tissue sarcomas [10]. DFSP typically occurs in the dermis and subcutis rather than in deeper soft tissue. The most frequently seen clinical feature is an asymptomatic multinodular bluish or brownish erythematous plate, with its typical 'protuberant' aspect, which develops over years [11,12]. The most commonly reported sites of DFSP are the trunk and extremities, with a benign aspect [13,14]. It is unusually reported in the neck and rarely in the breast where our case was detected [15]. The most potential clinical differential diagnoses for DFSP tumors include recurrent dermatofibroma, hypertrophic scars, keloid, skin manifestations of myofibroblastoma, metaplastic carcinoma, and fibromatosis [13].

Histopathology and immunohistochemistry are essential for DFSP diagnosis. Histopathologically, DFSP is characterized by the proliferation of plump, spindle cells arranged in a monotonous storiform pattern. The cells have little nuclear pleomorphism and low mitoses present. The main histologic differential diagnoses for DFSP are metaplastic carcinoma, fibromatosis, myoepithelioma, and Phyllodes tumor. A number of immunohistochemical markers are required to differentiate between these lesions. DFSP has positivity to CD34 in 84–100% of cases and to vimentin, which indicates the fibroblastic nature of the tumor. IHC markers for S-100, desmin, and actin are negative in DSFP as was the case in our patient [16].

Wide local surgical excision is the standard of treatment for localized DFSP tumors. Complete local surgical resection includes surgical margins of 2–3 cm and three-dimensional resection including skin, subcutaneous tissue, and underlying fascia [11,12]. The histological subtype, high cellularity, size, location on the head and neck, and high mitotic rate are associated with higher recurrence rates [17]. According to Dragoumis *et al*., the rate of local recurrence tends to decrease as surgical margins increases. Another study suggested that the surgical margin should not be <3.5–5 cm for a minimal recurrence rate [12]. As a first-line therapeutic option for cosmesis and to lower the recurrence rate, Mohs surgery is recommended by other authors [17-19]. However, this type of surgery needs the training of a specialized team and multiple stages. DuBay *et al*., in their clinical series published, had no DFSP recurrences identified after wide excision, Mohs surgery, or combination treatment over an average follow-up of 4.4 years [17].

In advanced cases where surgery is not an option, radiation therapy has been recommended as the sole treatment. A cure rate of more than 85% was found in cases where postoperative radiotherapy was used; however, this therapy carries a risk for the subsequent development of other skin tumors [14]. In our patient, we recommended adjuvant radiation therapy as it was a recurrent DFSP.

DFSP tends to be locally aggressive. Systemic dissemination is rare (1–4%) and usually occurs after multiple local recurrences. Metastases are hematogenous, with the lungs being the most common site. Extensive initial staging workup is not done routinely and is essential only for patients with suspected systemic disease [20].

Along-term follow-up is warranted as there have been recurrences reported after 5 years from the initial diagnosis. Due to its rarity, ability to mimic a variety of benign and malignant breast lesions, and increased readiness for accurate differentiation and management, DFSP in the breast is challenging to diagnose. To minimize recurrences, a personalized strict follow-up plan should be applied due to the lack of an existing evidence-based guideline.

CONCLUSION

DFSP is a rare breast neoplasm and possesses the potential for aggressive local behavior. It is a radioresponsive disease with

excellent local control after conservative surgery in combination with radiation therapy. Radiation therapy should be considered for patients with inoperable disease or recurrent tumors. Hence, a multimodality approach with conservative surgery and radiotherapy is needed for the treatment of DFSP breast.

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