

Recurrent Dermatofibrosarcoma Protuberans of the Chest Wall: The Sequential Ultrasonographic Features

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Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor with a propensity for local recurrence. Most DFSP recurrences occur within 3 years of the primary excision. The sequential ultrasonographic features from the time of the first diagnosis to the recurrence have not been previously described. We report here on a rare case of late local recurrence of DFSP and we describe the sequential imaging features, which would be helpful for the long-term evaluation of the excision scar.

Index words: Dermatofibrosarcoma protuberans; Neoplasm Recurrence, Local; Thoracic Wall; Ultrasonography

Introduction

Dermatofibrosarcoma protuberans (DFSP) accounts for approximately 6% of all soft tissue sarcomas and it typically presents as a small superficial soft tissue mass (1). Due to its infiltrating growth pattern, the tumor tends to extend far beyond the clinical margin. This explains the high recurrence rate of 20-15% after excision (2). Ultrasonography is considered an additional valuable method of monitoring this disease (3, 4). Although several reports have described the clinical and the ultrasonographic features of DFSP's local recurrence, to the best of our knowledge, no studies have reported the sequential ultrasonographic features of this disease from the time of the initial diagnosis to recurrence. We report here on a rare case of late local recurrence of DFSP and we describe the sequential ultra-

sonographic features.

Case

A 28-year-old female had a non-tender, reddish palpable mass in the previous excision scar of the left intra-clavicular chest wall. She had been aware of this lesion for 4 months and she complained that it was slowly growing. The scar had recently become nodular in consistency. Her family history was unremarkable, as well as her medical history, except for the fact that at the age of 23, she had a skin lesion excised from the same site of the left chest wall, which was reported to have been dermatofibrosarcoma protuberans (DFSP).

On physical examination, the patient had a well-circumscribed, reddish mass with a rubbery consistency on the scar tissue of the previous excision site. A clinical diagnosis of locally recurrent DFSP was strongly suspected.

Ultrasonography was performed with a linear transducer using a frequency of 5-12 MHz (HDI 5000, Advanced Technology Laboratories; Bothell, WA,

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USA) and with a 7.5–13 MHz linear transducer (EUP-L53; Hitachi Medical). Elastography was performed with a EUB-850 scanner (Hitachi Medical Corporation, Tokyo, Japan). Ultrasonography showed an oval, mixed echoic mass with a superficial hypoechoic lesion and a deep hyperechoic band extending from the dermis into the subcutaneous fat layer (Fig. 1). There was no vascularity seen on the color Doppler scan (Fig. 2). Elastography demonstrated the Ito score pattern 3 (the peripheral part of the lesion was green and the central part was blue) (Fig. 3). The initial ultrasonography showed an oval, mixed echoic mass in the dermis and subcutaneous fat layer. There were scattered areas of vascularity on the initial Color Doppler scan (Fig. 4). The serial ultrasonographic follow up images after the excision showed an irregular plate-like iso- to hyperechoic scar (Fig. 4). As compared with these follow up images, an irregular hypoechoic lesion had newly developed (Fig. 1), which was similar with the initial ultrasonography (Fig. 4). This lesion was considered to be a

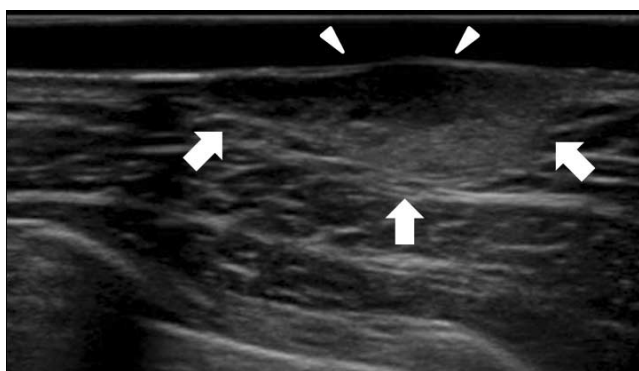


Fig. 1. Ultrasonography shows an oval, mixed echoic mass with a superficial hypoechoic lesion and a deep hyperechoic band extending from the dermis into the subcutaneous fat layer (arrows). Focal skin bulging is shown (arrowheads).

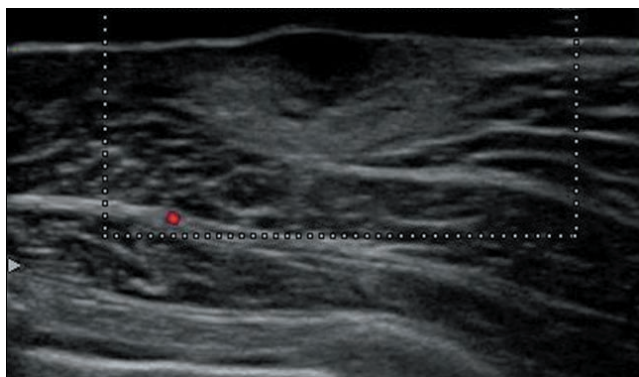


Fig. 2. Color Doppler scan shows no discernible flow signal.

recurrent tumor of BI-RADS category 4. Ultrasonographically guided percutaneous biopsy and a subsequent surgical wide excision demonstrated dermatofibrosarcoma protuberans (Figs. 5, 6). As compared with the initial excision pathology, the recurrence is more extensive with infiltrating the dermis and subcutis (Fig. 5), yet the microscopic features were similar to that of the initial pathology.

Discussion

The most frequent presenting location of DFSP is the skin of the trunk (50–60%) (3). DFSP is a relatively uncommon neoplasm of the deep dermis and subcutaneous tissue with low-grade malignant potential (2).

DFSP is a locally aggressive tumor with a high recurrence rate (2). Because of its infiltrating growth pattern, the tumor tends to extend far beyond the clinical margin. This explains the high recurrence rate of 15–20% after excision. Most recurrences of DFSP are detected within 3 years of primary excision (2). The histologic subtype, a high mitotic index, the cellularity and size, a location on the head and neck, and recurrent lesions are factors that are reportedly associated with higher recurrence rates. When the surgical margins are inadequate or conservative, the recurrence rates increase (2, 5). It always localizes to the initial site (6). In spite of the high rate of local recurrence, the US National Comprehensive Cancer Network recommends an overall clinical examination and inspection of the primary DFSP site

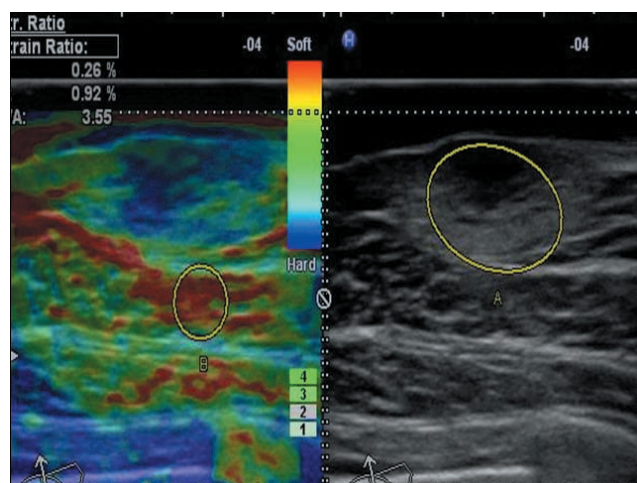


Fig. 3. Elastography shows the mosaic pattern of green and blue colors (score 2) and the strain ratio is 3.55.

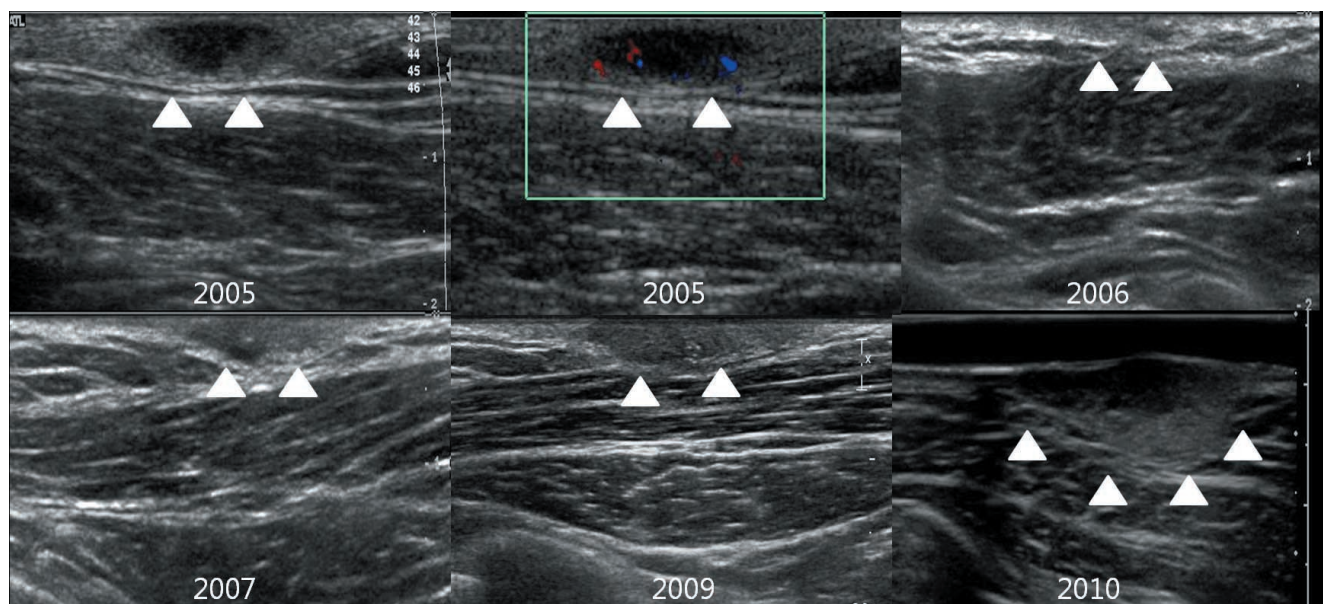


Fig. 4. The Serial ultrasonographic images demonstrate the lesion and the scar from 2005 to 2010. The initial ultrasonography and colour Doppler scan (2005) demonstrate an oval, mixed echoic mass in the dermis and subcutaneous fat layer and scattered areas of vascularity. The follow up ultrasonographic images (2006, 2007 and 2009) demonstrate an irregular iso- to hyperechoic scar on the excision site.

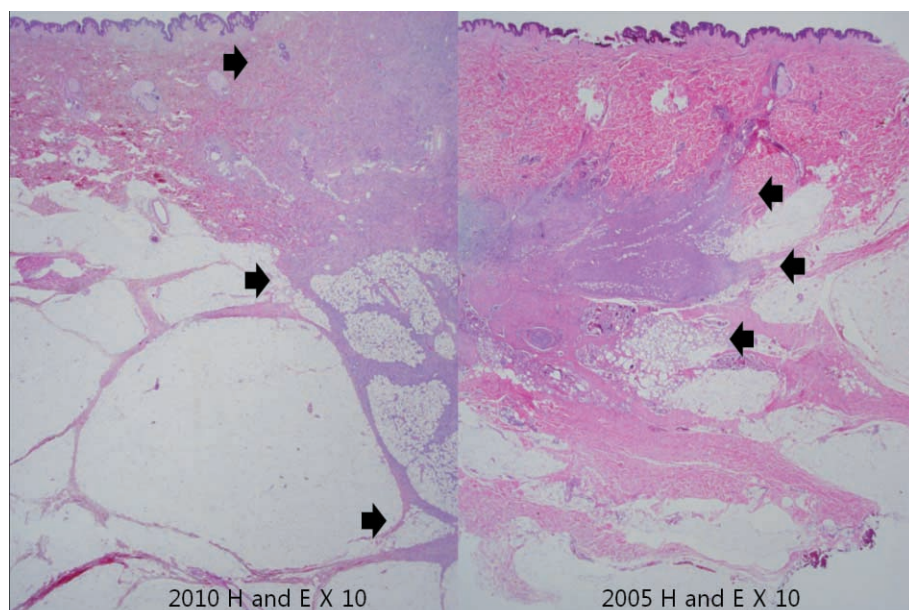


Fig. 5. Scanning view of the excision specimen (2005 and 2010) shows tumor cells in the deep dermis and they extend into the subcutis (arrows). Tumor involvement is more extensive in 2010 than that in 2005 (H and E, original magnification, $\times 10$).

every 6–12 months, with rebiopsy of any suspicious areas (3).

Ultrasonographically, the previously reported tumors were either broad-based against the dermis or they laid in the subcutis without a visible connection to the skin (7). The tumors were generally oval shaped and parallel in orientation. All the reported tumors were predominantly circumscribed, but some microlobulation might be seen. Some of the reported tumors were heteroge-

neously hypoechoic and others were mixed echoic with a hypoechoic area and wide hyperechoic bands (7, 8). Shin et al reported that the hypoechoic tumor demonstrates high cellularity, with spindle cells arranged in a distinct storiform pattern, whereas the hyperechoic bands exhibited a mixture of tumor cells and fibrous tissue infiltrating the subcutaneous fat (8). Our case, including the initial tumor and the recurrent tumor showed mixed areas of hypoechogenicity and hypere-

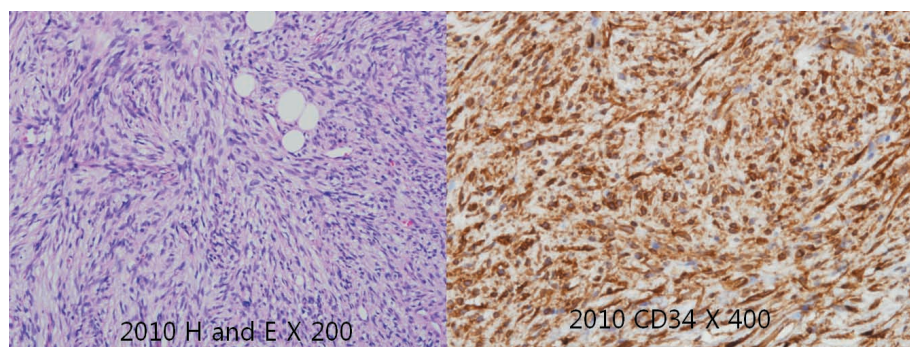


Fig. 6. High power view reveals short fascicles of densely cellular and uniform spindle cells in a storiform pattern (H and E, original magnification, $\times 200$). Immunohistochemical staining for CD 34 shows diffuse and strong positivity (CD 34, $\times 400$).

choic bands. DFSP shows scattered areas of hypervascularity throughout the tumor (7, 8) or flow signals along the periphery of the mass on a color Doppler scan (8). The initial tumor in our case showed increased vascularity, but there was no flow signal in the recurrent tumor. Elastography is a method of visualizing the elastic characteristics of a lesion. Several clinical studies have reported that elastography has the potential to differentiate benign from malignant solid breast masses (9, 10). The elastographic images are generally given one of five elasticity scores according to Itoh et al: score 1: the entire lesion is evenly shaded in green, score 2: the hypoechoic lesion has a mosaic pattern of green and blue, score 3: the peripheral part of the lesion is green and the central part was blue, score 4: the entire lesion was blue, but its surrounding area was not included, and score 5: both the entire hypoechoic lesion and its surrounding area are blue. Lesions with scores of 1–3 are classified as benign, while those with scores 4 and 5 are classified as malignant (10). Our case showed an elasticity score of 3 and this was classified as a benign pattern. There are few reports that have described the ultrasonographic feature of a recurrent DFSP (2) and there are no reports that have shown the sequential ultrasonographic features from the time of the initial tumor to recurrence. A recurrent DFSP was previously reported to be a circumscribed oval hypoechoic mass (2), which was similar to our case. This hypoechoic lesion in our case had newly developed at the scar site of the previously excised tumor.

Surgical excision remains the cornerstone of treatment for DFSP. Complete surgical resection is accepted

as the optimal treatment for primary or recurrent DFSP. Most authorities would suggest a margin of 2–3 cm of normal tissue from the gross tumor boundary, with a three dimensional resection that includes skin, subcutaneous tissue and the underlying fascia (2, 5).

Ultrasonographic follow-up would be helpful for the long-term evaluation of the scar after excision of DFSP

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흉벽에 재발한 Dermatofibrosarcoma Protuberans의 순차적 초음파 소견 : 증례 보고

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Dermatofibrosarcoma protuberans (DFSP)는 국소재발 성향을 보이는 드문 연조직 종양이다. 대부분의 DFSP는 제거 수술 후 3년 이내에 재발한다. 최초 진단에서부터 재발까지 순차적 초음파 소견에 대한 보고는 없었다. 저자들은 늦게 발생한 DFSP의 재발 증례를 보고하고 순차적 초음파 소견에 대해서 기술하고자 한다. 이러한 순차적 초음파 소견은 제거 수술 반환의 장기 추적검사에 유용하다.

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