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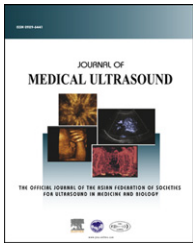

ORIGINAL ARTICLE

Ultrasound Appearances of Dermatofibrosarcoma Protuberans

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To analyze the ultrasound appearances of dermatofibrosarcoma protuberans (DFSP) and correlate these with pathologic findings. Nine tumors in eight patients were analyzed (one patient having two separate DFSP tumors). All ultrasound images were assessed and correlated with the histologic findings. Most (7/9, or 78%) tumors were located in the subcutaneous region. The shape of the tumor was round in 67% (6/9) and ovoid in 33% (3/9) of cases. The margin was poorly defined in one (11%) and well-defined or circumscribed in eight (89%) tumors. All the tumors showed a mildly lobulated border and had a heterogeneously hypoechoic matrix, often with rounded, ovoid, or occasionally linear discrete hypoechoic areas. Very small echogenic foci (<0.5 mm), usually without an accompanying comet tail artifact, were seen within the tumor matrix of all the cases. Posterior enhancement was also a feature of all the tumors. Most (67%) tumors showed moderate vascularity on color Doppler imaging. This vascularity tended to be more profound peripherally rather than centrally, and tended to be more organized rather than chaotic in distribution. Based on its quite characteristic ultrasound appearances, one should be able to either diagnose, or at least suggest, the likelihood of a DFSP tumor. In such circumstances, either percutaneous biopsy or *en-bloc* resection with wide margins is recommended.

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a low-grade spindle cell sarcoma accounting for about 6% of all soft tissue sarcomas. It is, however, the most common primary sarcoma to occur in the subcutaneous tissues [1]. It was originally described as a distinct clinicopathologic entity in 1924 by Darier and Ferrand [2], with the terminology being coined by Hoffman 1 year later [3]. DFSP is a mesenchymal tumor nearly always arising from the dermis and usually associated with typical clinical appearances [1].

Clinically, DFSP starts as a small nodule that will grow into a medium-sized erythematous or bluish lesion protruding from the skin [1]. This nodule may eventually ulcerate [1]. Imaging is performed in those clinical situations where the typical clinical appearances are not present, or where the clinician is either unaware of the typical clinical appearances or is unclear as to the depth of invasion.

The magnetic resonance imaging (MRI) and computed tomography (CT) appearances of DFSP have been described as nonspecific [4,5]. The sonographic appearances of non-breast DFSP have been described in one case report and one case series [6,7]. The ultrasound features of breast DFSP have also been specifically reported [6,7]. Overall, the appearances of breast DFSP are similar to those of DFSP tumors occurring outside the breast [6,7].

This study was undertaken to document in greater detail the ultrasound appearances of DFSP lesions in eight patients, particularly with a view to identifying any characteristic ultrasound appearances that would enable preoperative recognition of this tumor.

Materials and methods

The pathology database at our institution was retrospectively accessed to identify tumors with a pathologic diagnosis of DFSP presenting between May 1999 and October 2010. A total of 52 patients with histologically confirmed DFSP were retrieved, of whom 13 (23%) had undergone ultrasound prior to excision. The ultrasound images of the DFSP were available for review in eight of these 13 patients. The mean patient age of these eight patients was 49 $\pm$ 10.3 (standard deviation) years, range 30–60 years, with a slight female predominance (female:male = 5:3). All had undergone both grayscale and color Doppler ultrasound with one of four musculoskeletal radiologists (with 5–15 years' musculoskeletal ultrasound experience), using either high-resolution 12–17 MHz linear transducers for superficial lesions or moderate resolution (5 MHz) linear transducers for deeper lesions (Sonoline Elegra, Siemens, Issaquah, WA, USA; iU22, Philips, Bothell, WA, USA).

Two fellowship-trained musculoskeletal radiologists (with a musculoskeletal sonography experience of 5–15 years) retrospectively reviewed, in consensus, the ultrasound images of these DFSP tumors, noting the following imaging features: the location (dermal, subcutaneous, intramuscular), shape (rounded, ovoid), margin (well defined, ill defined), border (smooth, spiculated, lobulated), principal echogenicity (anechoic, hypoechoic, isoechoic, or hyperechoic to subcutaneous fat), additional echogenic areas (anechoic, hypoechoic, isoechoic, or hyperechoic to subcutaneous fat), central tiny echogenic foci (absence or presence), posterior enhancement or shadowing, tumor rim (absence or presence), internal vascularity during color Doppler imaging (peripheral or central), and whether this vascularity was organized (i.e., with vessels dispersed at regular intervals) or chaotic (i.e. the vessels irregularly dispersed) [8]. The electronic medical records were accessed to record the preimaging clinical diagnosis, biopsy findings, surgical findings, and clinical recurrence. The histology slides of both the biopsy and excision specimens of the tumors under review were retrieved and reassessed by a senior pathologist experienced in assessing soft tissue tumors.

Results

Imaging data

A total of nine tumors were analyzed (one patient had two separate large tumors of the knee region). The pre-ultrasound clinical diagnoses were DFSP (2/9, 22%), soft tissue sarcoma (5/9, 56%), hemangioma (1/9, 11%), and breast carcinoma (1/9, 11%). Ultrasound was performed on all nine tumors. Additional MRI, CT, and positron-emission tomography CT (PET-CT) studies were performed in Case 1, Case 3, and Case 8, respectively, for presurgical assessment.

Ultrasound appearances

For the nine tumors studied, the maximum dimension ranged from 1.8 to 11.0 cm, with a mean of 6.7 cm. Most [78% (7/9)] tumors were located in the subcutaneous tissues, with one located intermuscularly (between the tibialis anterior and extensor digitorum longus muscles) and one located just deep to the investing fascia of the upper leg. Most tumors were mainly round [67% (6/9)] in shape rather than ovoid [33% (3/9)]. One (11%) tumor showed a poorly defined margin, while the remainder [89% (8/9)] were well-marginated or circumscribed. All tumors showed a mildly lobulated border, while one tumor (11%) had a focal appendage-like elongation.

All the tumors had a heterogeneously hypoechoic matrix (compared to subcutaneous fat), often containing a rounded, ovoid, or occasionally linear more discrete hypoechoic area or areas. Very small echogenic foci ($<$0.5 mm), usually without an accompanying comet tail artifact, were seen within the tumor matrix of all the cases. Posterior enhancement was also a feature seen in all the tumors.

Most [67% (6/9)] tumors showed moderate vascularity on color Doppler imaging. This vascularity tended to be more profound peripherally than centrally, and tended to be more organized rather than chaotic in distribution [8]. Ultrasound-guided core biopsies using a 16 G Tru-Cut needle and a coaxial system were performed on all tumors. These biopsies revealed features consistent with DFSP in all cases.