Sonography of Dermatofibrosarcoma Protuberans in the Skin Over Breast

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We describe here the sonographic manifestation of the soft tissue neoplasm, dermatofibrosarcoma protuberans, which rarely occurs in the breast. Four patients had clinical features of breast mass. Sonography was performed with a high-frequency transducer (7.5–12.0MHz). Distribution of blood flow inside the tumor was studied by color Doppler. All the cases of dermatofibrosarcoma protuberans observed on the sonograms were hypoechoic, with a heterogeneous internal echo. An oval-shaped and circumscribed margin was seen in two of the tumors, whereas a lobulated shape and slightly obscured margin was noted in the other two tumors. On color Doppler, arterial blood flow signals were noted inside the tumor in three of the cases, and no significant flow signal was noticed in the remaining case. In three of the cases, the masses were in the dermal region, without infiltrating the mammary tissue, and they were diagnosed as a dermal mass. However, in the remaining case, the mass infiltrated the mammary tissue and was thus misdiagnosed as a breast mass.

KEY WORDS — color Doppler, dermatofibrosarcoma protuberans, ultrasonography

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor of cutaneous origin with an intermediate level of malignancy. DFSP was reported in as early as 1890, but it was first described by Darier and Ferrand [1] in 1924, as a distinct cutaneous disease entity called progressive and recurring dermatofibroma. It was first referred to as DFSP by Hoffmann in 1925 [2]. Several studies have revealed a homogenous sex distribution with a slight bias towards men [3]. It usually occurs between the second and fifth decade of life [4] and most frequently appears on the trunk and the extremities. However, its occurrence in the breast is rare [5]. Therefore, sonographic manifestation of dermatofibrosarcoma protuberans in the breast is rarely reported. We present four cases of DFSP in the breast that were recorded in our hospital from January 2002 to December 2008. The value of sonography in imaging DFSP and its sonographic features are briefly discussed.
Case Reports

Case 1
A 35-year-old male patient who was suspected as having fibroadenoma and lipoma, and had undergone surgery of his left breast three times within a period of 10 years, presented with a complaint of pain and aggressive growth in the past 2 years. A physical examination revealed a soft tissue tumor in the previously resected surgical area, accompanied by redness and swelling. The size of the tumor was 3.2 cm × 3.7 cm. Routine laboratory examinations were unremarkable. Ultrasonography showed a hypoechoic mass with an oval-shaped and circumscribed margin in the dermis with a heterogeneous internal echo (Fig. 1). Swelling of the axillary lymph node with clear corticomedullary differentiation was noted. Doppler sonography showed no remarkable blood flow inside the tumor. A benign tumor of dermal origin was suspected. Aspiration biopsy was not carried out. The patient was referred to a surgeon for their opinion. Once the intraoperative histopathologic examination confirmed DFSP, surgery was carried out with a wide excision margin.

Case 2
A 21-year-old woman had noticed a growth in her left breast for 3 years, which had already been operated upon, and was beginning to grow rapidly accompanied by redness and swelling in the 4 months leading up to her being admitted to our hospital. Her family and past history were unremarkable. Clinical examination showed a tumor in her left breast with remarkable redness and swelling. Ultrasonography revealed a hypoechoic mass with a heterogeneous internal echo with size of 5.3 cm × 4.9 cm. An oval-shaped and slightly obscured margin was seen. Color Doppler revealed a profuse blood flow signal inside the entire tumor (Fig. 2). Axillary and supraclavicular lymph nodes were normal. As the mass was seen in the glandular tissue, a benign mass originating from glandular tissue was reported. Since the intraoperative pathologic examination revealed DFSP, surgery was carried out with a wide excision margin.

Case 3
A 29-year-old woman had a history of a right breast mass since the previous year. She had rapid growth of the mass, excessive redness of the skin and discharge from the nipple in the preceding 4 months. Thus, she presented to our hospital. Her family and past history were insignificant. A physical examination showed a mass in her right breast with significant redness and swelling. Ultrasonography revealed a hypoechoic mass with a heterogeneous internal echo with size of 5.3 cm × 4.9 cm. An oval-shaped and slightly obscured margin was seen. Color Doppler revealed a profuse blood flow signal inside the entire tumor (Fig. 3). Axillary and supraclavicular lymph nodes were normal. As the mass was seen in the glandular tissue, a benign mass originating from glandular tissue was reported. Since the intraoperative pathologic examination revealed DFSP, a wide excision margin was carried out.
Case 4
A 30-year-old woman had a history of fibroadenoma on her left breast and had undergone surgery twice within 2 consecutive years. She was admitted to our hospital with a soft tissue mass on the previously resected area. During the preceding 2 months she had noticed that redness of the skin, pain, and swelling were aggravated. Sonography showed a large mass with a size of 5.2 cm × 3.0 cm × 5.4 cm on the dermis over the breast. The mass was hypoechoic, oval-shaped, with a circumscribed margin and had a heterogeneous internal echo. Color Doppler showed a relatively increased blood flow signal (Fig. 4). Swelling of the axillary lymph node that was 1.9 cm × 0.8 cm with a clear corticomedullary differentiation was also noted. Considering the sonographic manifestations, a benign solid mass originating from the dermis was suspected. With a provisional diagnosis of benign mass, she underwent surgery. Once the histological examination showed that the mass was DFSP, surgery was performed with a wide margin.

Discussion
DFSP is a rare low-grade soft tissue neoplasm that originates from the dermis and accounts for approximately 1.8% of all soft tissue sarcomas [6,7]. Previous reports have shown that men are affected more frequently than women [3]. The tumorigenesis of DFSP is still not clear. A history of trauma with the tumor originating in surgical scars [8], trauma scars [9] and burn scars [10] has been previously reported as the possible causes for inducing DFSP. However, recent cytological investigations have shown that tumor cells with chromosomal anomalies of monocellular origin to be the main cause. Translocation of chromosomes 17 and 22 resulting in unregulated expression of platelet-derived growth factor has been shown to induce the pathogenesis of DFSP.

DFSP may occur in any area but is most frequently seen on the trunk and extremities and is rarely seen in the mammary region. Therefore, diagnosis is often difficult, as it is easy to misdiagnose as a breast tumor. Although DFSP is a slow growing tumor, it has a high tendency of local infiltration, and therefore, the surrounding fat, fascia, muscle, mammary gland and even bone may become infiltrated. Remote metastasis rarely occurs, and when it does occur, the common sites are the lungs and the regional lymph nodes [11,12].

Sonography, a noninvasive and nonionizing tool, has been used for imaging and assists in the diagnosis of superficial soft tissue lesions of subcutaneous tissue from the very early period. Diagnosis of superficial lesions may be suspected on the basis of
the tumor's clinical appearance. However, large lesions can infiltrate the glandular tissue and can be easily confused with higher grade sarcomas. The primary origin of the lesion is often difficult to detect. Sonography is able to determine the location, infiltration, nature and vascularity of the lesion, particularly within a very short time. In our hospital, the number of diagnosed cases of DFSP from January 2002 to December 2008 was 15, and 4 of the cases were in the breast region. Since it rarely occurs in the breast, care should be taken to avoid misdiagnosing during the process of imaging. A case of DFSP in the breast of a Japanese woman was reported by Fukushima et al [5], which was sono-graphically diagnosed as a benign tumor. Similarly Kau et al [13] reported a case of DFSP in the groin. Shin et al [14] reported four cases of DFSP and correlated the sonographic features with pathology examination results. A case of pancreatic metastasis of DFSP was also reported by Yokoyama et al [15]. They found that there was metastasis to the pancreas and the left gluteus maximus muscle after the operation and radiotherapy were carried out for the primary lesion in the left lower abdomen. Similarly, Chattopadhyay et al [16] reported 10 cases of DFSP in the All India Institute of Medical Sciences, New Delhi, India. In their study, only two cases occurred in the chest region. Thus, there are few reports of DFSP in the breast; the features of DFSP are difficult to diagnose correctly. Most of the DFSPs are usually characterized by painless, atrophic plaques and occasionally present as red plaques with irregular borders, mimicking hemangioma [6]. It is difficult to make a correct diagnosis by clinicians merely on the basis of examination of the tumor because it may be confused with localized scleroderma or congenital solitary fibromatosis [6,17].

In our cases, sonography showed an oval-shaped, hypoechoic mass with an internal irregular echo just below the skin in the dermal region. Color Doppler showed that three of the four cases had arterial blood flow signals inside the tumor. The remaining case had axillary lymph node enlargement with clear corticomedullary differentiation. Moreover, three of the four cases were reported to have originated from the dermis of the breast. One case was misdiagnosed because the mass was wrongly suspected of having originated from the glandular tissue of the breast. Although it was DFSP, a large mass was seen in the mammary tissue with sonography. We did not observe local invasion, and therefore, it was misdiagnosed. However, the other three masses were reported to have originated from the dermis. Therefore, we believe that sonography is helpful in the differentiation of a dermal lesion DFSP from a primary breast lesion. According to Shin et al [14], if a sonogram reveals an oval mass in the subcutaneous tissue that is abutting against the skin and has a focal lobulated margin with hypoechogenicity or an irregular margin with mixed echogenicity, a diagnosis of DFSP should be considered. The hetero-echogenicity of the mass is due to the internal components of the tumor cells and the fibrous tissue. When examined histologically, the tumor parenchyma is characterized by a distinct storiform pattern, created by spindle-shaped cells arranged in an irregularly whorled pattern. According to Laskin [11], the regional lymph nodes are the primary sites of metastasis. Metastasis in DFSP is also rare, and therefore, no abnormal regional lymphadenopathy was noted in the four cases. Color Doppler is another technique used in detecting blood flow in breast tumors. Vessels in a malignant tumor tend to have a higher velocity as well as pulsatility flow than those in benign tumors. Neovascularization in a malignant tumor gives rise to the characteristic flow that is different from benign tumors [18]. Doppler revealed that among our four cases, two different patterns of blood flow signal were noted within the tumor. The first was distinguished by the absence of a small amount of blood flow through the peripheral portions of the tumor. The second pattern was a profuse blood flow signal noted through the tumor (Figs. 2, 3 and 4). Interestingly, two of our four cases of DFSP had originated in surgical scars. Cases of DFSP occurring in surgical scar sites have also been previously reported [6].

It is not easy to make a correct diagnosis of DFSP solely depending on sonography. Clinicians need
to be aware that during a sonogram, a number of neoplasms having mixed echogenicity appear to originate from the dermis, e.g. hamartoma, chronic lipid cyst, lipoma with fibrous contents, and fat necrosis. These lesions can be differentiated from DFSP by mammography, as these lesions contain fat but DFSP does not. Similarly, an epidermal inclusion cyst of a superficial location should be taken into consideration as a differential diagnosis. However, they usually have little or no blood flow signal on color Doppler [19]. Sonography can still greatly assist in substantial prediagnosis of DFSP. Sonography-guided percutaneous biopsy and core needle biopsy are also challenging methods in the diagnosis of soft tissue sarcoma. Many studies have been carried out to examine the effectiveness and accuracy for diagnosis of DFSP using these two methods. However, these methods do not have the same level of high accuracy and effectiveness as intraoperatively carried out excisional biopsy [20,21]. Therefore, intraoperative histopathologic examination is carried out to rule out DFSP, as it is the only gold standard method to date. Wide local excision or Mohs micrographic surgery is the preferred treatment method. Failure to completely excise the peripheral projectile structures of the tumor is the major factor leading to recurrence. Due to technical or cosmetic reasons, for those patients who cannot undergo wide surgical excision, the combination of conservative resection and postoperative radiotherapy might be considered as an alternative [22,23]. In a series of 19 DFSP cases treated with radiation as adjuvant therapy after surgical resection at MD Anderson Cancer Center (Houston, Texas), only a single recurrence was reported, occurring in a patient who had received radiotherapy as a definitive treatment for gross postoperative disease.

In summary, DFSP is a rare soft tissue tumor arising from the superficial layer of the skin. Whenever a sonogram reveals a well-circumscribed hypoechoic mass in the superficial layer of the breast, DFSP should be considered as a differential diagnosis. Doppler sonography may be helpful, since most of DFSPs are hypervascular.

References

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