An unusual case of cauliflower-like tumor of breast

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To the Editor: Dermatofibrosarcoma protuberans is a type of poor to moderately differentiated tumor that accounts for 0.1% of all malignant tumors.[1] This tumor type mainly occurs in the superficial skin and the subcutaneous tissue of the trunk and is rare in patients with breast cancer.[2] Here, we report a patient with dermatofibrosarcoma protuberans of the breast whose tumors are irregular in shape.

A 24-year-old woman was admitted to the Central Theater Command General Hospital of the Chinese People's Liberation Army with a tumor in the left breast that was discovered 2 years earlier. Physical examination: A 6.0 cm × 5.0 cm tumor was observed in the upper quadrant of the left breast that was prominent in the skin and exhibited an inflamed, red appearance. In the lower quadrant of the left breast, a 4.0 cm × 5.0 cm mass was observed that was prominent in the skin; it was dark red in color, hard in texture, exhibited no obvious tenderness, showed moderate activity, and had unclear boundaries with surrounding tissues. The skin of the left breast exhibited a patchy appearance. The skin color of the right breast was normal, and no nipple inversion was found [Figure 1A]. Bilateral breast color Doppler ultrasonography revealed the following. Left breast: multiple cauliflower-like exophytic masses were observed, including a solid mass 6.4 cm × 4.2 cm in size in the upper quadrant of the nipple involving the subcutaneous tissue and skin layer, deep to the muscular layer, and it had a smooth border and irregular shape. Right breast: an uneven parenchymal echo, a "leopard" pattern, and visible patches were observed. Hypoechoic areas of shape and strip were not detected by mass or mass echoes. Bilateral axilla: there were several solid hypoechoic nodules with blurred boundaries and middle lymphatic hilum structure, of which the left side was 1.4 cm × 0.5 cm and the right side was 1.7 cm × 0.5 cm. Color Doppler flow imaging showed that there was abundant blood flow in the solid mass in the quadrant above the left breast papilla. No abnormal blood flow signal was found in the right breast. Stellate blood flow signals were observed in bilateral axillary hypoechoic nodules [Figure 1B]. Ultrasound-guided biopsy under local anesthesia showed the following: (left) considering the duration of dermatofibrosarcoma protuberans, combined with the general clinical findings, immunohistochemical results were vimentin (+), desmin (−), smooth muscular actin (−), CD34 (+), CD31 (−), S-100 (−), Nestin (+), Ki67 (+5%), and beta-catenin (serum +) [Figure 1C]. Chest (bilateral) computerized tomography (CT) plain scan: multiple irregular soft tissue masses approximately 51 mm × 66 mm in size with clear margins were found in the left breast and left chest wall; no consolidation was found in either lung, the hilar bronchial opening was unobstructed, and no enlarged lymph node shadow was observed in the mediastinum [Figure 1D and 1E]. There were no abnormalities in the relevant examinations, including three routine examinations, namely, liver and kidney function and coagulation function evaluations. Because the patient was diagnosed in our hospital, it was recommended that the patient seek further surgical treatment in another hospital as soon as possible.

Dermatofibrosarcoma protuberans is a poor to moderately differentiated malignant tumor occurring in the dermis and subcutaneous mesenchymal tissue. This tumor grows slowly and locally and has a high local recurrence rate. However, distant metastasis is rare.[3] Protuberant dermatofibrosarcoma accounts for approximately 2% to 6% of soft tissue sarcoma and 0.1% of all malignant tumors. The annual incidence of protuberant dermatofibrosarcoma is approximately (0.8–4.5)/1 million.[1] The peak age of the disease is 20 to 50 years old, and it affects more males than females. The initial stage of the disease can present as a small nodule, which grows slowly. The course of the disease can be over several years to decades. With the progression of the disease, the tumor size can increase rapidly. The disease mainly occurs in the superficial skin and subcutaneous tissue of the trunk (50%–60%), followed by the limbs (20%–30%) and then the head and neck (10%–15%). This condition is rare in breast cancer patients. The etiology of dermatofibrosarcoma protuberans (DFSP) is still unknown. Rutger *et al*[2]

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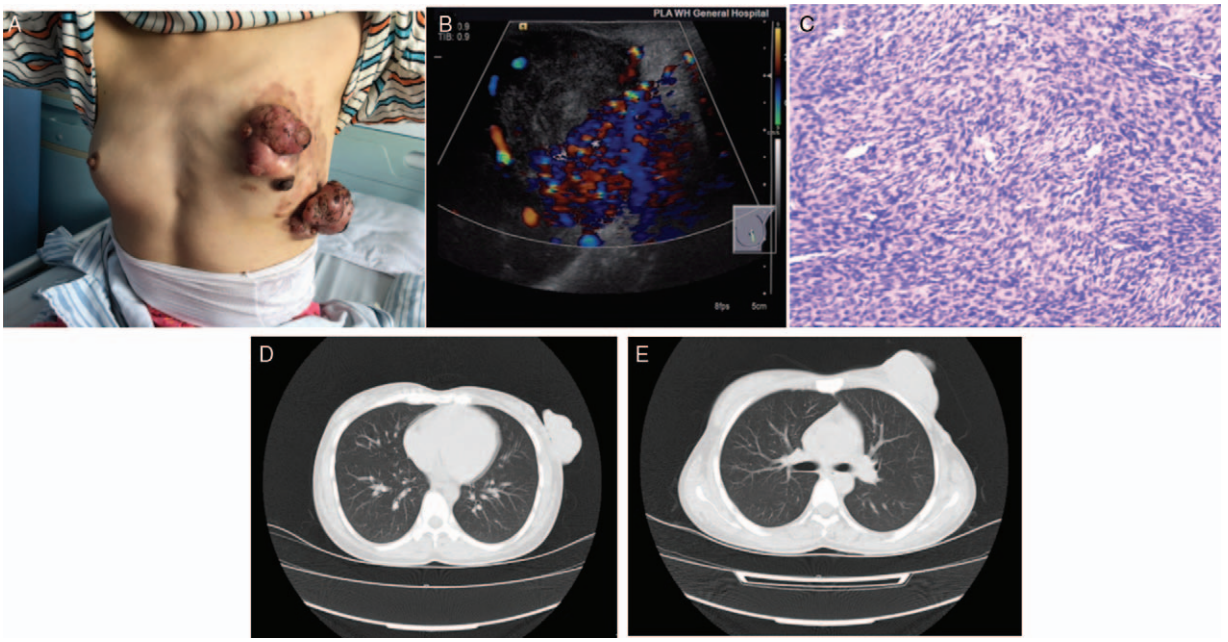

Figure 1: A large dermatofibrosarcoma protuberans of the breast in a 24-year-old female. (A) Tumor morphology at admission on July 24, 2017. (B) Patients underwent ultrasound-guided puncture biopsy under local anesthesia on July 25, 2017. (C) Pathological diagnosis of breast protuberant dermatofibrosarcoma (H&E staining, original magnification ×100). Tumor cells were short bundles, closely arranged in spokes, with long spindle nuclei and clear cytoplasm. (D) Chest CT plain scan shows the oval slightly low-density shadows on the lower quadrant of the left breast with uniform density and clear boundary. (E) Chest CT plain scan shows the oval slightly low-density shadows on the upper quadrant of the left breast with uniform density and clear boundary. CT: Computerized tomography; H&E: Haematoxylin and eosin.

reported 264 cases of patients with dermatofibrosarcoma protuberans, suggesting that the occurrence of this disease may be related to the excessive proliferation of damaged tissues, while Xia Tian *et al*[4] reported a case of dermatofibrosarcoma protuberans of the breast in a patient with a history of breast trauma. However, the specific biological differences of dermatofibrosarcoma protuberans occurring in the breast and other parts of the body have not yet been reported and need to be further studied. This tumor rarely metastasizes and is classified as an intermediate type of fibroblast/myofibroblastic tumor in the 2013 World Health Organization classification.

If a subcutaneous tissue-like round mass adjacent to the skin appears as a lobulated hypoechoic region or irregular mixed echoes on ultrasound, then the possibility of dermatofibrosarcoma protuberans of the breast should be considered. Ultrasonography showed that the surface of the breast involved the subcutaneous tissue and skin layer and the deep to muscular layer, it exhibited a smooth border and irregular shape, and the blood flow signals in the solid mass above the left nipple were abundant. Kransdorf *et al*[5] reported the CT findings of six cases of dermatofibrosarcoma protuberans, which showed clear isodense or slightly hypodense shadows with uniform density and clear boundaries. Chest CT showed irregular isodensity with a clear boundary. Imaging examinations can provide the location, size, shape, margin, internal structure, degree of skin and deep invasion, and the relationship with the surrounding tissues. This analysis can provide vital information for the pre-operative evaluation of protuberant dermatofibrosarcoma, especially for patients with recurrence after the operation.

Pre-operative imaging examination of this disease mainly includes mammography, ultrasonography, CT, and magnetic resonance imaging, and the final diagnosis depends on pathological examination. The epidermis of dermatofibrosarcoma protuberans is often atrophic. The dermis is composed of spindle-like fibroblasts of the same size. Tumors are rich in cells. Around the center, the cells are arranged in a radial shape or in sheets. Tumor cells can grow like crab claws, invading subcutaneous tissue, interlobular septa or muscular layers to form honeycomb-like or multi-layered parallel arrangements. CD34 positivity is found in all types of dermatofibrosarcoma protuberans. CD34 was positive in this patient, and the histopathological findings were consistent with those of dermatofibrosarcoma protuberans. In addition, more than 90% of dermatofibrosarcoma protuberans gene analyses exhibit translocation of chromosomes 17 and 22 (17; 22), which could be used as a new diagnostic method for dermatofibrosarcoma protuberans.

Tumor cells of dermatofibrosarcoma protuberans are insensitive to chemotherapy, and thus, radical surgical resection is still the first-line treatment. Surgical margins should be 3 to 5 cm away from the tumor tissue. The Mohs operation can also be performed, which effectively reduces the recurrence rate. The patient in this study underwent enlarged resection of the tumors after diagnosis in our hospital. For patients with large tumors, negative margins, metastasis or recurrence, adjuvant radiotherapy or imatinib mesylate, a tyrosine kinase inhibitor, can be used. Tyrosine kinase inhibitors selectively inhibit the bcr gene (bcr/abl), stem cell factor receptor (Kit), the human CSF-1 receptor gene (c-FMS), Arg (abl-related gene), platelet-derived

growth factor receptor A and B (PDGFRA and PDGFRB), prevent tyrosine kinase receptor phosphorylation, suppress abnormal signaling pathways, and inhibit the proliferation of dermatofibrosarcoma protuberans.

Differential diagnosis should be performed to distinguish this disease from breast fibroadenoma and phyllodes tumors. The former mainly appears as a round-like mass with smooth and sharp edges, some of which show calcification, good mobility, and no adhesion to the skin; the latter is lobulated, smooth edge and highly dense mass, but the surface of the skin and subcutaneous fat are still relatively complete.

In summary, breast protuberant dermatofibrosarcoma is rare, and the size of the tumors in this patient was approximately 6.0 cm × 5.0 cm. Final diagnosis can be confirmed by imaging examination and ultrasound-guided puncture biopsy. At present, surgical resection is the preferred treatment for this disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient provided her consent for the images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

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