Primary Granulocytic Sarcoma Presenting as Bilateral Breast Masses

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Primary granulocytic sarcoma (GS) of the breast is rare. The sonographic finding of GS is nonspecific, mainly noncalcified hypoechoic lesion with ill-defined margin and hypervascularity. Correct diagnosis is important to avoid unnecessary surgery. However, distinguishing GS from metastatic malignancies or other hematologic malignancies is difficult if there is a lack of clinical information of known hematologic disorders in the patient. We describe a 51-year-old woman who initially presented with bilateral breast masses in whom histologic study revealed a pattern of GS. She was finally diagnosed to have acute myelogenous leukemia.

KEY WORDS — acute myelogenous leukemia, breast tumor, granulocytic sarcoma, ultrasound

Introduction

Granulocytic sarcoma (GS) is a rare tumor composed of immature myeloid cells in extramedullary sites [1]. It generally occurs either during the natural course of acute myelogenous leukemia (AML) or is associated with other myeloproliferative disorders. However, GS may rarely represent the initial manifestation of the disease [2].

Case Report

A 51-year-old woman presented with palpable nodules over bilateral breasts of 2 weeks’ duration. Excisional biopsy of a left breast lesion was done at a local hospital and the frozen section showed an invasive tumor. This woman had a past history of hysterectomy without hormone replacement therapy.

Mammography was performed and two lobulated nodular lesions in the breasts were demonstrated. One was found in the right subareolar region and the other in the left subareolar region, respectively (Fig. 1). No definite microcalcification could be identified. Ultrasound showed irregular shaped heterogeneous hypoechoic nodular lesions with ill-defined margins and mild posterior enhancement over bilateral breasts (Fig. 2).

Excisional biopsy was performed and two greenish-yellow soft tissue nodules were obtained, one from each breast. Microscopic study showed tumor cells with granular cytoplasm diffusely infiltrating in the breast stroma. Immunohistochemical stains for myeloperoxidase were diffuse and strongly

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Fig. 1. Mammography shows two lobulated nodular lesions in the breasts: one in the right subareolar region (arrows) and the other in the left subareolar region (arrowheads).

Fig. 2. Sonography shows two irregular heterogeneous hypoechoic nodular lesions with ill-defined margins and minimal distal acoustic enhancement: (A) right breast (arrows); (B) left breast (arrowheads). Color Doppler study of the lesions shows minimal peripheral color flow signals: (C) right breast (arrow); (D) left breast (arrowheads).
positive; CD117 was focally positive, while stains for CD34 and CD68 were negative. The histopathologic study was compatible with GS. Bone marrow biopsy was normal at the beginning. Two months later, anterior chest pain was noticed and bony metastatic lesion was depicted over the sternum on X-ray studies. Sternal bone marrow biopsy showed hypercellularity with blast cells over 90%; AML was subsequently diagnosed.

Discussion

GS, previously called extramedullary myeloblastoma or chlora, is an unusual variant of myeloid malignancy. This entity mainly consists of myeloblastic leukemia, presenting as a solid tumor mass composed of aggregates of immature granulocytic precursors in extramedullary sites such as the central nervous system, bones, soft tissues of the head and neck, skin and breast [3]. It occurs during either leukemia relapse or remission, but rarely presents as a primary extramedullary lesion. GS developing in the breast is a rare entity [4]. Isolated GS in the breast without bone marrow evidence of leukemia is exceedingly rare [3].

GS generally occurs either during the natural course of AML or after remission [2]. In the absence of any clinical history or hematologic abnormality, GS may be misdiagnosed, depending on the degree of myeloid differentiation present within the tumor. The differential diagnoses include large-cell non-Hodgkin’s lymphoma, lobular carcinoma of the breast, undifferentiated carcinoma, malignant melanoma, extramedullary hematopoiesis and inflammation [5].

In addition to AML, other hematologic abnormalities can also be associated with GS. GS has been reported to develop in patients with myelodysplastic syndromes and advanced-stage chronic myeloid leukemia (CML) [4]. The overall prevalence of GS in leukemia patients is about 7% (8% in AML, 4% in CML) [5–8].

Primary GS presents most commonly in skin and lymph nodes. The breast is an uncommon site for development of primary GS [3]. Therefore, when it does present in the breast, misdiagnosis is common. Primary breast GS is most frequently misdiagnosed as lymphoma or sarcoma. Histologic testing and immunostaining are essential for proper diagnosis [4].

The presence of immature eosinophils was an important clue. Leder’s chloroacetate esterase stain, immunostaining for myeloperoxidase, CD34, CD43, CD68, and lysozyme, and ultrastructural finding of cytoplasmic lysosomal granules and Auer bodies all aided in confirming the diagnosis. It is imperative to recognize GS to avoid unnecessary surgery. GS should be included in the differential diagnosis of breast tumors, especially in patients with a known history of hematologic disease, and in patients who have breast tumors with diffuse proliferation of small tumor cells [8].

GS appears on mammograms as single or multiple noncalcified, irregular masses with poorly defined margins [6,7]. In our patient, noncalcified nodules with a lobulated ill-defined margin over bilateral breasts were noted. Most GS lesions present as round or oval homogeneously hypoechoic nodules with irregular or circumscribed margins on sonography. Unusually hypoechoic circumscribed nodules with central high echogenicity and a heterogeneously hypoechoic mass with lobulated margins were shown. No definite lateral shadowing or posterior enhancement was seen in the lesions, nor was parenchymal distortion or skin thickening visualized [6]. In our patient, sonography showed irregularly shaped, heterogeneously hypoechoic nodular lesions with ill-defined margin and minimal posterior enhancement. Color Doppler study showed minimal peripheral color flow signal. These findings are different from those with hypervascularity on color Doppler reported in other articles [2,9].

References


