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Primary Breast Lymphoma

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We report the case of a 20-year-old female with lymphoma of the breast. Mammography showed an asymmetric pattern of confluent densities without any discrete mass. Sonography revealed diffuse heterogenous echoic mass intermingled with low-and medium level echoes. We present the clinical, radiographic and histologic features of primary breast lymphoma with a brief review of the literatures.

Key Words: *Breast, Primary breast lymphoma*

INTRODUCTION

Primary lymphoma of the breast is rare, accounting for an estimated incidence less than 1% of all malignant breast primary tumors (Pope et al., 1985; El-Ghazawy and Singletary, 1991). It comprises 2.2% of all extranodal non-Hodgkin's lymphoma (NHL) and 0.38% of all NHL (Misra et al., 1991).

We have recently seen a case of primary lymphoblastic lymphoma of the breast. We present the clinical, mammographic, sonographic and pathologic features of this disease and include a brief review of the literatures.

CASE REPORT

A 20-year-old woman found a huge mass in her left breast with a 2 month history of rapid growth. Her past medial history was unremarkable, and family history was negative for breast malignancies. Examination revealed a 10×10cm, well-demarcated movable tender mass in the left breast. There were no palpable nodes in either axilla, and the right breast was normal. The liver and spleen were not palpable. Bilateral mammography using a dedicated CGR free-standing unit showed an asymmetrical confluent increased density in the left breast without discrete mass margin. There were no associated findings of skin thickening or parenchymal distortion (Fig. 1).

Sonography using a dedicated breast ultrasound unit (ATL-60) showed an irregular large heterogenous echoic mass containing scattered low- and medium level echoes (Fig. 2). There was no significant posterior shadowing of the mass. The normal parenchymal architectures were almost all replaced by the mass. The preoperative diagnosis was of a diffuse infiltrating ductal carcinoma or abscess. The patient underwent a biopsy of the left breast mass, which yielded lymphoma, lymphoblastic type (Fig. 3).

She then underwent abdominal and pelvic ultrasonography, which were normal. Iliac crest bone marrow biopsy yielded permeation of malignant lymphoma. She refused treatment and discharged herself. 1⅔ month later, she was re-admitted with a high fever and palpable enlarged neck lymph nodes. Physical examination of the breasts revealed multiple masses on both sides. Systemic chemotherapy was given.

The chemotherapy regimen included cytoxan, methotrexate, vincristine and adriamycin. During the chemotherapy, the patient died of sepsis due to opportunistic infection.

DISCUSSION

The incidence of primary breast lymphoma (PBL) varies from 0.04% to 0.51% of all breast malignancies (Misra et al, 1991; Hugh et al., 1990). The original criteria for PBL suggested by Wiseman and Liao (1972) are as follows:

1. The availability of adequate pathology material.

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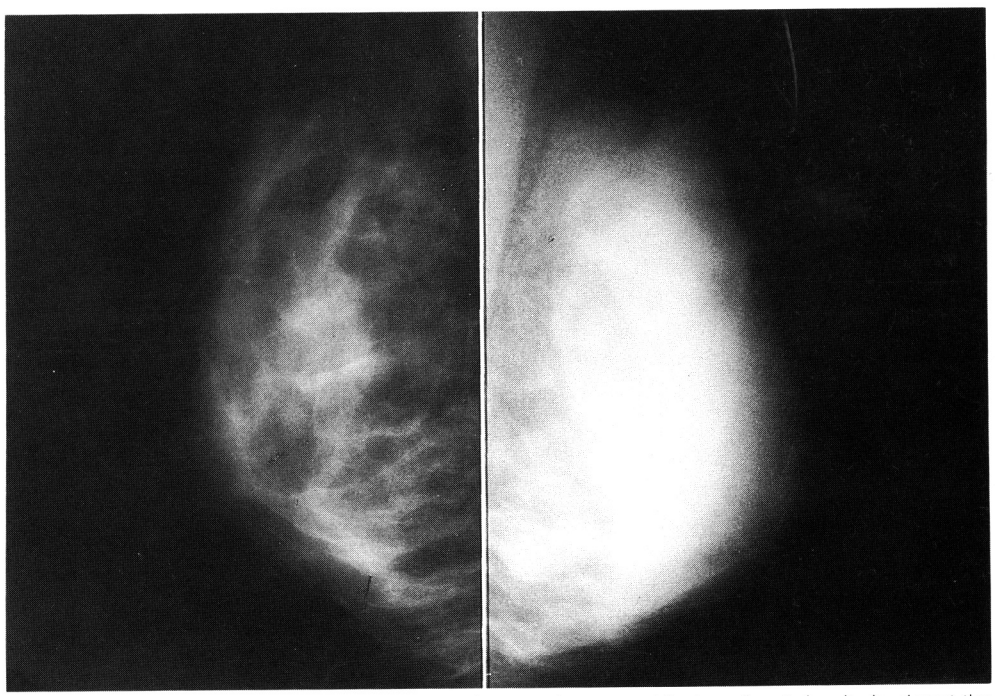

Fig. 1. Mediolateral oblique view of mammography reveals asymmetrical confluent density in almost the entire left breast without any definite margin of mass. There is no overlying skin thickening.

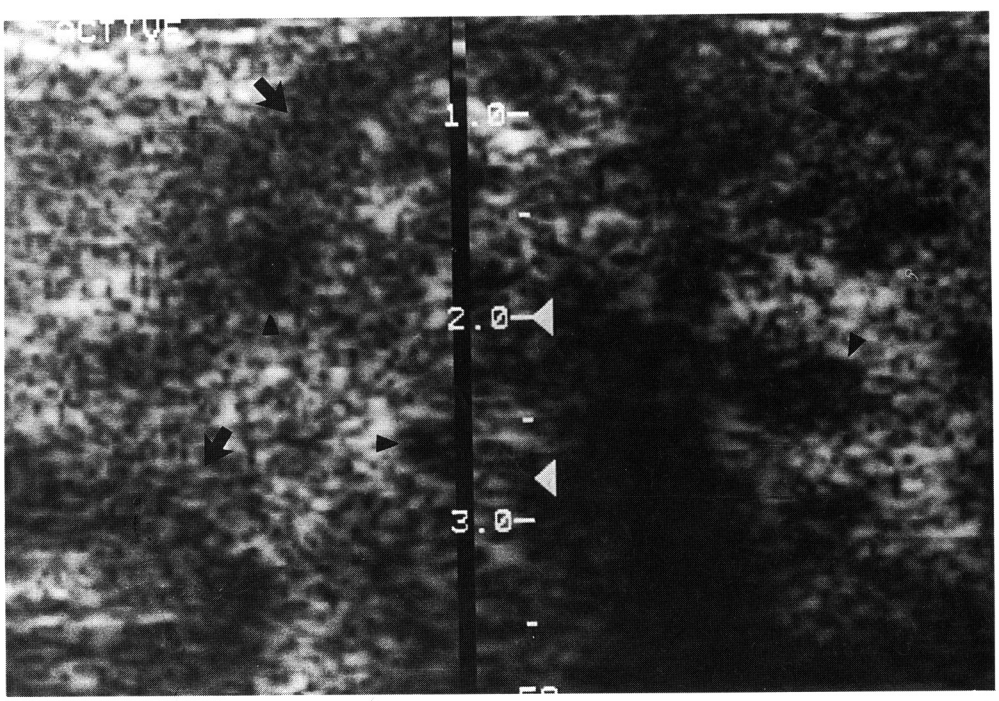

Fig. 2. Sonography of the left breast mass shows a diffuse heterogenous echoic mass. Multiple low (arrows) and medium (arrowheads) level echoes are intermingled within the mass.

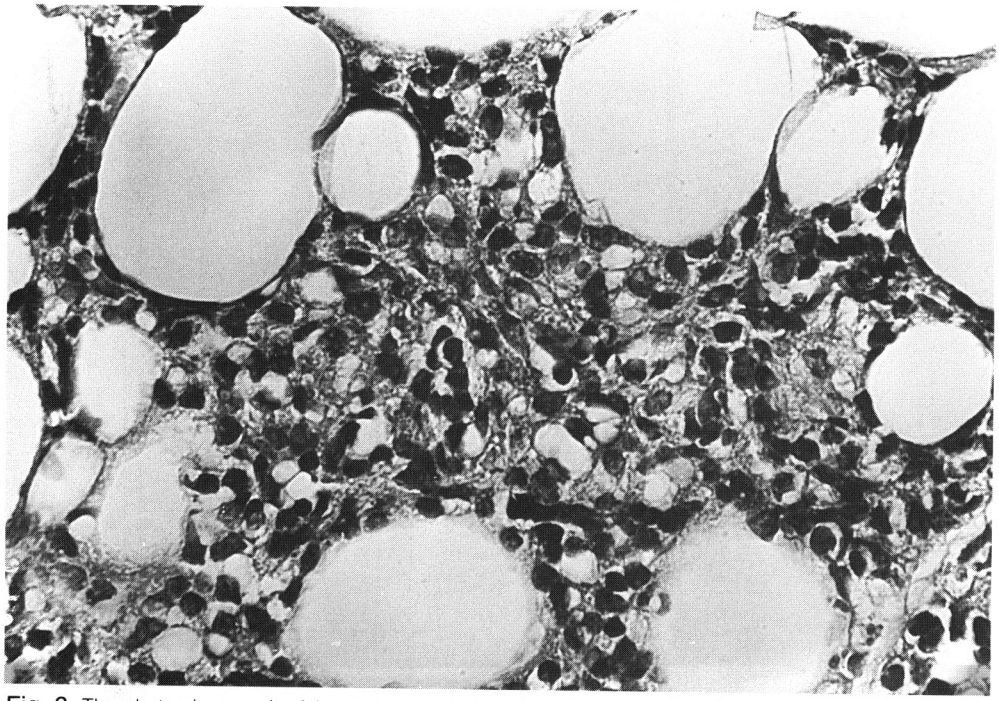

Fig. 3. The photomicrograph of the mass reveals the tumor cells consisted of small lymphoid cells with few indentations.
The tumor cells are positive for leukocyte common antigen (Hematoxylin-Eosin, ×400).

2. Both mammary tissue and lymphomatous infiltrate present.
3. No widespread disease or preceding extramammary lymphoma.
4. Homolateral axillary lymph node involvement, which is considered acceptable.

Even if on subsequent staging investigations the lymphomatous process was shown to involve distant nodal sites (stage III) or bone marrow (stage IV), we diagnosed this case as a PBL as other investigators (Huge et al., 1990; Brunstein et al., 1987; Mambo et al., 1977) did, because the breast was the first or major site of presentation.

The age incidence of PBL is highly variable. It occurs between 9 years to 85 years, but frequently has been seen in the fifth decade, which is also the mean age of women with adenocarcinoma of the breast (El-Ghazawy and Singletary, 1991; Liu and Clark, 1986). Only a a few cases have been reported in males. Sixty percent of the patients showed localization in the superior outer quadrant. Bilaterality is noteworthy, an incidence of 6% to 13% (Wiseman and Liao, 1972; Schouten et al., 1981). There is a right-sided predominence which is difficult to explain. Clinical features are short history, rapidly enlarging breast mass, absence of nipple discharge and retraction. "B" symptoms (10% weight loss, night sweats, fever) are unusual features. And laboratory evaluation is usually normal (Liu and Clark, 1986).

Mammography is of questionable value as a diagnostic aid. But in nodular lymphoma, the typical features are a well-rounded mass, a regular and smooth outline, and a thin perimeter of radiolucency without tumor spiculations or malignant-appearing calcifcations. The mass is somewhat oblong and showed no evidence of desmoplastic reaction. Diffuse lymphoma typically shows generalized asymmetric increased desity and skin thickening (Pope et al., 1985, El-Ghazaawy and Singletary, 1991; Misra et al., 1991; Schouten et al., 1981). But our patient showed only a large asymmetric confluent density without skin thickening, so proper diagnosis could not be made with mammography alone.

Typical ultrasonographic appearance is a well-marginated hypoechoic mass in nodular lymphoma, which is similar with nodular lymphoma elsewhere in the body. In diffuse type, ill-defined hypoechoic area replaces the normal breast parenchyma (Pope et al.,

1985; Zonderland et al., 1991). In our case, the sonography showed an irregular shaped heterogenous echoic mass without posterior shadowing. Multiple low- and medium level echoes were intermingled within the mass. The medium level echoes were more intense than the echoes in carcinoma. Pope (1985) reported a homogenous echo-poor mass containing scattered low-and medium level echoes. Zonderland (1991) presented similar sonographic findings in extramedullary hematopoiesis of the breast.

Histologically most of the breast NHL are of diffuse large-cell (histiocytic) type, however other types of lymphoma are the subject of various reports (El-Ghazawy and Singletary, 1991; High et al., 1990; Wiseman and Liao, 1972; Liu and Clark, 1986; Schouten et al., 1981). The breast is a hormone-dependent member of the mucosa-associated lymphoid tissue and therefore lymphoma may be positive for estrogen receptor. In our case, the tumor cell consisted of small lymphoid cells with few indentations, and were positive for leukocyte common antigen and T-cell markers (UCHL-1), indicating that this was a T-cell lineage tumor. B-cell markers, cytokeratin and EMA (epithelial membrane antigen) were negative.

Initial reports of the therapy for PBL described surgery, consisting of radical or simple mastectomy with or without axillary node dissection, but recent published reports indicate that radical surgery is to be avoided. Subsequently radiotherapy is most commonly used to treat the local site of disease in conjunction with mastectomy or excisional biopsy. For stage III and IV patients, aggressive combination chemotherapy can achieve complete remission rates approaching 50% (Schouten et al., 1981). Only a few authors have compiled 5 year survival figures for PBL. Freeman (1972) has reported a 5 year survival of 33%; a median survival of 60 months has been reported by Liu and Clark (1986). Diffuse histiocytic lymphoma tends to be associated with an unfavorable prognosis. Hugh (1990) reviewed 26% of the recurrent cases at least once with the time to recurrence ranging from 1 to 176 months (median, 8 months).

Although mammography alone shows non-specific findings of breast lymphoma, corrective diagnosis of PBL can be made preoperatively associated with the sonographic finding, similar echo-pattern involving other site.

REFERENCES

Brustein S, Filippa DA, Kimmel M, Lieberman PH, Rosen PP: *Malignant lymphoma of the breast: A study of 53 patients. Ann Surg 205:144-150, 1987.*

El-Ghazawy IMH, Singletary SE: *Surgical management of primary lymphoma of the breast. Ann Surg 214:724-726, 1991.*

Freeman C, Berg JW, Cutler SJ: *Occurrence and prognosis of extranodal lymphomas. Cancer 29:252-260, 1971.*

Hugh JC, Jachson FI, Hanson J, Poppema S: *Primary breast lymphoma. An immunohistologic study of 20 new cases. Cancer 66:2602-2611, 1990.*

Liu FF, Clark RM: *Primary lymphoma of the breast. Clin Radiol 37:567-570, 1986.*

Mambo NC, Burke JS, Butler JJ: *Primary malignant lymphomas of the breast. Cancer 39:2033-2040, 1977.*

Misra A, Kapur BML, Rath GK: *Primary breast lymphoma. J Surg Oncol 47:265-270, 1991.*

Pope TLJr., Brenbridge ANAG, Sloop FBJr., Morris JRIII, Carpenter J: *Primary histiocytic lymphoma of the breast: Mammographic, sonographic, and pathologic correlation. JCU 13:667-670, 1985.*

Schouten JT, Weese JL, Carbone PP: *Lymphoma of the breast. Ann Surg 194:749-753, 1981.*

Wiseman C, Liao KT: *Primary lymphoma of the breast. Cancer 29:1705-1712, 1972.*

Zonderland HM, Michieles JJ, Kate FJWT: *Case report : Mammographic and sonographic demonstration of extramedullary haematopoisis of the breast. Clin Radiol 44:64-65, 1991.*