Extranodal Lymphoma of the Breast

Brandi T. Nicholson, MD[a,*], Rahat M. Bhatti, MD[b], Leonard Glassman, MD[c]

KEYWORDS

• Extranodal lymphoma • Breast • Primary breast lymphoma • Secondary breast lymphoma
• B-cell lymphoma

KEY POINTS

- Extranodal lymphoma (ENL) of the breast is an uncommon disease and typically presents in middle-aged women as a unilateral palpable breast mass.
- B-cell lymphoma is the most common ENL of the breast.
- Imaging is unable to distinguish lymphoma from breast cancer, which is more common; therefore, biopsy is done to establish the diagnosis.

INTRODUCTION

The breast is an uncommon location for ENL to present, occurring in only 2% of all localized ENLs,[1] and ENL encompasses fewer than 0.5% of all breast malignancies.[2] Overall leukemia and lymphoma, however, are the most common sources of extramammary cancer involving the breast.[3,4] Breast lymphoma (BL) typically presents in 1 of 2 clinical scenarios. These include primary BL (PBL) and secondary involvement of breast tissue by a systemic lymphoma/leukemia, or secondary BL (SBL).[5,6] The term PBL is used when a patient has a malignant lymphoma primarily occurring in the breast without previous known lymphoma.[7] A vast majority of lymphomas involving the breast are seen in women; however, rarely, cases have occurred in men.[8–12]

The origins of BL are not completely understood. It has been postulated that the breast can act as a mucosal immune system site with development of lymphoid tissue associated with autoimmune mechanisms or other inciting events, or in general terms the breast may behave as a mucosal-associated lymphoid tissue.[4,13] Some studies have shown higher rates of BL in patients with autoimmune diseases, in particular Sjögren syndrome and systemic lupus erythematosus.[14,15] Given that not all BLs are associated with autoimmune disease, however, other mechanisms must exist. Some have postulated that BL may arise from intraparenchymal lymphatics or intramammary lymph nodes.[13]

SBL is more common than PBL. When PBL occurs, the most common type is diffuse large B-cell lymphoma (DLBCL). DLBCL typically occurs in the 5th or 6th decade with a unilateral, solitary, palpable mass, indistinguishable from a breast cancer.[2,9,16–18] **Fig. 1** shows the common imaging appearance and various pathology of B-cell lymphoma. These patients uncommonly have the systemic symptoms of mastalgia or B symptoms (fever, chills, and night sweats); therefore, diagnosis is usually only obtained after a breast

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[a] Department of Radiology & Medical Imaging, University of Virginia, Charlottesville, VA 22908, USA; [b] Department of Pathology, University of Virginia, Charlottesville, VA 22908, USA; [c] Washington Radiology Associates PC, Washington, DC 20037, USA
* Corresponding author.
E-mail address: bte6v@virginia.edu

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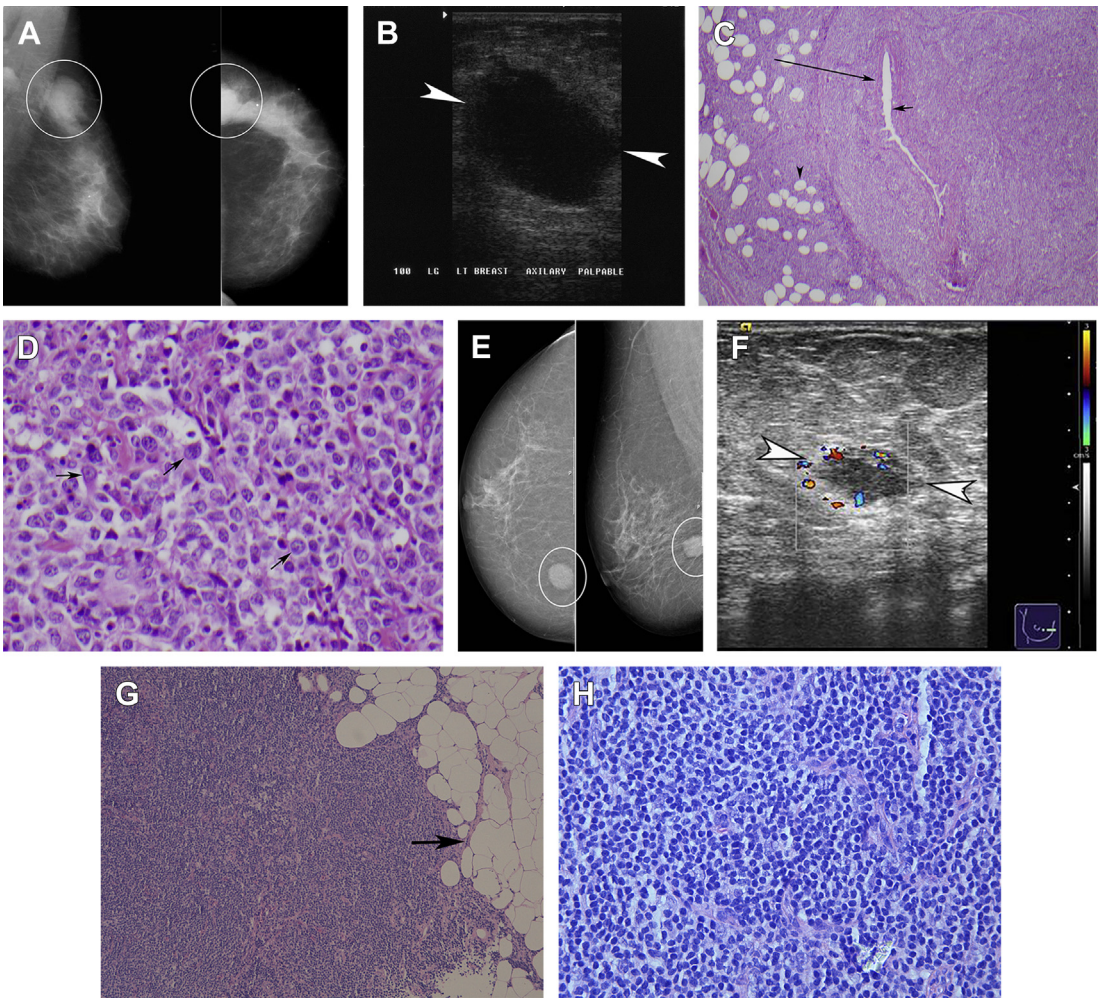

Fig. 1. B-cell lymphoma, both (*A–D*) high-grade (DLBCL) and (*E–H*) low-grade tumor types. (*A*) Diagnostic mammogram, mediolateral-oblique (MLO) and cranial-caudal (CC) views, performed for a palpable left breast mass in a middle-aged woman reveals a round, obscured, equal-density mass in the upper outer left breast (*circles*). (*B*) Subsequent ultrasound shows a round, indistinct, hypoechoic solid mass (*arrowheads*). (*C*) Hematoxylin-eosin stain at low power demonstrates sheets of tumor cells (*long arrow*) in close approximation to a duct (*short arrow*) and mammary adipose tissue (*arrowhead*). (*D*) Hematoxylin-eosin stain at high power shows tumor cells that have large, irregular nuclei consistent with a high-grade process (*arrows*). (*E*) A second patient presented with a screening detected oval, circumscribed, equal-density right breast mass (*circles*) on CC and MLO views. (*F*) Ultrasound shows an oval, indistinct, and hypoechoic solid mass with increased vascularity (*arrowheads*). (*G*) Hematoxylin-eosin stain at low power shows sheets of small cells infiltrating fat (*arrow*) with a diffuse growth pattern. (*H*) Hematoxylin-eosin stain at high power shows small cells with a monotonous pattern, consistent with a low-grade process.

biopsy.[19] Less commonly, PBL presents in a pregnant or recently pregnant younger woman as bilateral high-grade malignant lymphoma. This type of PBL, which is commonly a rapidly disseminating Burkitt lymphoma, tends to spread to the central nervous system, ovaries, gastrointestinal tract, or endocrine organs. **Fig. 2** shows the imaging appearance of a more aggressive Burkitt lymphoma.

Given the rarity of BL, it is not usually suspected in the presentation of a palpable breast mass. It is important to secure a diagnosis via biopsy because the treatment of BL is different from that for breast cancer. Surgery may be done; however, the most successful treatment of BL is a combination of chemotherapy and radiotherapy. Outcomes of PBL differ from those for breast cancer. For PBL, the reported 5-year survival rate is 42% to