

Mucosa-associated lymphoid tissue (MALT) lymphoma of the breast: A case report

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Abstract: Primary breast lymphoma (PBL) is a rare tumor of the breast. Likewise, it is one of the rare manifestations of extranodal lymphoma. Herein we report a case of a 51-year-old Thai woman who presented with a palpable painless mass at the right breast for 10 years without constitutional symptoms, no fever, and no weight loss. Her mass was in ovoid shape, 4.3x5 cm. in size, movable, non-tender but firm consistency and she had no other nodes and no hepatosplenomegaly. The mass was totally excised and its pathological diagnosis was MALT lymphoma. The chest film and the ultrasonography of the whole abdomen were unremarkable, the serum LDH level was normal and the bone marrow was not involved by the lymphoma. The final clinical diagnosis was MALT lymphoma, stage IAE (breast). In general, the cell type of the primary breast lymphoma in almost all cases is DLBCL while the MALT lymphoma that is far more commonly found at the stomach and the salivary glands, has occasionally been mentioned in the breast. Because the MALT lymphoma usually runs rather indolent course and the definite guideline for the treatment of primary lymphoma of the breast has not been yet established, no specific therapy was offered to her. After the total excision of the breast mass, she can survive well without the recurrence at the old or new locations in two years while this paper is written.

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มะเร็งน้ำเหลืองที่เต้านม เป็นมะเร็งที่หาได้ยากของเต้านมขณะเดียวกัน ก็ถือว่าเป็นอาการแสดงที่พบได้ยากอย่างหนึ่งในบรรดามะเร็งน้ำเหลืองที่เกิดนอกต่อมน้ำเหลืองด้วยกัน รายงานนี้เป็นของผู้ป่วยหญิงไทยอายุ 51 ปี มาพบแพทย์ด้วยอาการคล้ำได้ก่อนที่เต้านมขวาเป็นเวลา 10 ปี โดยไม่มีอาการเจ็บปวดที่ก้อนหรืออาการเจ็บป่วยร่วมอย่างอื่น ๆ เช่น ไข้ น้ำหนักลด ตรวจร่างกายพบว่าเป็นก้อนรูปไข่ ขนาดประมาณ 4.3x5 เซนติเมตร ขยับได้ กดไม่เจ็บ แข็งปานกลาง ไม่มีต่อมน้ำเหลืองที่ใด ๆ โต ตับม้ามไม่โต ก้อนเนื้ออกได้รับการตัดออกจนหมด ผลการตรวจชิ้นเนื้อทางจุลพยาธิพบเป็นมะเร็งน้ำเหลืองชนิด MALT เนื่องจากเอ็กซเรย์ปอดและการตรวจด้วยเครื่องสะท้อนคลื่นเสียงความถี่สูงของช่องท้องทั้งหมดปกติ ระดับ LDH ในเลือดปกติและไขกระดูกก็ตรวจไม่พบว่ามีมะเร็งน้ำเหลืองเข้ามาแทรกแซงจึงได้ให้การวินิจฉัยทางคลินิกว่าผู้ป่วยเป็น MALT lymphoma, ระยะ IAE (เต้านม) โดยทั่วไป มะเร็งน้ำเหลืองที่เกิดที่เต้านม เกือบทั้งหมดมักเป็นชนิด diffuse large B cell lymphoma (DLBCL) ในขณะที่ชนิด MALT lymphoma ซึ่งพบได้บ่อยที่ตำแหน่งกระเพาะอาหารและต่อมน้ำลาย พบได้น้อยที่ตำแหน่งนี้เนื่องจากมะเร็ง MALT lymphoma มักมีการดำเนินโรคที่ค่อยเป็นค่อยไปและแนวทางการรักษาโรคมะเร็งต่อมน้ำเหลืองที่เต้านม ก็ยังไม่ได้มีการสรุปให้เป็นที่แน่ชัด การรักษาในผู้ป่วยรายนี้หลังจากผ่าตัดก้อนเนื้ออกในเต้านมออกจนเกลี้ยงแล้ว จึงเป็นเพียงการเฝ้าสังเกตอาการเท่านั้น ขณะที่เขียนรายงานนี้ผู้ป่วยอยู่ได้นาน 2 ปี แล้ว โดยไม่มีการกลับเป็นซ้ำที่ตำแหน่งเดิมหรือที่อื่น ๆ

Introduction

Non-Hodgkin's lymphoma (NHL) is the malignant neoplastic disease of the lymphoid cells however around 10-35 % of the cases of this disease may develop the initial lesion outside the usual lymphoid tissue, the so-called primary extranodal lymphoma. The most common site of the extranodal lesion is the gastrointestinal tract, followed by the skin. Other less common extranodal sites involved by the aggressive NHLs as the primary presentation include the testis, bone and kidney whereas the rarer sites

include the prostate gland, bladder, ovary, orbit, heart, breast, salivary glands, thyroid, and adrenal glands⁽¹⁾.

The breast is considered one of the unusual sites of the extranodal lymphoma, viz., the primary breast lymphoma (PBL) approximately accounts for 1 to 2 % of all extranodal lymphoma. And also, it is a very rare malignancy of the breast, around 0.04 to 0.5 % of all breast cancers⁽²⁾. Other form of breast lymphoma is the secondary involvement of the lymphoma as a part of systemic dissemination⁽³⁾.

In Thailand, lymphoma is rather common but the PBL has been rarely reported^(4,5). Of 389 cases, 214 cases (55.0 %) are found to be the primary extranodal lymphoma and only one case (0.5 %) originates at the breast⁽⁶⁾. Herein, we report one additional case of PBL with unusual microscopic pathology.

Case Report

A 51-year-old Thai woman presented a painless persistent mass at the right breast for 10 years without any constitutional symptoms such as fever, weight loss. During this period, she did not have any significant illness.

The physical examination revealed the mass in ovoid shape, 5x6 cm, at the right breast which was firm in consistency, movable, non-tender and did not fix with the underlying tissue and the covering skin appeared normal. She had no axillary or other lymph nodes, no hepatosplenomegaly and no discharge from the nipple.

Blood tests: Hb 10.9 g%, WBC 6,600/mm³, N 59 %, L 39 %, platelet 291,000/mm³, MCV 64.1 fl, MCH 20.4 pg, RDW 23.0 %, Hb typing: A₂ A, Hb A₂ 2.3 %. The liver and kidney function tests, uric acid and serum LDH were within the normal ranges. The HIV antigen / antibody, VDRL, anti-HCV and HBsAg were all negative.

The chest film and the ultrasonography of the whole abdomen showed unremarkable findings. The mammography showed one large, speculated, solid, heterogeneous mass, 4.3x5 cm, in the right breast, highly suggestive malignancy.

The mass was totally excised under the general anesthesia and the pathological diagnosis was

most likely MALT lymphoma with follicular colonization (CD20+, CD5-, CD23+, cyclinD1-, CD10-, BCL6 non-specific staining, kappa-, lambda-, IgD-). The bone marrow aspiration and biopsy were found unremarkable studies.

The definite clinical diagnosis was MALT lymphoma, stage IAE (breast). And after the total excision of the tumor, no chemotherapy or radiotherapy was offered to her. While this report is written, she can still survive well during two-year follow-up without the recurrence at the previous site or anywhere else.

Discussion

Because the PBL cannot be distinguished from other breast cancers by the palpation and/or the imaging procedures, the definite diagnosis of MALT lymphoma of the breast in our case is established with using the tissue biopsy specially stained by the histoimmunochemistry and it usually displays the characteristic histological features, including the lymphoepithelial lesions and a diffuse infiltrate of marginal-zone "centrocyte-like" B cells⁽⁷⁾.

In most studies, DLBCL is the most commonly mentioned immunophenotype in almost all cases of PBL⁽⁸⁻¹⁰⁾ whereas MALT lymphoma has been hardly seen⁽¹¹⁻¹³⁾. In fact, MALT lymphoma is mostly found at the stomach which is much more common than the whole non-gastric sites including the salivary glands, ocular adnexa, lung and the skin⁽⁷⁾. MALT lymphoma of the breast accounts for 2 % of all non-gastric MALT lymphomas⁽¹⁴⁾. In a review of 203 cases of PBL, 102 (47.9 %) are identified as DLBCL. Thirty-three (15.5 %) are follicular lymphomas, only 26 (12.2

%) are identified as MALT lymphoma, and 22 (10.3 %) as Burkitt or Burkitt-like tumor⁽¹⁵⁾. In Thailand, the phenotype in the majority of PBL is DLBC while in the minority, it is low grade B cell^(4,5).

MALT lymphoma may be found associated with chronic immunologic stimulation, e.g., *H. pylori* infection in case of the gastric MALT lymphoma and the autoimmune diseases such as Sjogren's syndrome in non-gastric MALT lymphoma but our case does not have any chronic infection or autoimmune disease⁽¹⁶⁾.

So far, there is no definite guideline for the treatment of MALT lymphoma of non-gastric sites. Many authorities offer local modalities, such as radiotherapy and/or surgery with or without systemic chemotherapy for the disease within the stage I or II⁽¹⁷⁾. And with these various treatments, the overall response rate is 98 %. The complete remission rate for patients receiving radiotherapy is 96 % vs. only 66.7 % of the patients receiving chemotherapy. Locoregional recurrence (75 %) is more common than distant site recurrence in the patients with limited stage, regardless the treatment modality. The 5-year progression-free survival and overall survival rates are 74.7 % and 95.9 %, respectively. For our patient, we offer her only local surgery because her mass takes a very long time (ten years) to gradually grow and she can survive without any recurrence in two years when this paper is written.

Conclusion

A 51-year old Thai woman felt a painless mass at the right breast for 10 years without local or constitutional symptom. The mass is totally excised and the pathologic diagnosis is MALT lymphoma. The

cell type of the primary breast lymphoma is mostly diffuse large B cell while the MALT lymphoma has been rarely mentioned. And she can survive at least two years after the total excision of the breast mass without chemotherapy or radiotherapy.

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