Bilateral primary breast lymphoma (PBL); case report

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Introduction
Extranodal lymphomas account for 10–40% of all malignant lymphomas [1]. The most common anatomic site of a primary extranodal lymphoma is the gastrointestinal tract (stomach and ileocecal region), but virtually any extranodal location may be a primary site including the skin, CVS, bone, testis, soft tissue, thyroid, etc.

Primary breast lymphomas (PBL) account for 1.7% to 2.2% of extranodal lymphomas, 0.04% to 1.1% of breast neoplasms, and 0.38% to 0.7% of non-Hodgkin's lymphomas [2, 3, 4]. The predominant pathologic type of PBL is diffuse large B-cell lymphomas, but other types can be found [3, 5, 6]. The median age of patients diagnosed with PBL is between 40 and 67 yr, but the range is broad; thus, these tumors can appear in teenagers or patients in their 90’s, the peak age incidence usually is during the sixth decade [7, 15]. All published series reported an overwhelming female predominance [7-9]. The clinical presentation of lymphoma in the breast is similar to other breast malignancy, with a single or multiple painless masses in breast parenchyma. About 13% of patients have bilateral breast involvement. Axillary nodes are involved in 30-40% of cases. Radiographic imaging features of PBL are nonspecific, with the exception that calcifications are rare [10, 11].

In the past, radical mastectomy with postoperative irradiation to the chest and regional nodes was widely used for treatment. Because these tumors respond readily to radiation and to chemotherapy, it seems that these patients could be managing with biopsy or local excision followed by radiation or chemotherapy, usually both.

The role of surgery in PBL should be limited to acquisition of adequate material for diagnosis, typically with a biopsy either from the breast mass or from an involved lymph node. Treatment by mastectomy offers no survival benefit or protection from recurrence [3]. Systemic chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (RCHOP) is currently the standard of care for patients with diffuse large B-cell, and this regimen should be used in patients with PBL as well [12-14]. We report a patient with PBL and solitary pancreatic mass.

Case Report
A 17-year-old girl was referred to Iranian Center for Breast Cancer surgery clinic because of bilateral painless breast mass in November 2006. Family history was negative for breast cancer. Physical exam of the breasts showed bilateral breast mass with irregular margins. The mean
diameter of the masses was 5cm. There was no palpable lymphadenopathy. Ultrasonography revealed bilateral breast mass suspicious to phyllodes tumor. An excisional biopsy of the breasts mass was performed. The pathology report was malignant lymphoma, large B cell type with involvement of surgical margins in parts. The patient underwent staging procedures including ultrasonography and CT-Scan of chest, abdomen and pelvic and bone marrow aspiration. There was a well defined mass; 2.5 cm was detected in body of pancreas. There was no evidence of metastases in other organs.

The patient was referred for chemo–radiotherapy. She received combination chemotherapy CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone). After two course of chemotherapy the pancreatic mass disappeared. At the end of forth cycle she developed severe headache, vomiting and diplopia due to increase of intracranial pressure. Computerized tomography of brain was normal without space occupied lesion. The patient underwent lumbar puncture. Cytology report showed atypical lymphoid cells. Intravenous high dose dexamethason started and the patient symptoms improved. Because of probable central nervous system involvement the patient candidate for eight cycle chemotherapy. General condition of the patient was poor, and she suffered from oral ulcers, loss of appetite, headache, nausea and vomiting. After six course of chemotherapy, echimotic lesions due to thrombocytopenia and perianal abscess worst the patient condition. She was admitted in hematology ward for antibiotic therapy and perianal abscess drainage. Unfortunately after two days the patient was died due to septic shock.

Conclusion
Primary breast Lymphomas is rare compared with primary breast carcinoma. Because of its low incidence, malignant lymphoma rarely is considered in the preoperative evaluation of patients with breast tumors. Furthermore, according to the literature, there are no specific clinical or radiological findings that can lead the clinician to suspect a lymphoma preoperatively (15-19). So, it should be considered in the differential diagnosis of breast masses.

References