

## Case Report



# Primary Cutaneous B-Cell Lymphoma: Diffuse Large B-cell Lymphoma, Leg Type

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### ABSTRACT

Primary cutaneous B-cell lymphoma has only recently been recognized as a distinct clinical entity. Primary cutaneous diffuse large B-cell lymphoma of the legs generally occurs in elderly patients, in particular women. A 70-year-old female patient was admitted to our hospital with her nodular erythematous lesions localized on the left thigh. We performed an ultrasound and color Doppler ultrasound as well as computed tomography. This article will discuss the diagnostic and the treatment options and prognosis of primary cutaneous B-cell lymphoma.

**Key words:** Cutaneous, B-cell lymphomas, Ultrasound.

### ÖZET

#### Primer Derinin B-Hücreli Lenfoması: Diffüz Büyük B-Hücreli Lenfoma, Bacak Tipi

Derinin primer B hücreli lenfoması son zamanlarda ayrı bir klinik antite olarak kabul edilmiştir. Bacaklardaki derinin primer büyük B hücreli lenfoması genellikle yaşlı, özellikle kadın hastalarda görülmektedir. 70 yaşındaki kadın hasta, sol uyluk üzerinde lokalize nodüler eritematöz lezyonlar ile hastanemize başvurdu. Ultrasonografi ve renkli Doppler ultrasonografinin yanı sıra bilgisayarlı tomografi yaptık. Bu makalede, derinin primer B hücreli lenfomasının teşhis ve tedavi seçenekleri ve prognozu tartışılacaktır.

**Anahtar Sözcükler:** Deri, B hücreli lenfomalar, Ultrasonografi.

According to their immunohistological findings, skin lymphomas are classified as B- and T-cell lymphomas, which form two distinct groups in terms of the clinical course and prognosis (1). Extranodal non-Hodgkin lymphomas choose the skin as their second most common location after primary gastrointestinal lymphomas (1, 2).

Primary cutaneous B-cell lymphoma belongs to a group of rare B-cell lymphoproliferative disorders that present in the skin and have no evidence of extracutaneous manifestation at the time of initial diagnosis (3).

Primary cutaneous B-cell lymphoma has been recently described (4, 5). Most of the cases present with nodules and/or tumors. Violet-red colored, flat surfaced nodules and tumors are usually surrounded with small papular lesions, mildly infiltrated plaques and/or figurate erythematous lesions. Deeply localized nodules are commonly situated on lower extremities (1, 6).

With the advent of ultrasound (US) probes with high resolution, there have been great advances in the detection and evaluation of dermatological diseases and

US has gained a critical role in evaluating the diseases affecting skin and subdermal tissues (7). As a result of a case we had, we investigated the contribution of radiological imaging techniques to the diagnosis in primary cutaneous B-cell lymphoma.

### CASE REPORT

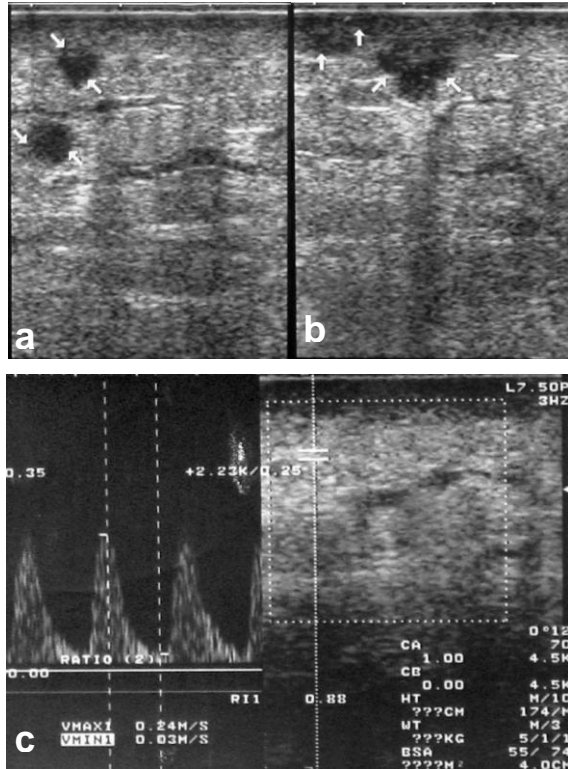
A 70-year-old female patient developed a bluish-black walnut-sized, painless, non-draining mass below her left knee one year ago. The patient was operated on and was free from symptoms until 2.5 months ago. She suddenly started experiencing pain, swelling and skin lesions on the left leg again. Upon physical examination, together with a generalized edema of the left leg, there were nodular erythematous lesions localized on left thigh.

In our department, we performed a US and color Doppler US (CDUS) with a 7.5MHz superficial probe and also computed tomography (CT) on this patient. On the US, on the left hip and thigh region, together with diffuse thickness and echogenicity increase of the skin and subdermal tissues there were multiple small hypochoic nodular lesions (Figure 1a) as well as

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Received/Geliş Tarihi: 09.12.2013

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Accepted/Kabul Tarihi: 17.05.2014

lobulated contoured lesions (Figure 1b). CDUS revealed an increased arterial blood flow rate in this region (Figure 1c). Additionally, there was a hypoechoic mass lesion of 8x6x5cm with lobulated borders on the left femoroinguinal region with subdermal localization.



**Figure 1.** On US, on left hip and thigh region, together with diffuse thickness and echogenicity increase of the skin and subdermal tissues, multiple small hypoechoic nodular lesions (arrows) (a). On US, lobulated contoured lesions were delineated (arrows) (b). CDUS revealed an increased arterial blood flow rate in this region (c).

On the CT, on the region with the lesion, there was a significant degree of diffuse thickening and density increase of the skin and subdermal tissues. On the left femoroinguinal region, the hyperdense mass lesion of 8x6x5cm with lobulated contours, which was localized between subdermal tissues and muscular tissue, was evaluated as conglomerated lymphadenopathy (LAP). The skin that was neighboring the conglomerated LAP anteriorly was thickened and retracted (Figure 2). A chest CT of the case revealed mediastinal LAP. The skin biopsy that was performed reported the diagnosis as diffuse large B-cell lymphoma.



**Figure 2.** On axial CT, on the region of the lesion, there were significant degree of diffuse thickening and density increase of the skin and subdermal tissues, associated with conglomerated lymphadenopathy.

### DISCUSSION

Primary cutaneous B-cell lymphoma represents a heterogeneous group of entities which show variation in histology, immunophenotype and in prognosis. These are follicular lymphomas, marginal zone B-cell lymphomas and diffuse large B-cell lymphomas (1-3, 8). Diffuse large B-cell lymphomas are of the leg type; they manifest in the lower extremity location, and affect elderly patients (mean age 76 years), especially females (1, 3, 9-11). Diffuse large B-cell lymphomas with leg type is an intermediate-grade B-cell lymphoma that comprises only 1% to 3% of all cutaneous lymphomas and approximately 10% to 20% of primary cutaneous B-cell lymphomas (3).

Primary cutaneous diffuse large B-cell lymphoma, leg type, may show features that overlap with other lymphomas. While a variety of primary cutaneous and systemic/extracutaneous lymphomas may show similar features, the combination of clinical findings, morphology, and immunophenotype helps to distinguish this lymphoma from other diagnostic considerations, with both important prognostic and treatment implications for patients (11). Punch (4–6 mm), wedge-incisional, or excisional biopsies are most frequently performed for the diagnosis of primary cutaneous B-cell lymphoma (3).

According to their US appearance, the lesions in non-Hodgkin lymphomas of the cutaneous kind are classified as lesions with focal and diffuse patterns. The focal pattern is described as small (0.4-1.8cm), hypoechoic and with well-defined nodules (Type I), or with multiple nodular structures with the same characteristics, which tend to form a polylobulated hypoechoic patchy area (Type II). These lesions are localized in the dermis, in the subcutaneous layer or in both. The diffuse pattern is described as homogeneous hyperechoic thickening of the dermis (Type III) or as diffuse and unhomogeneous infiltrate involving both the dermis and subcutaneous tissue (Type IV) (7).

B cell lymphoma can demonstrate nodular and/or diffuse patterns on US. In B-cell lymphomas, solitary or

few non-ulcerated regular contoured nodules are together with frequent and early neoplastic lymph node involvement (7). Our case presented a mixture of focal and diffuse pattern findings on US and conglomerated LAP US accompanied the picture.

Although the value of CT is limited in cases at an early stage, dermal thickening and subdermal invasion are investigated by CT. In addition to cutaneous lesions, accompanying LAPs are also identified by CT (12, 13). Miketic (12) in none of the 17 stage I cases, and Bass (13) in 32 of 43 stage I patients could not identify any describable abnormality by CT. On the CT of our case, in the skin and subdermal tissue with the lesion, there was severe diffuse thickening, a density increase and conglomerated LAP in the femoroinguinal region neighboring this region. Additionally, we identified mediastinal LAP on the thoracic CT of our case.

Magnetic resonance imaging (MRI) with good resolution is clinically advantageous in primary cutaneous lymphomas. MRI can preoperatively evaluate the depth and extension of the primary or recurrent skin tumors (14).

Diffuse large B-cell lymphomas are one of the main reasons for the peripheral paralysis of the cranial nerves (15). Although recurrences are quite common in the clinical course, extra dermal invasion and internal

organ invasion are very rare (4). Our case did not have internal organ involvement.

The majority of studies indicate that primary cutaneous B-cell lymphoma is highly responsive to radiotherapy. Polychemotherapy should be reserved for involvement of noncontiguous anatomic sites or those with extracutaneous spread (4). There are publications recommending a combination of chemotherapy and radiotherapy as well (3, 8, 10). The prognosis of primary cutaneous B-cell lymphomas is good (4, 8). Diffuse large B-cell lymphoma, leg type has a poorer prognosis. The 5-year disease-specific survival rates ranged from 43% to 63% in studies (3, 4).

Although high resolution US has a high sensitivity in skin lymphomas, most of the US patterns are nonspecific and can be found in other dermatological diseases as well. Therefore, the specificity of the US is very low without making a histological examination. However US can be effective in select cases such as cases with suspected lesions and skin lesions that develop following the treatment (7). For cases at early stages, CT is of limited value (12, 13).

In conclusion, for cases that are thought to be cutaneous lymphomas as a result of clinical and radiological investigations, the diagnosis should be confirmed with skin biopsy.

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