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Primary cutaneous diffuse large B-Cell Lymphoma, Leg Type: A case report

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Abstract

A rare case of primary cutaneous diffuse large B-cell lymphoma-leg type (PCDLBCL-LT) with abdominal wall lesions was reported. The patient was a 76-year-old female, and a mass on the abdominal wall and left inner thigh was found on CT examination, and the morphology, density, and signal intensity of the lesion were similar on CT and MRI, which the imaging presentation had distinctive features. The final pathology confirmed the diagnosis of primary cutaneous diffuse large B-cell lymphoma-leg type.

Keywords

Lymphoma; Computed tomography; Magnetic resonance imaging; Ultrasonic.

Introduction

Primary cutaneous lymphoma is a type of non-Hodgkin's lymphoma [1]. The primary cutaneous diffuse large B-cell lymphoma-leg type (PCDLBCL-LT) belongs to a subtype of primary cutaneous B-cell lymphoma [2]. The incidence is low, accounting for about 4% of primary cutaneous lymphomas [3], mostly in elderly female patients. The disease is an aggressive lymphoma with a poor prognosis, with an overall 5 annual survival of only 41% [4].

Case Report

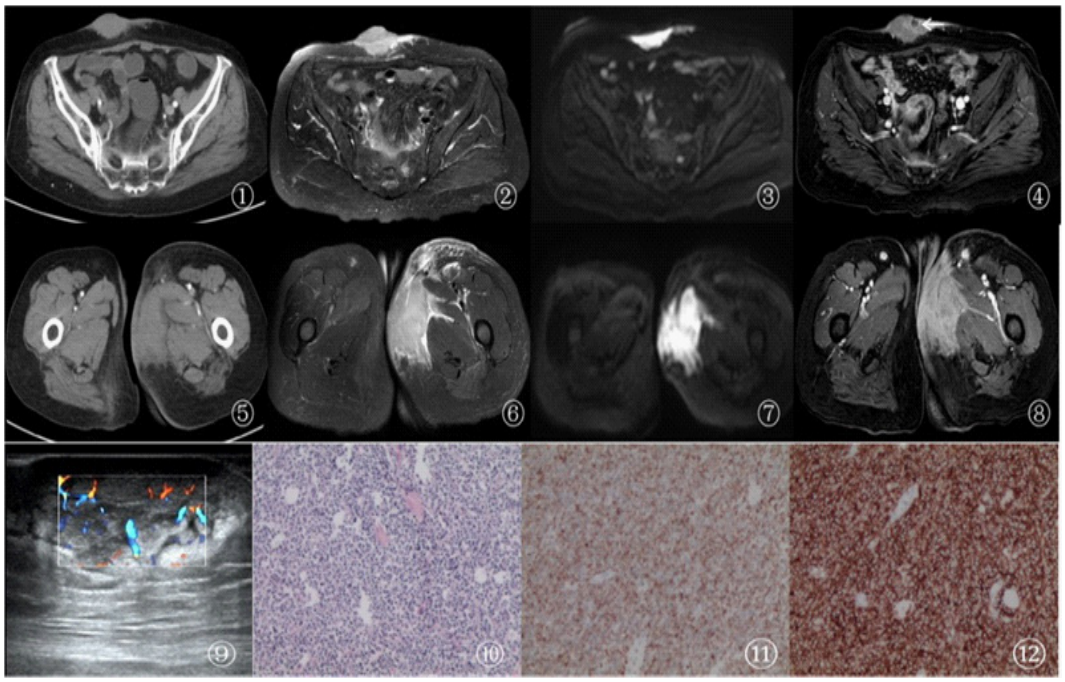
A 76-year-old Chinese female was admitted to the hospital with a subcutaneous mass with rupture in the lower abdomen without apparent cause. The patient presented with a history of subcutaneous mass with growth in the right lower abdominal wall 1 month ago, which recently increased in size with rupture. **Previous medical history:** Surgical treatment for right foot osteomyelitis 10 years ago and history of smoking and alcohol consumption. Physical examination revealed a red, ruptured mass of approximately

5.0 cm in the right lower abdominal wall and swelling in the left medial thigh. **Laboratory tests:** NSE (Neuron-Specific Enolase) 37.00 (<17.00) ng/ml, LDH (Lactate Dehydrogenase) 2731 (109~245) U/L, α-HBDH (Alpha-Hydroxybutyrate Dehydrogenase)2350 (72~182) U/L.

Imaging of the right lower abdominal wall: CT scan showed a spindle-shaped mass of about 5.2 cm × 3.0 cm in size under the skin of the right abdominal wall with a CT value of about 39 HU, uniform density, thickened surrounding skin, and small nodules were seen under the skin in its lower right side. On CT-enhanced scan, the mean CT values were 57, 66, and 61 HU in the arterial phase, portal phase, and delayed phase (Figure 1). **The final CT imaging diagnosis was:** ligamentous fibroma. MRI revealed intermediate signal intensity at T1-weighted imaging, high signal intensity at T2WI-SPAIR (Figure 2), and significantly high signal intensity at DWI sequence (Figure 3), compared with the surrounding muscle. Strips of high signal intensity at T2WI-SPAIR were seen in the adjacent subcutaneous soft tissues. MRI enhancement scan showed that the lesion was homogeneously enhanced, and local ulcerative necrosis was not enhanced (Figure 4). **The final MRI imaging diagnosis was:** lymphoma/sarcoma. Ultrasound demonstrated that a heterogeneous hypoechoic with inhomogeneous internal echogenicity and poorly defined borders was seen in the sub umbilical abdominal wall. The soft tissue around the hypoechoic area was edematous. Red-blue dotted-strip blood flow was evident within the hypoechoic area (Figure 9).

Imaging of the left thigh: CT scan showed increased density of soft tissue in the left inguinal and medial thigh roots, and flocculent shadows were seen. The left thin femoral muscle was poorly displayed, and the CT values were similar to those of the abdominal wall lesion (Figure 5). MR revealed mounded and irregular lamellar high signal intensity at T2WI-SPAIR (Figure 6), and significantly high signal intensity at DWI sequence (Figure 7) in the left groin, perineum, and medial thigh (femoral thin muscle and subcutaneous and skin), which with moderate enhancement (Figure 8). Cotton-shaped high signal intensity at T2WI-SPAIR was seen in the soft tissues surrounding the lesion (Figure 6).

To clarify the diagnosis, further surgery was performed to remove the abdominal wall mass. **Pathological examination:** one piece of dermal tissue, size 10.0 cm × 6.5 mm, with grayish-white and tough cut surface. **Microscopic observation in HE section:** skin tissue with epidermal ulcer formation, diffuse infiltration of tumor cells in the dermis, and subcutaneous adipose tissue with single morphology and loss of follicular structure. Tumor cells were central mother cells with large nuclei, obvious nucleoli, and easy-to-see nuclear division images (Figure 10). Immunohistochemistry demonstrated that the tumor cells expressed CD20, Bcl-2, MUM-1, Ki-67 expression rate was 80% (Figures 11,12); while CK pan, CD3, CD5, CD10, Bcl-6, CyclinD1 were negative. **Pathological diagnosis:** Highly aggressive lymphoma (abdominal), considered primary cutaneous large B-cell lymphoma, leg type.



Figures 1-12: Right lower abdominal wall mass, CT-enhanced scan **(Figure 1),** T2WI-SPAIR **(Figure 2)**, DWI **(Figure 3)**, and MR enhancement **(Figure 4)** show a right lower abdominal wall mass with relatively uniform CT enhancement, slightly high signal intensity in T2WI-SPAIR, stripes of high signal intensity in the surrounding skin, high signal intensity in DWI sequence, showing diffusion restriction, and local ulcer necrosis without significant enhancement in the enhanced scan (white ↑). **Figure 5-8:** Left thigh mass, CT enhancement scan **(Figure1)**, T2WI-SPAIR **(Figure 2)**, DWI **(Figure 3)**, and MR enhancement **(Figure 4)** showed that the CT enhancement and MR signal intensity were similar to the right lower abdominal wall mass, and the local demarcation with the left femoral thin muscle and subcutaneous and skin was unclear. **Figure 9:** Ultrasound showed that the right lower abdominal wall mass was rich in blood supply and the echogenicity was not homogeneous. **Figure 10:** HE low magnification showed diffuse infiltration of tumor cells with a single morphology and loss of follicular structure. The tumor cells were central mother cells with large nuclei, obvious nucleoli, and easy-to-see nuclear fission images. **Figure 11:** Tumor cell pulp was positive for Bcl-2. **Figure 12:** Tumor cell membrane is positive for CD20.

Discussion

Primary cutaneous diffuse large B-cell lymphoma-leg type is most common in the lower extremities bilaterally or on one side, but skin involvement outside the legs is rare [5]. The lesions are mostly purplish-red or red nodules, often growing rapidly, and may ulcerate and spread easily to other parts of the skin. This case is consistent with the relevant clinical manifestations. The density or signal of the solid mass of PCDLBCL-LT, in this case, was relatively homogeneous and not well-defined. Subcutaneous strips of abnormal signal or density shadow reflect the infiltration and edematous presentation of lymphoma, which is consistent with the majority of cases summarized by Carroll et al. Therefore, in middle-aged and elderly patients (aged≥50 years or older) who present with apparent swelling and thickening of the limb, rapid enlargement of subcutaneous nodules, and protrusion toward the skin surface with ulcerative erosion, meanwhile, CT or MRI suggests a nodular solid mass with uniform density or signal, and abnormal density or signal in the subcutaneous fat layer in the form of strips, cutaneous lymphoma should be considered when antibiotic treatment is ineffective.

PCDLBCL-LT has a low prevalence, diverse clinical manifestations, and very few relevant diagnostic imaging aspects are reported in the literature, making it highly susceptible to misdiagnosis, and therefore

needs to be differentiated from the following lesions [2]. **Nodular fasciitis:** It is common in young adults and has a short disease duration, mostly seen in superficial subcutaneous areas as a solitary, solid soft tissue mass. The MRI signal intensity depends on the ratio of intracellular cytoplasm to collagen, extracellular water, and the degree of vascular enrichment in the lesion, The image shows a well-defined mass with few invasions, no edema, and moderate to severe heterogeneous or circular enhancement [7,2]. **Cellulitis:** Mostly seen in the skin, subcutaneous tissue, appendix, and other lax tissues. CT demonstrates inflammatory infiltration of subcutaneous fat with abscess formation, and MRI shows striated or reticulated subcutaneous fat layer and fascia, with slightly low signal intensity in T1WI and high signal intensity in T2WI, but mild signal changes in nearby muscles and no soft tissue mass manifestation [8,3]. **Ligamentous fibroma:** lesions vary greatly in size depending on the site of origin, with diameters generally ranging from 3-20 cm. The image appears as a homogeneous or inhomogeneous mass, and necrosis is rare. Heterogeneous high signal intensity is seen on TWI-FS, and internal bands of the low signal are seen on all sequences [9]. Most of them show more than moderate enhancement with stripes of the non-enhancing low signal intensity internally.

In summary, PCDLBCL has a low incidence, diverse clinical manifestations, and is sometimes difficult to distinguish from some tumors and inflammatory lesions. Imaging plays an important role in the differential diagnosis of PCDLBCL, and clinical confirmation often requires repeated, multifocal sampling with histopathological and immunohistochemical confirmation of the diagnosis.

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