

Case Report

Extra-nodal lymphoma presenting as an abdominal wall mass: a case report

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Abstract: Lymphoma is one of the most common types of malignancy in humans. Approximately 40% of lymphoma patients will manifest extra-nodal lymphoma in certain organs and tissues, such as in the gastrointestinal tract and on the skin. Primary extra-nodal lymphoma that manifests as a mass in the abdominal wall is a rare condition. Only a few case reports have been published on abdominal wall lymphoma. A 69-year-old Chinese woman developed a mass in her abdominal wall because of lymphoma. The initial symptom of the patient was a soft lump fixed on her abdominal wall. The findings of auxiliary examination were consistent with low-grade B-cell lymphoma. The patient received four courses of chemotherapy (rituximab plus cyclophosphamide, doxorubicin, vincristine and prednisone) at three-week intervals as treatment. Diagnosis of lymphoma in primary abdominal wall remains difficult due to the lack of unique characteristics of this disease at first presentation. Image-guided core needle biopsy is essential in diagnosis. MRI may also help in detecting soft tissue lymphoma.

Keywords: Extra-nodallymphoma, abdominal wall, MRI, core needle biopsy, chemotherapy

Introduction

Lymphoma is the seventh most common type of malignancy in human beings [1]. Approximately 40% of lymphoma patients will develop extra-nodal lymphoma in certain organs and tissues [2]. Primary extra-nodal lymphoma that manifests as mass in the abdominal wall is rare; only a few reports have been published on this case in Western studies [3, 4]. A systematic literature review shows that the present study is the first attempt to explore this rare condition in Asia.

Case presentation

A 69-year-old Chinese female came to our hospital to seek medical assistance for a fixed solid mass in her abdomen. She first noticed the mass 4 years ago, but ignored the initial lump and did not go to a hospital. The mass was enlarged in the past month and caused a painful swelling in her abdomen. She did not experience fever, night sweats, or weight loss.

The patient does not smoke nor drink alcohol. One year ago, she underwent a resection of the right saphenous vein because of varicose veins in her right leg. Family history did not include malignancies among first-degree relatives, and the patient did not have previous tumor-related medical history.

Physical examination revealed an average-sized female with no acute distress. Primary signs were as follows: temperature, 99.5 F; pulse, 100 beats per minute; blood pressure, 139/78 mm of Hg; and respiratory rate, 20 per minute. Several enlarged lymph nodes were palpated at both sides of the groin and right axilla. The abdomen appeared swollen on examination, and faint bowel sounds were detected in all four quadrants. A defined and moderately hard mass was palpated in the right anterior abdominal wall measuring 70 mm × 150 mm. The mass appeared fixed to the surrounding tissues.

Peripheral blood count and tumor marker tests were all within the normal range, except for an

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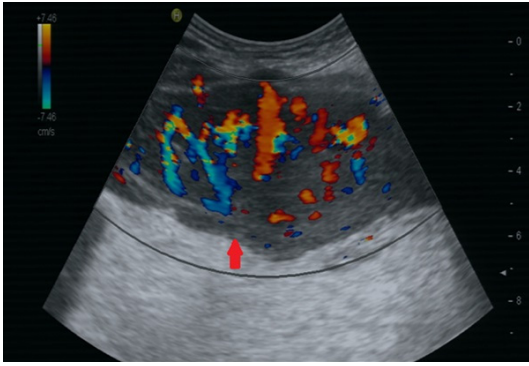


Figure 1. Ultrasonography showed a highly vascularized prismatic mass in the abdominal cavity with heterogeneous hypoechogenicity. And the mass was partly infiltrating into the abdominal muscle layer and even intruded upon fat layer.

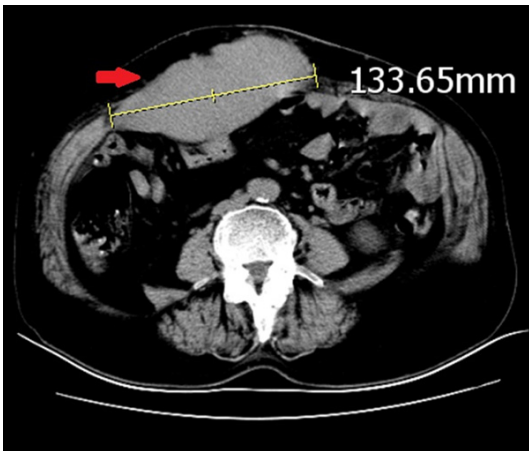


Figure 2. Before treatments, computed tomography of the upper abdomen shows a large soft tissue mass on the right anterior abdominal wall measuring 69*133 mm.

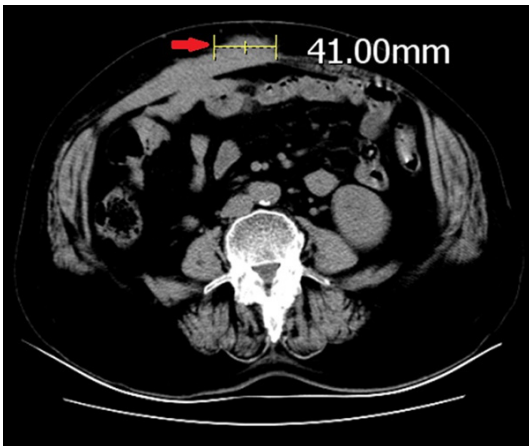


Figure 3. After treatments, computed tomography of the upper abdomen shows a remarkable decrease

of the size of the mass, from 69*133 mm to 41*23 mm.

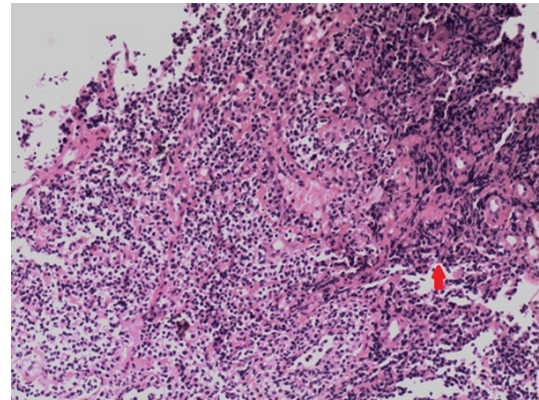


Figure 4. Histopathology results showed a lymphoid proliferation comprising of small-sized cells with round nuclei and dense chromatin.

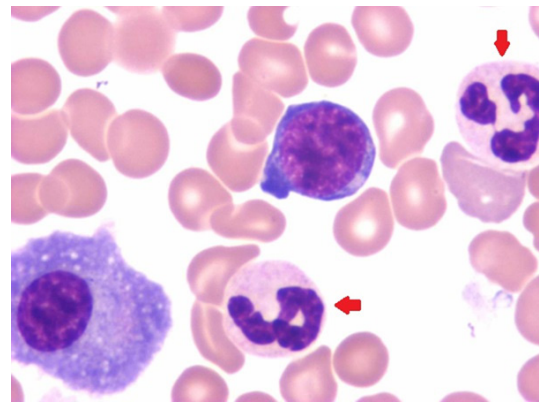


Figure 5. Bone marrow biopsy revealed an active proliferation of bone marrow and a slight decrease of myeloid erythroid ratio.

abnormally increased carcinoembryonic antigen (CEA), which measured 15.3 ng/ml. Ultrasonography was initially used to determine the nature of mass. The result (**Figure 1**) showed a highly vascularized prismatic mass in the abdominal cavity with heterogeneous hypoechogenicity. The upper bound of the mass was under the right costal margin, whereas the lower bound was above the pubic symphysis. The mass was partly infiltrating into the abdominal muscle layer and even intruded the fat layer. To exclude a possibility of involvement of other organs, the CT scan was performed. The CT (**Figure 2**) of the abdomen and pelvis showed that the mass was characterized by mildly heterogeneous soft-tissue density that measured

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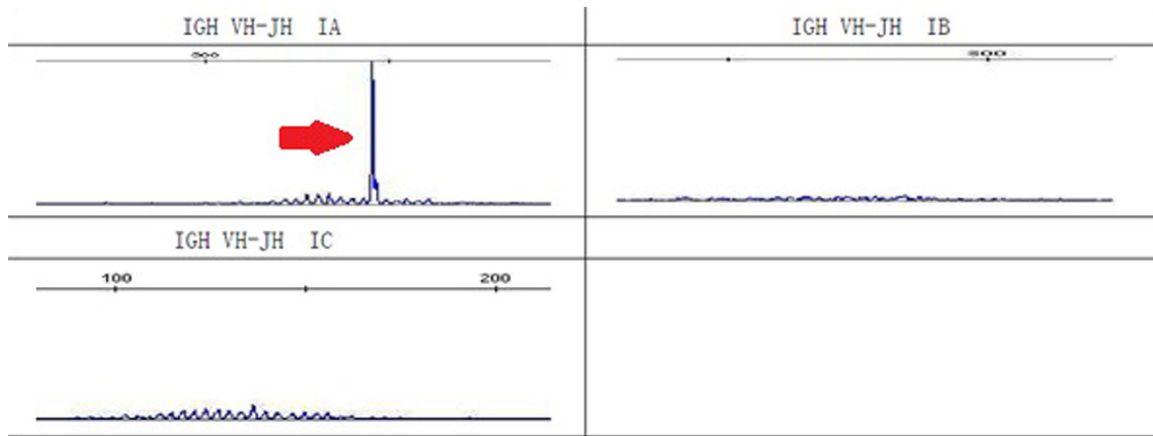


Figure 6. IGH rearrangement test came out to be positive.

69 mm × 133 mm. Several homogeneous nodular masses were observed in the groins and pelvic wall. Further examinations did not find other lesion of cancer. Thus, a malignant tumor probably of lymphocytic origin, such as lymphoma, was suggested.

An ultrasonography-guided core needle biopsy of the abdominal mass was performed on admission to determine the exact source of the mass. Histopathological results (**Figure 4**) showed a lymphoid proliferation consisting of small-sized cells with rounded nuclei and dense chromatin. Immunohistochemical stains on neoplastic cells were positive for CD20 and Bcl-2, negative for CD10 and Cyclin-D, and weakly positive for CD3 and CD5 thereby suggesting a B-cell origin. Ki-67 proliferation rate ranged from 5% to 10%, which was consistent with low-grade B-cell lymphoma. A bone marrow biopsy was performed to determine the subtype and progression stage of the mass and recommend effective chemotherapy regimen. The results (**Figure 5**) revealed an active proliferation of bone marrow and a slight decrease of myeloid erythroid ratio. IgH rearrangement test (**Figure 6**) was positive. A final diagnosis of primary extra-nodal low-grade B-cell marginal zone lymphoma from the soft tissues of the anterior abdominal wall was made.

The patient then received four courses of systemic chemotherapy (RCHOP regimen) at three-week intervals as treatment. Each cycle of chemotherapy last for 5 days, consisting of 1200 mg cyclophosphamide (day 1), 80 mg epirubicin (day 1), 30 mg vinorelbine (day 1), and 90 mg prednisone (days 1 to 5). Six months after

initial of treatment, the size of the mass showed a sharp decrease on CT scan from 69 mm × 133 mm to 41 mm × 23 mm (**Figure 3**). CEA level also decreased to 9.8 ng/ml. No severe side effect was observed. The current condition of the patient is improving.

Discussion

Extra-nodal lymphoma is one of the most common manifestations of lymphoma. Approximately 40% of non-Hodgkin's lymphomas (NHL) manifest as primary extra-nodal disease, which mostly appear in the gastrointestinal tract and skin; CNS is also involved in some cases [2, 5-8]. Extra-nodal disease often ranges from intermediate to high-grade [9]. Coy F. et al. [10] reported that extra-nodal disease occur to persons of all ages, but more than 75% of patients are over the age of 50. Overall five-year survival rate is about 41%; middle-aged female patients have better prognosis than others [10]. Primary soft tissue lymphoma is rare and accounts for 0.1% to 1% of new cases of NHL and 1.2% to 2% of all soft tissue lymphomas [11]. Diffused large B-cell lymphoma is the most common type of soft tissue lymphoma [12]. A review of relevant literature published in English revealed that primary lymphoma in the abdominal wall has been disclosed in two cases only [3, 4]. The present article reviews related papers and provides an additional case of primary extra-nodal lymphoma in a Chinese woman that manifested as a mass in the abdominal wall mass. This is the first case of this rare condition to be reported in Asia.

Clinical features of primary extra-nodal lymphoma vary with manifestation sites [2, 5-8]. For

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Table 1. Clinical information and diagnosis of extra-nodal lymphoma for reported cases

Sources	Age (yr)	Sex	Site of primary lymphoma	B symptoms	CT	Mass size (mm)	Immunohisto-chemical staining	Pathology results	Chemotherapy	Outcomes (months)
Present case	69	Female	Right anterior abdominal wall	NO	Mildly heterogeneous soft-tissue density mass	69*133	CD20 (+) Bcl-2 (+) CD5 (-/+) CD3 (-/+) Cyclin-D1 (-) Ki67: 5%-10%	Low grade B-cell marginal zone lymphoma	RCHOP	6
Alexios M et al. 2013	84	Female	Anterior and left anterolateral abdominal wall	NO	NA	120*93*30 30*40*18 43*31*22	CD19 (+) CD20 (+) slgM (+) Bcl-6 (+) CD5 (-) CD10 (-) MUM1 (-)	Diffuse large B-cell lymphoma	RCHOP	10
George B et al. 2006	68	Male	Left posterolateral abdominal wall	NO	Mildly heterogeneous Soft tissue density mass	155*95*61	CD20 (+) CD5 (-) CD30 (-) Bcl-6 (-)	Diffuse large B-cell lymphoma	RCHOP	10
E Curakova et al. 2014	57	Female	Peritoneum	NO	Thickened nodular and irregular peritoneum affecting abdomen and anterior abdominal wall	NA	CD20 (+) Ki67: high	Diffuse large B-cell lymphoma	NA	Died in 22 days
Catarina O et al. 2014	61	Male	Peritoneum	Loss weight	Diffuse peritoneal thickening	NA	CD20 (+) Bcl-6: 0.5 Ki67 > 90%	Burkitt lymphoma	COPADM	NA
Tina W et al. 2011	38	Female	Left small-bowel mesentery	NO	A large lobulated mass	84*68*75	CD20 (+) CD10 (+) Bcl-2 (+) Bcl-6 (+)	Non-Hodgkin's lymphoma	RCHOP	2.5
Sakshi K et al. 2014	69	Female	The lower abdomen	Loss 20 pounds in 6 months	Two large heterogeneous masses; intraperitoneal implants	200*170*140 65*45	CD20 (+) CD45 (+) CD10 (+) Bcl-6 (+) CD5 (-) Ki67: 95%	Double hit B-cell lymphoma	EPOCH	NA
Paul P et al. 2009	68	Male	Left upper chest wall	NA	Mass suspicious for mesothelioma	60*60	NA	Large cell non-Hodgkin's lymphoma	NA	NA
Xiaoming Q et al. 2014	62	Female	Left anterior chest wall	NO	Solid round mass, involving second and third cartilages	70*70	CD20 (+) Bcl-6 (+) CD10 (-) CD3 (-) Ki67: 60%-70%	Diffuse large B-cell lymphoma	NA	17

RCHOP, rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone; COPADM, cyclophosphamide, vincristine, prednisolone, doxorubicin and methotrexate. EPOCH, etoposide, prednisone, vincristine, cyclophosphamide and doxorubicin. NA: not available.

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conditions in the gastrointestinal tract, patients often complain about difficulty in eating and sharp weight loss in a few weeks [13]. Patients may suffer from mycosis fungoid with cutaneous lymphoma [14]. Pain and fatigue and swelling in the ipsilateral extremity are often observed in primary extremity lymphoma [6]. In conclusion, most of patients with extra-nodal lymphoma do not manifest typical lymphoma symptoms. However, certain changes in experimental indexes, such as abnormal increases of lactate dehydrogenase and abnormal rise of tumor biomarkers, will be readily observed in extra-nodal lymphoma [2, 5-8].

Similar to the other two cases of primary abdominal wall lymphoma, our patient first developed a soft mass in her right anterior abdominal wall. During the evolution of the mass, none of the three patients reported lymphoma-associated symptoms, such as fever, night sweat, steep weight loss, and anemia. The CT scan performed on two of the patients revealed two masses with mildly heterogeneous soft-tissue density fixed on the abdominal wall. The remaining patient underwent MRI. The result showed three enhanced masses in the anterolateral abdominal muscles. Immunohistochemical stains of specimens demonstrated that these lymphomas originated from B cells. The previous two cases were diffused large B-cell lymphomas. Our case was low-grade B-cell marginal zone lymphoma. Both patients received RCHOP regimens with good prognoses. We also examined similar cases, including abdominal cavity lymphoma and chest wall lymphoma, to facilitate comparison with our case. **Table 1** provides additional details and information.

After diagnosis, the lymphoma was classified either as primary or secondary lesion; chemotherapy is usually the first choice of treatment in non-lethal conditions [15]. Clinical features may help distinguish lymphoma in abdominal wall from tumors of mesenchymal origin, such as sarcomas and metastatic carcinomas in soft tissue [6]; however, these features are far from sufficient [16]. Auxiliary examinations should be conducted. Compared with CT and ultrasonography, MRI may have better advantage in differentiating abdominal wall lymphoma from other similar diseases. Chun CW et al. [17] reported that muscle lymphomas are isointense to normal muscle on T1-weighted

images with increased or intermediate signal intensity compared with fat on T2-weighted images with homogeneous diffuse enhancement in most cases.

The cornerstone of accurate diagnosis is pathological examination. Core needle biopsy is an important diagnostic method for excluding potential malignancy or obtaining a histological specimen in known malignancy, usually lymphoma, that requires reclassification for therapies [18]. Biopsy can also offer immunohistochemical outcomes to ensure further identification of subtypes. Image-guided core needle biopsy also reduces the rate of diagnostic surgeries. Compared with surgical examinations, this minimally invasive method will reduce costs and postoperative complications [19]. Core needle biopsy established diagnosis with a sensitivity of 93% in one case series [5].

Chemotherapy is commonly the first choice of treatment for lymphoma. RCHOP showed a remarkable beneficial effect on the outcome of our patient. Coiffier B et al. [15] found that RCHOP increased complete response rate and prolonged event free and overall survival of elderly patients with diffused large B-cell lymphoma without marked clinical increase in toxicity.

Primary abdominal wall lymphoma is a rare form of extra-nodal lymphoma. The various manifesting characteristics of this disease and its atypical clinical and image features lead to diagnostic misconception and sub-optimal treatments. Prompt diagnosis and early chemotherapy are of utmost importance for curing the disease. Image-guided core needle biopsy is essential for diagnosis, whereas MRI may help detect and assess soft tissue lymphoma.

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Disclosure of conflict of interest

None.

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