Case Report

Camper's and Scarpa's fasciae: Anatomic landmark to differentiate between lymph node and abdominal wall involvement in a case of lymphoma

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Abstract

Cutaneous involvement of MALT lymphoma can either be primary or secondary, Secondary spread to the skin is considered disseminated extra-nodal disease involvement, which is classifies as a stage IV disease.

We described a case of a 44-year-old woman with MALT lymphoma of the lacrimal glands with biopsy-proven lymphoma involvement in the subcutaneous tissue of the anterior abdominal wall. This involvement is manifested as multiple small discrete solid enhancing subcutaneous nodules. Although such findings on conventional CT scans are usually non-specific and may present in various systemic conditions, the knowledge of the patient's prior clinical history should alert interpreting radiologists to be aware of cutaneous and subcutaneous involvement of lymphoma. This awareness can help plan the use of imaging-guided tissue biopsy for a minimally-invasive and safe acquisition of tissue specimen for histopathological diagnosis.

Keywords: Cutaneous involvement, Extra-nodal lymphoma, Fascia, MALT lymphoma.

Introduction

The extranodal lymphoma occurred more frequently for non-Hodgkin lymphoma than Hodgkin lymphomas [1]. The most common types of extranodal lymphoma (ENL) are diffuse large B-cell lymphoma (DLBCL) and mucosa associated lymphoid tissue (MALT) lymphoma.

The extranodal involvement is commonly localized in the gastrointestinal tract, followed by head and neck, lung, skin, bone, and brain. Extra-nodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT lymphoma) is a relatively rare subset of non-Hodgkin lymphoma, a cancer of the lymphatic system which arises from the B-cell lineage of lymphocytes. It involves the mucosal epithelium of various organs, such as the stomach, salivary glands, lungs, small bowel, thyroid, ocular, adnexa, and skin. The stomach is the most frequent site of involvement with a strong correlation with Helicobacter pylori infection [2].

As part of initial staging of MALT lymphoma, conventional computed tomography (CT) scan or magnetic resonance imaging (MRI) and imaging of the orbits and salivary glands is acquired. Initial staging is primarily based on Lugano classification [2, 3]. Positron emission tomography (PET) scans in cases of marginal zone lymphoma have been shown to be useful only when localized treatment is planned [2]. This differs from recommendation for primary nodal lymphoma, where the use of PET-CT is considered the gold standard for staging of fluorodeoxyglucose (FDG)-avid nodal lymphoma which includes all histology except chronic lymphocytic leukemia/small lymphocytic lymphoma, lymphoplasmacytic lymphoma/Waldenström macroglobulinemia, mycosis fungoides, and marginal zone NHLs, unless there is a suspicion of aggressive transformation [3].

Cutaneous involvement of MALT lymphoma can either be primary or secondary, where secondary spread to the skin is considered disseminated extra-nodal disease involvement which classifies as a stage IV disease [3]. The primary cutaneous marginal zone lymphoma (PCMZL) presented with multifocal plaques or nodules localized on the trunk and arms [4]. The age of patients with PCMZL

is younger than those with secondary cutaneous marginal zone lymphoma (SCMZL). The average age is being 50 years in PCMZL and 57 years in SCMZL [5]. Imaging findings of cutaneous lymphoma of T-cell lineage are typically described as panniculitis-like, seen as a subcutaneous nodule or thickening, whereas a cutaneous lymphoma of B-cell lineage usually presents with enhancing nodules and infiltrative cutaneous and subcutaneous masses [6]. Bone marrow biopsy and excisional biopsy of three subcutaneous lesions verified DLBCL, stage IV [7].

In this article, we present a classic case of lacrimal gland MALT lymphoma with multiple abdominal subcutaneous enhancing nodules located below Camper's fascia and Scarpa's fascia. This patient was diagnosed with lymphoma stage IV. The whole abdominal CT of this patient showed multiple enhancing nodules beneath Scarpa's fascia and Camper's fascia, which could be misled, as these might be mistaken for lymph node metastasis and determined incorrect staging. Owing to the detection of multiple nodules in CT scans for initial disease staging, the nodules were biopsied and proven to be lymphoma involvement on histopathology. Subsequent resolution on imaging occurred following treatment with chemotherapy.

Case Summary

A 44-year-old woman without an underlying disease or a history of trauma, presented with swelling of both eyelids. She reported no palpable abdominal mass or skin lesions elsewhere. The initial physical examination showed movable rubbery consistency masses at the lateral aspect of both eyelids without any other palpable masses.

Her initial complete blood count (CBC) showed mild anemia with elevated LDH, and blood chemistry was within normal limits. Further CT scan of both orbits was performed which revealed enhancing masses at both lacrimal glands (Figure 1). The patient was then referred to our institution for a right lacrimal gland biopsy, which showed extra-nodal marginal zone lymphoma of the mucosa-associated lymphoid tissue (MALT lymphoma).

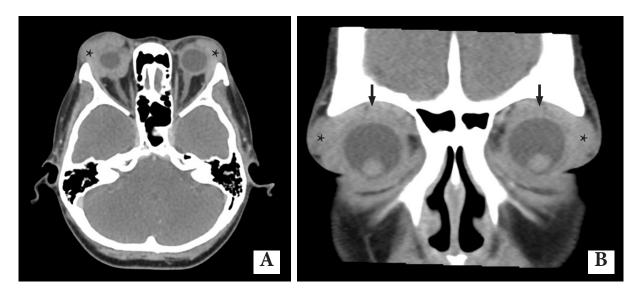


Figure 1. Conventional CT of the orbit in axial (A) and coronal (B) views show multiple enhancing masses involving both lacrimal glands (*), upper eyelids (arrow), and the intra/extra-conal spaces of both orbits. Pathology proven is extranodal marginal zone lymphoma of the mucosa-associated lymphoid tissue (MALT lymphoma).

Additional bone marrow biopsy, and conventional CT scans of the neck, chest, and abdomen were carried out as part of her initial staging. The bone marrow biopsy revealed 60% cellularity with presence of all the cell lineages with maturation. A few scattered small B-cells (CD20+) and T-cells (CD3+) were detected without presence of malignant cells. In our institute, PET scan was not performed.

Her abdominal CT scan revealed borderline hepatosplenomegaly and multiple well-defined enhancing nodules along the subcutaneous layer of the abdominal wall (Figure 2). No other significant lymphadenopathy was detected in the neck, chest, or abdomen.

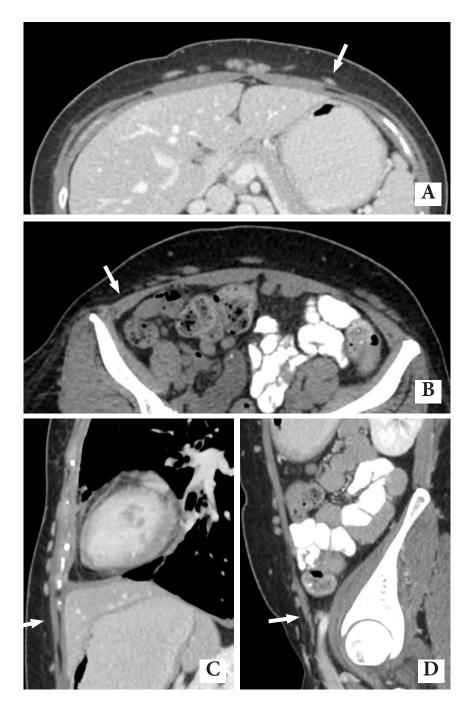


Figure 2. Conventional CT of the whole abdomen axial upper level (A), axial lower level (B), sagittal (C, D) show multiple well-defined enhancing nodules along the subcutaneous layer of the abdominal wall located beneath Camper's and Scarpa's fasciae (arrow).

Due to the patient's underlying MALT lymphoma, cutaneous involvement of lymphoma was suspected. An ultrasound-guided biopsy of the abdominal wall nodule was performed. Ultrasonography revealed an ill-defined subcutaneous hypoechoic lesion at the anterior abdominal wall. The lesion was biopsied, and histopathological results showed a small B-cell neoplasm (CD20+, CD79a+, Bel2+, CD5-, CD23-, CD10-, cyclinD1-, CD3-, CD138+ few plasma cells with kappa light chain restriction (Figure 3).

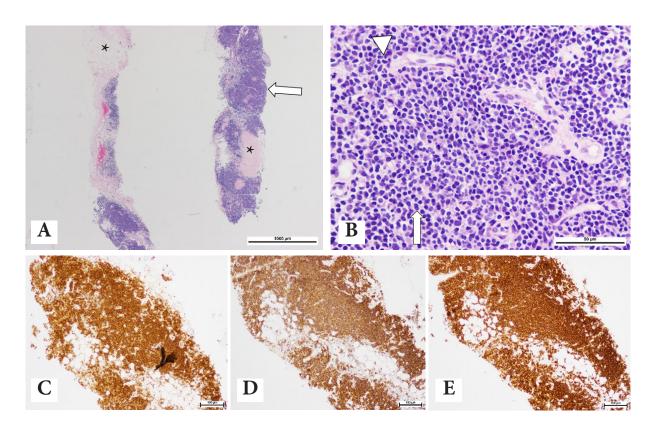


Figure 3. Core tissue needle biopsy of the abdominal wall (A, B) shows atypical lymphoid cells (arrow) with small to medium sizes, infiltrating in fibroadipose tissue (*). Small vessel (arrowhead), lined by normal endothelial cells are noted. (Hematoxylineosin stain; original magnification, x20 and x400). Atypical lymphoid cells are highlighted with immunohistochemical stain for BCL2 (C), CD20 (D), and CD79a (E). The most likely diagnosis of this case is MALT lymphoma.

The patient was diagnosed with stage IV MALT lymphoma with abdominal wall and liver involvement, and was treated with chemotherapy (CHOP). An interim follow-up CT after three cycles of chemotherapy showed disappearance of the lacrimal gland masses and almost complete disappearance of the subcutaneous abdominal wall nodules (Figure 4).

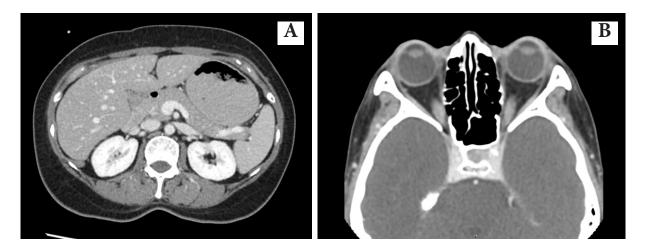


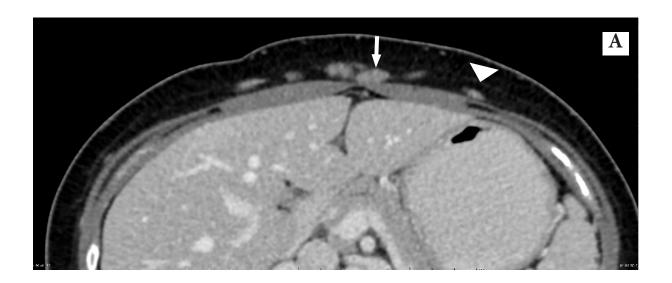
Figure 4. After three cycles of chemotherapy (CHOP regimen) in a case of MALT lymphoma. Conventional CT of the whole abdomen axial (A) shows disappearance of multiple well-defined enhancing nodules along the subcutaneous layer of the abdominal wall. Conventional CT of the orbit in axial (B) shows disappearance of multiple enhancing masses involving both lacrimal glands, upper eyelids, and the intra/extra-conal spaces of both orbits.

Discussion

In our case, the abdominal CT showed multiple small, discrete, solid enhancing subcutaneous nodules without internal calcification or fat density. These enhancing nodules were located beneath Scarpa's fascia and Camper's fascia (Figure 5), which could be misleading as these may metastasize to lymph nodes. According to anatomy of superficial lymphatic drainage of the abdominal wall, lymphovenous anastomosis (LVA) were found originating from the umbilical cord, a midline watershed area, with small diameters (<0.1cm), and above Scarpa's fascia (Figure 5), immediately deep to subdermal venules [8]. Hence, from CT imaging, these nodules located below the Scarpa's fascia and Camper's fascia, suggest secondary subcutaneous involvement. These findings are non-specific, and can be found in various systemic diseases such as post-transplant lymphoproliferative disorders, soft tissue sarcoma, metastasis, or even from subcutaneous injections, resulting in the formation of injection granuloma. Differentiation of these conditions based on imaging findings alone is difficult, and the key to interpreting a diffuse abdominal wall process is knowledge of the patient's clinical history [9]. Nonetheless, histopathology remains the reference standard for the diagnosis of skin pathology, and the role of cross-sectional imaging is primarily for baseline tumor staging in the case of suspected malignancy, preoperative planning, and assessment of treatment response.

Due to our patient's known underlying MALT lymphoma of the lacrimal gland and absence of a prior history of subcutaneous injections, cutaneous involvement of lymphoma was, therefore, suspected. Ultrasound-guided biopsy was chosen due to the superficial location of the subcutaneous nodule and was successfully performed without post-procedural complications.

Interim follow-up CT after three cycles of chemotherapy showed almost complete disappearance of the subcutaneous abdominal wall nodules with a slightly smaller size of the borderline hepatosplenomegaly (Figure 5), suggesting a good response to treatment. In the patient's latest documented follow-up, the fourth cycle of CHOP was administered and the patient will continue to be followed at our institution.



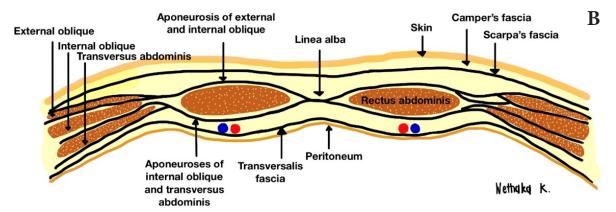


Figure 5. CT abdomen (A) shows multiple small solid enhancing subcutaneous nodules (arrow) located beneath Camper's and Scarpa's fasciae (arrowhead). The drawing (B) demonstrates anatomy of anterior abdominal wall including Camper's and Scarpa's fasciae [10].

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