CT Imaging of Orbital Lymphoma with Pathological Correlation: Case Series

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ABSTRACT
Orbital lymphoma represents a small fraction of all systemic lymphomas that has been accounted for approximately in 1-2% of non-Hodgkin lymphomas (NHL). Some diagnostic delays might be occurred due to the fact that there are non-specific clinical signs. Furthermore, an orbital CT scan can determine the location of the orbital mass and assist in ophthalmological surgery as well as to obtain an optimal sample for an accurate diagnosis. CT scan was performed and revealed the pattern of orbital lymphoma in order to pursue the proper diagnosis. In accordance to the imaging result, immunohistochemical studies were performed from biopsy specimens of all three patients. It showed that there were a non-Hodgkin's lymphomas originating in the orbit, lids, and oculi adnexa.

Keywords: orbital lymphoma; non-Hodgkin lymphomas; orbital CT scan

1.BACKGROUND

Orbital lymphoma is a type of non-Hodgkin lymphoma (NHL) that can originate from the conjunctiva, lacrimal gland, eyelid soft tissue, or extraocular muscles. Nevertheless, it has mostly occurred in the extraconal.[1] Some studies showed that the most common lymphoma has been aroused from the tissues around the eye. It is a low-grade B-cell lymphoma and is also known as Extranodal Marginal Zone Lymphoma (MALT). Sjö LD et al. (2009) stated that more than 50% of orbital and ocular adnexal lymphomas originated from the MALT lymphoma subtype; however, diffuse large B-cell lymphoma (DLBCL) predominated in the intraocular lymphoma.[2,3] Furthermore, these lymphomas have been aroused in lymphoid tissue and acquired at certain extranodal sites because of chronic inflammation or autoimmune disorders. Such tumors were likely to be found in the 5 to 7 decades of life which were predominantly occurred in females. The occurrence of lymphoma orbital infiltration is characterized by a palpable mass' texture can be hard or spongy.[2] As a result, the emergence of pain is an uncommon symptom.[4] In terms of characterizing and evaluating the extent of this disease, imaging plays an important role. In addition, proper consideration should be accounted to the anatomical compartment involved and the tissue which was originated from the aroused neoplasm. This mechanism is conducted to evaluate the imaging of all orbital disease processes. In this study, the following features as the clinical, histopathological, and imaging of orbital lymphoma in the adult population were discussed.[5]

2. CASE

2.1. CASE 1

A 50-years-old woman presented with bilateral orbital swelling and pain. Moreover, an orbital CT scan has been performed and revealed a solid extraconal mass found in the lateral-inferior lid of the right oculi with involvement of the right lacrimal gland and the inferior lid of the left oculi (Figure 1). In

addition, histopathological examination and immunohistochemical analysis of incision biopsy were obtained from the right and left eyelid and suggesting a Mantle Cell Lymphoma.

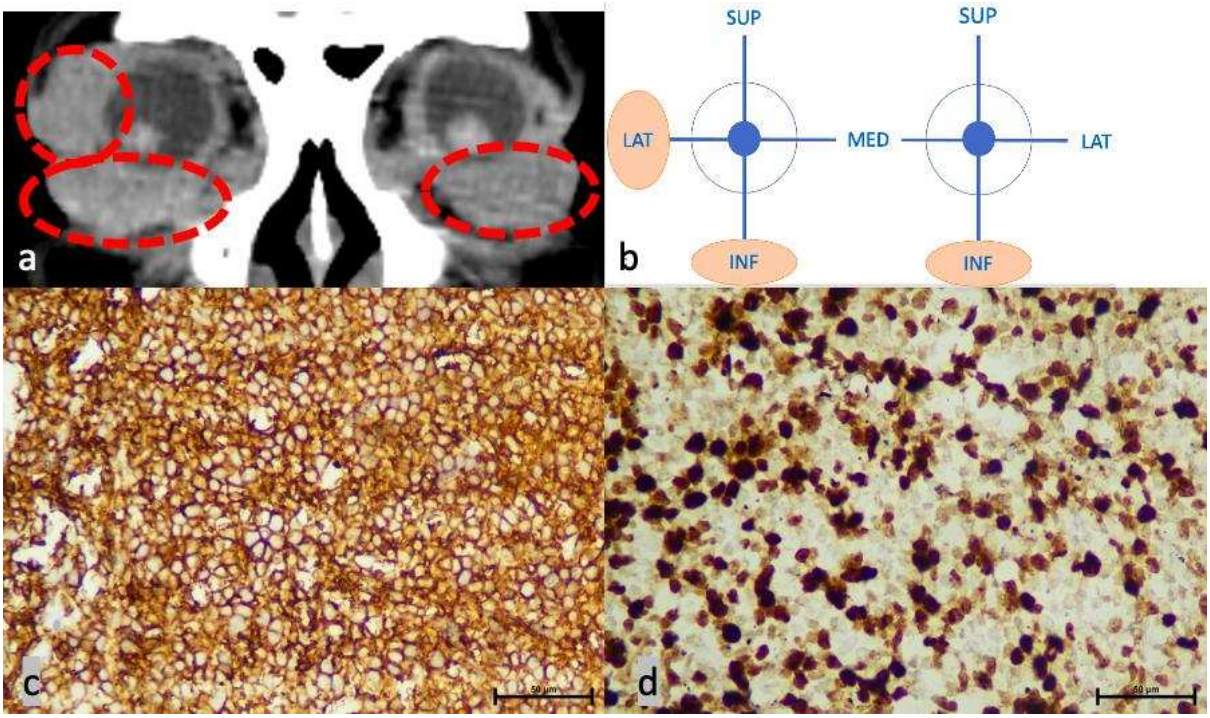

Figure 1. Mantle Cell Lymphoma, High Grade in a 50-years-old-woman. (a) The coronal view revealed bilateral orbital lymphoma: homogenous masses were indicated in a red circle in lateral et inferior quadrant of the right oculi and inferior quadrant of the left oculi (c) Immunohistochemical studies revealed that the neoplastic cells expressing CD20 (400x magnification) (d) Ki-67 is performed for evaluation of the mitotic index, which highlighted 80 percent of the neoplastic cells (400x magnification)

2.2. CASE 2

A 74-years-old woman with a history of right orbital proptosis for a year. In this case, an orbital CT scan has been performed and revealed a solid mass in the right orbital cavity filled intra and extraconal which extended to the superolateral side of the right periorbita. Moreover, it infiltrated the lateral, superior, inferior, and partially medial rectus muscles, superior et inferior oblique muscles, right optic nerve. Thus, it extended anteriorly to push the right bulbi oculi causing proptosis (Figure 2). Orbitotomy was performed and the histologic features of orbital tissue demonstrated Extranodal Marginal Zone Lymphoma of Mucosa-Associated Lymphoid Tissue (MALT) type.

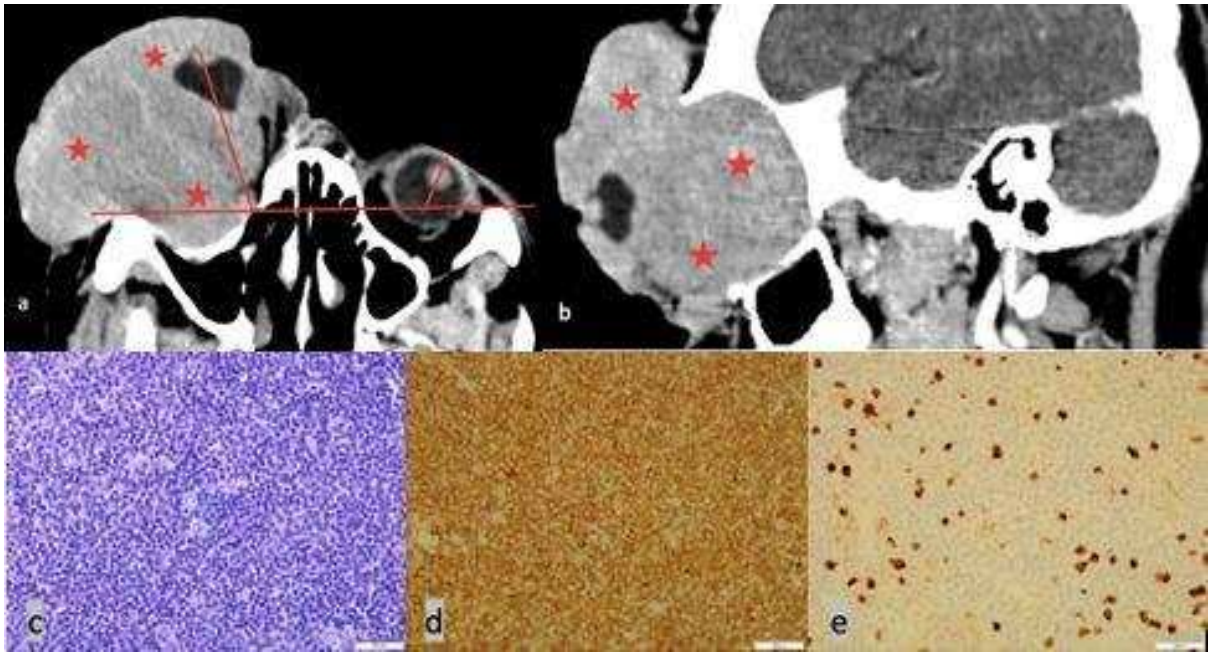

Figure 2. Extranodal Marginal Zone Lymphoma of MALT type found in a 74-years-old-woman.(a) Axial and (b) sagittal view revealed a solid mass (red star) in the right orbital cavity extending anteriorly to push the right bulbi oculi causing proptosis (c) The smears revealed proliferation of anaplastic lymphoid cells with round nuclei, small to medium-sized monotonous population, clumped chromatin (H and E; 400 x magnification) (d) Immunohistochemical studies revealed the lymphoid cells were CD20-positive (400x magnification) (e) The figure corresponding of T-cell marker revealed negative immunostaining with CD3 (400x magnification). So, the presence of a monotonous population of CD20-positive intraepithelial lymphocytes supported a diagnosis of MALT-type

2.3. CASE 3

A 43-years-old man presented with left proptosis and indicated limited eye movement. Furthermore, an orbital CT scan has been performed and revealed a solid mass in the left orbital cavity filled intra and extraconal which infiltrated the lateral, medial, superior, inferior rectus muscles, superior et inferior oblique muscles, encasement of the left optic nerve. It caused proptosis of the left bulbi oculi to the anteroinferior side (Figure 3). The result of excisional biopsy from left ocular adnexa demonstrated Extranodal Marginal Zone Lymphoma of Mucosa-Associated Lymphoid Tissue MALT type.

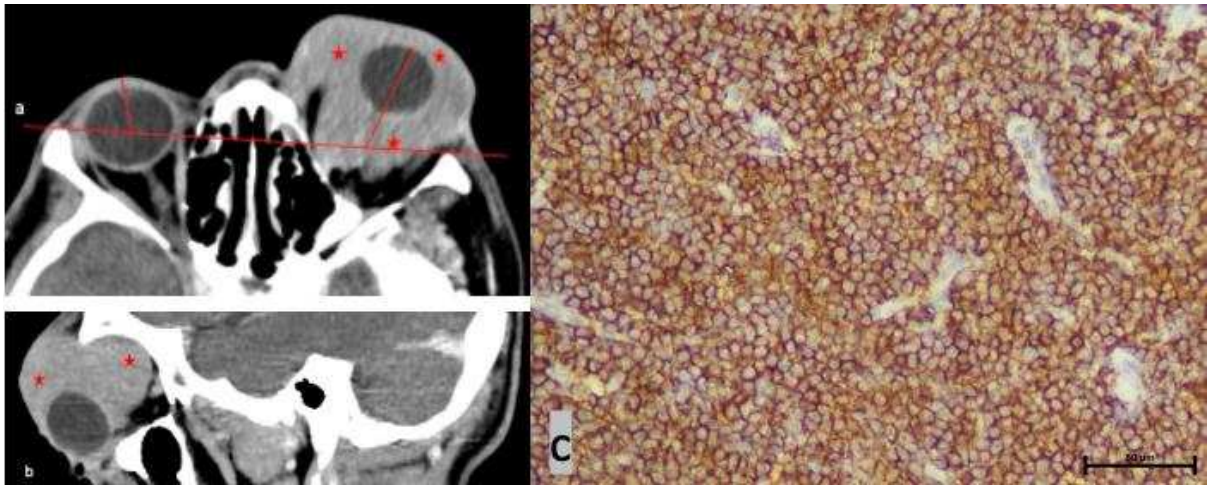

Figure 3. Extranodal Marginal Zone Lymphoma of MALT type found in a 43-years-old-man (a) Axial and (b) sagittal view revealed a solid mass (red star) in the left orbital cavity causes proptosis of the left bulbi oculi to the anteroinferior side (c) Immunohistochemical studies revealed the lymphoid cells were CD20-positive (400x magnification)

3.DISCUSSION

Orbital lymphoma is a rare case and represents about 1 to 2% of lymphomas; however, these entities represent the majority of orbital malignancies. The predominant subtype of non-Hodgkin's lymphoma involving the orbit and ocular adnexa is Extranodal Marginal Zone Lymphoma. In addition, less common subtypes of B-cell lymphoma includes; follicular lymphoma, diffuse large B-cell lymphoma, plasmacytoma, lymphoplasmacytic lymphoma, mantle cell lymphoma, and even hairy cell leukemia.[6]

The most common symptom of orbital lymphoma is unilateral proptosis. In terms of physical examination, orbital lymphoma may have the classic "salmon-patch" appearance i.e. restriction of eye movement, periorbital edema, ptosis, hyperemic conjunctiva, and occasionally a firm palpable mass. Moreover, the timing of symptoms may vary between lymphoma subtypes. Because the symptoms are non-specific, the diagnosis of orbital lymphoma is often delayed.[7]

Therefore, radiological imaging studies could not assess the possibility to differentiate between benign and malignant lymphoproliferative disorders. Most of the lesions found in 76% of cases are categorized as unilateral and often extraconal. As estimated as 40% of the cases involve the lacrimal gland. In terms of an orbital CT scan, it was found that half of the lesions are diffuse with ill-defined borders, the other half is smooth and well-defined masses.[5] The solid mass was homogeneous, isodense, and showed moderate homogeneous contrast enhancement on contrast administration.[3] In addition, orbital lymphoma rarely involves bone erosion; however, it can occasionally occur in diffuse large B-cell lymphoma.[5]

In this study, we discussed the pattern of involvement in diagnosing orbital lymphoma including quadrant, bilateral, distribution, and orbital structure.

3.1. Quadrant

In terms of the number of affected quadrants, all cases were occurred in the lateral-inferior quadrant. The superior-medial quadrant was involved secondary.

3.2. Bilaterality

Most cases had unilateral involvement which was found in two cases, whereas bilateral involvement was presented in a case.

3.3. Distribution

According to the classical division, the orbit is divided into an extraconal compartment, which is located on the outside of the muscle cone, and an intraconal compartment, which is located on the inside of the muscle cone. In this study, the distribution of orbital lymphoma cases was based on their frequency. The findings were two cases occurred in the intra and extraconal and only a case occurred in the extraconal.

3.4. Orbital Structure

The orbital structures which are involved by orbital lymphoma, the lateral, inferior rectus muscles and lacrimal glands occurred in almost all cases. Furthermore, involvement with the optic nerve was reported in two cases.

Table 1. Findings from the three cases of orbital lymphoma

Quadrant	Bilaterality	Distribution	Orbital Structure	Histological
Case 1**: Lat-Inf**	Bilateral	Extraconal	Rectus muscle, lacrimal gland	Mantle Cell Lymphoma
Case 2**: Sup-Lat-Inf**	Unilateral	Intra-Extraconal	Rectus muscle, lacrimal gland, optic nerve	Extranodal Marginal Zone Lymphoma of MALT-type
Case 3**: Sup-Lat-Inf-Med**	Unilateral	Intra-Extraconal	Rectus muscle, lacrimal gland, optic nerve	Extranodal Marginal Zone Lymphoma of MALT-type

4.CONCLUSION

This study portrays a pattern of lymphoma localization that will be beneficial to radiologists in the diagnosis of orbital masses differently. Furthermore, an orbital CT scan can lead to the proper diagnosis of orbital masses, although a biopsy examination is still required. In short, imaging can assist the process of sampling biopsy and surgery for the ophthalmologist.

Conflict of Interest

The author affirms no conflict of interest in this study

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