CASE REPORT

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An Unusual Presentation of Orbital Lymphoma with a Temporal Mass

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Abstract

Orbital lymphomas (OLs) account for 2% of all lymphomas and only 1% of all non-Hodgkin's lymphoma (NHL). However, in patients >60 years of age, lymphomas are the most common primary orbital tumor. The majority of cases are primary, low-grade, B-cell, NHL.The most common sub-type is extranodal marginal-zone B-cell lymphomas of mucosa-associated lymphoid tissue (MALT) type lymphomas. OLs other than MALT lymphoma have different histologies, with diffuse, large, B-cell lymphoma being relatively common. The presentation of OL is variable based on the involved part of the orbit. Accurate diffferentiation of OL from benign lymphoproliferative disorders is crucial due to the differences in management. Imaging characteristics and the clinical findings should be carefully evaluated, and lymphomas should be considered in the differential diagnosis of slowly growing periorbital/orbital region masses in adults. We report an unusual presentation of a primary OL with a rubbery temporal region mass in a 59-year-old woman.

Keywords: Orbital lymphoma, temporal mass, benign lympho-prolferative disorders

INTRODUCTION

Lymphoproliferative diseases of the orbit is a spectrum of disorders that includes lymphoid hyperplasia, atypical hyperplasia, and lymphoma. Orbital lymphoma (OL) may be a manifestation of systemic lymphoma or may occur primarily in the orbit. OLs account for 2% of all lymphomas and only 1% of all non-Hodgkin's lymphomas (NHLs) (1). However, in patients >60 years of age, lymphomas are the most common primary orbital tumor (2). The majority of cases are primary, low-grade, B-cell, NHL.The most common sub-type is extranodal marginal-zone B-cell lymphomas of mucosa-associated lymphoid tissue (MALT) type lymphomas (3,4). Although it is unclear whether MALT exists normally in the ocular adnexa, including the conjunctiva, lacrimal gland, orbital fat, eyelid, and lacrimal sac, the orbit is otherwise believed to be devoid of lymphatic tissue or lymphatic drainage. Therefore, this lymphoid tissue is thought to be acquired as a result of chronic inflammation or autoimmune disorders like systemic lupus erythematosus (3,5). One of the recently described possible associations of ocular adnexial MALT lymphoma (OAML) is *Chlamydia psittaci* infection, which has lead to an antibiotic therapy being used to reduce the size of the tumor, and in some cases to remission (3,6,7). On the other hand, OLs other than OAML tumors have different histologies with diffuse, large, B-cell lymphoma being relatively common (8). The presentation of OL is variable based on the involved part of the orbit. We aimed to report a non-OAML primary OL presenting with a rubbery temporal region mass in a 59-year-old woman.

CASE PRESENTATION

A 59-year-old woman presented to our hospital with a left frontotemporal swelling involving the outer periorbital area which had become more obvious in the last 1 month but was present for about 6 months. Physical examinations including neurological evaluations did not reveal any pathological findings. Globe movements were normal in all directions. Magnetic resonance imaging (MRI) was performed and revealed a 90x33x250 mm sized left temporal region mass with a large

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extracranial temporal component covering the suprazygomatic temporal muscle, which ended inferiorly by obliterating the retroantral (buccal) fat. A small, intraorbital, extraconal extention of the mass at the region of the lacrimal gland in the left superolateral orbital quadrant was revealed without an intraconal component. The mass was isointense to extraocular muscles on T1w images and hyperintense on T2w images. It was involving the lacrimal gland and the lateral rectus muscle. Mild proptosis was noted. After gadolinium, moderate homogenous enhancement was present. The globe, retrobulbar fat, and the optic nerve appeared to be normal (Figure 1). On non-enhanced CT (NECT), the mass was homogenous and slightly hyperdense to the extraocular muscles with no adjacent bony destruction (Figure 2). She had no relevant medical history of chronic inflammatory or autoimmune diseases. Fine needle aspiration biopsy was performed, and histological examination proved non-OAML OL (Figure 3) which was revealed to be a primary by systemic evaluation.

DISCUSSION

OLs may be seen as smooth, circumscribed, unifocal lesions affecting any orbital structure or may emerge as diffuse, infiltrative masses. However, different from the other aggressive intraorbital malign lesions, they have a tendency to slowly mold the adjacent orbital structures without infiltration or destruction. In these cases, the adjacent intraorbital structures are usually seperately identifiable from the tumor as not being the origin. Although close contact may occur, since direct infiltration of the globe and optic nerve is rare, vision is preserved in the majority of cases (3,9). There may be bony remodeling without erosion or hyperosteosis. The majority of OLs are unilateral and are typically located in the superior quadrants, particularly at the lateral part in close proximity to the lacrimal gland, which is involved in nearly 40% of cases (2). Patients usually present with a palpable, firm, soft tissue mass with possible proptosis, exophthalmos, diplopia, or restricted ocular motions depending on the involved orbital structures (3,4). In patients with conjunctival involvement, which accounts for 25% of the cases, a salmon-red patch of a

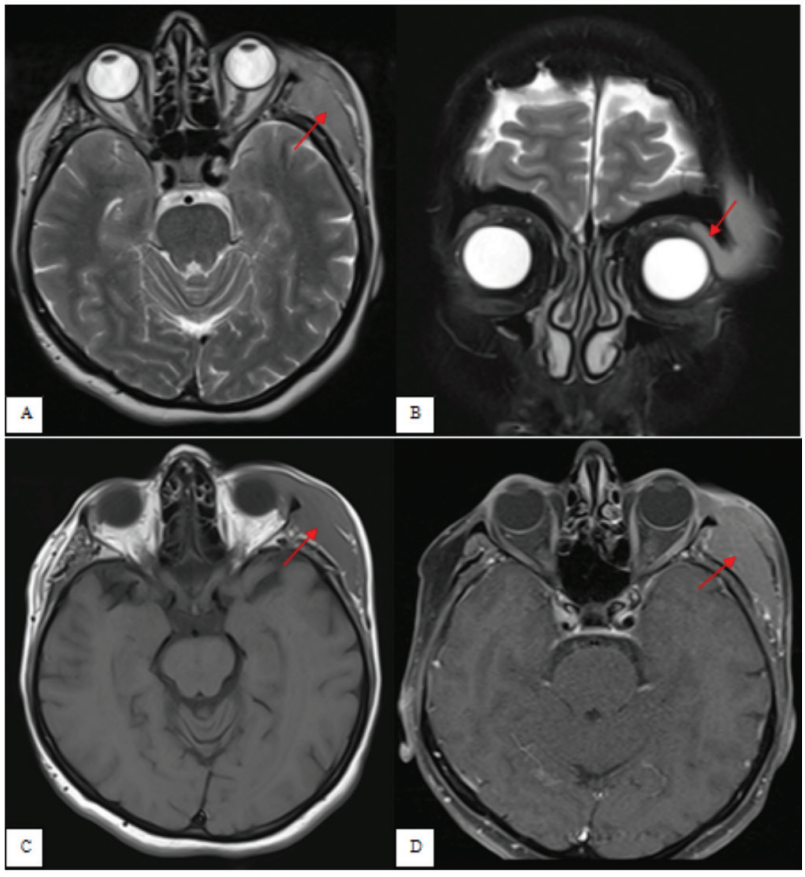

Figure 1. MRI of the brain and the orbit showing the left temporal region mass on axial T2w image (A, arrow). Intraorbital extention of the mass near the lacrimal gland in the left superolateral orbital quadrant is shown on coronal T2w image (B, arrow). The mass is hyperintense to extraocular muscles on T2w images (A, B) and isointense on T1w image (C). On postcontrast T1w image, homogenous moderate enhancement of the mass is seen (D, arrow)

MRI: Magnetic resonance imaging

swollen conjunctiva may be the presenting manifestation, which is usually seen in cases of OAML (3). Although pain is uncommon as a general feature of lymphoproliferative lesions, it may occur in a small subset of patients with inflammatory changes like periorbital edema (3). Most of them are extraconal with a possible intraconal extention for large sized tumors, but intraconal occupation is predominantly seen in cases of orbital lymphoid hyperplasia and is not expected from OL (9,10). On NECT, the mass is usually homogeneous in attenuation, isodense, or slightly hyperdense compared to the extraocular muscles. On MRI, the mass is iso- to hypointense to the extraocular muscles on T1w images and iso- to hyperintense to muscles on T2w images. Following administration of contrast on either CT or MRI, homogeneous moderate enhancement is demonstrated, similar again to extraocular muscles and the lacrimal gland (3,10,11). In the differential diagnosis in our patient, due to the homogenous and non-destructive nature of the lesion, aggressive orbital and lacrimal tumors or metastasis have not been considered. The densely cellular nature of OL with a high nucleus-to-cytoplasm ratio results in relatively specific imaging manifestations including

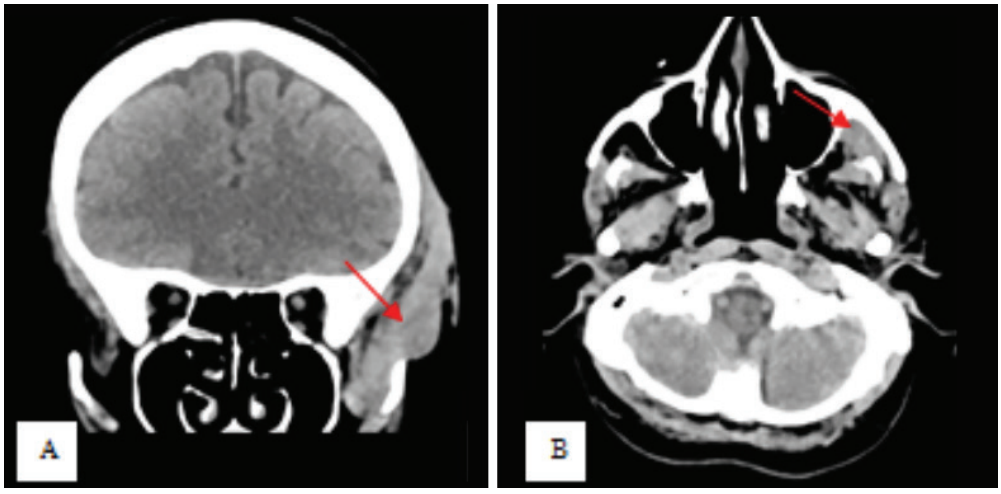

Figure 2. CT of the head showing the isodense left temporal region mass on coronal image (A, arrow). On axial images (A, B), the inferior extention of the mass obliterating the retroantral (buccal) fat at the left infratemporal fossa is seen (B, arrow). Small intraorbital part of the mass is shown (C, arrow)
CT: Computed tomography

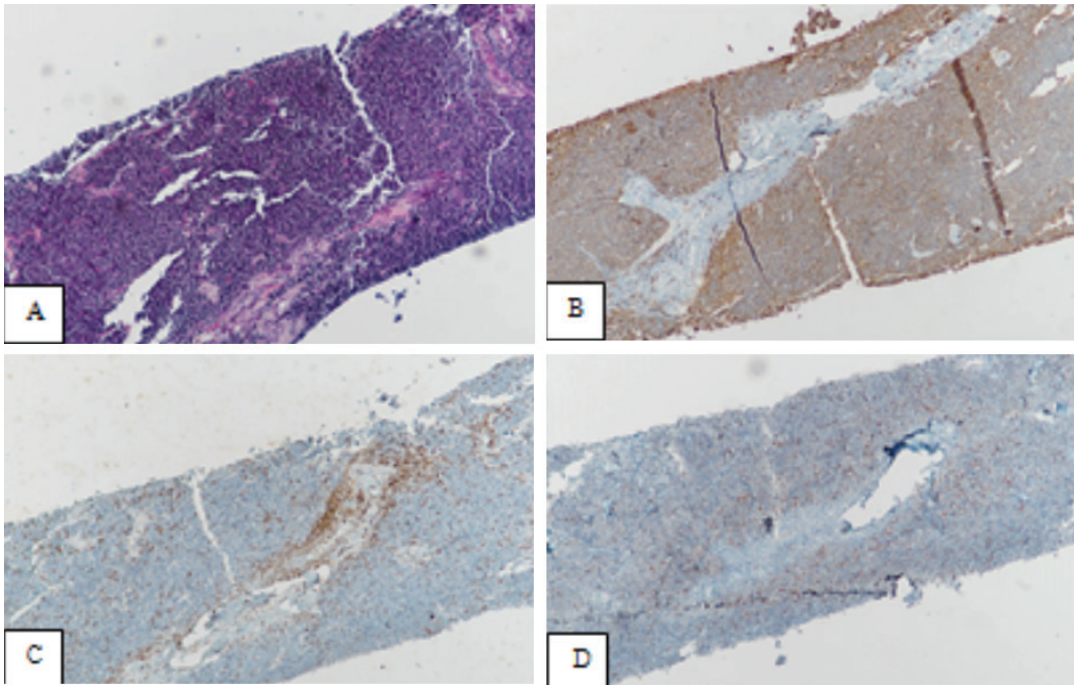

Figure 3. Histologic (A) and immunophenotypic (B-D) features of the lesion. The neoplastic cells are small in size, monomorphic in appearance, and are characterized by a slightly irregular nuclei, inconspicuous nucleoli, and narrow cytoplasm (A, 400). The neoplastic cells are positive with *CD-20* with occasional *Bcl-6* and *CD-5* positivity (B-D, 400)

higher attenuation values on NECT and restricted diffusion on MRI (3,11,12). However, it is still challenging to differentiate it from some inflammatory conditions sharing similar features on imaging. The most challenging imaging differential of OL is idiopathic inflammatory orbital pseudotumor (IIOP) which is a non-granulomatous inflammatory disease responding well to steroids. Among its five forms including anterior, apical, diffuse and sclerosing, myositic, and lacrimal types, the most common is the myositic form characterized by uniform enlargement of one or more extraocular muscles and their tendons. Since our patient did not have extraocular muscle enlargement, this was not considered in the differential diagnosis, like thyroid ophtalmopathy. The rare lacrimal sub-type was considered; however, orbital pseudotumor is characterized by a relatively acute onset of pain, which was not present in our patient. Although DWI was not included in the MRI of our patient, the presence of restricted diffusionl if demonstrated, is a feature that differentiates OL from pseudotumor. *IgG4*-related orbital disease is a diffuse or mass forming fibroinflammatory reaction rich in *IgG4*-positive plasma cells which may also involve the orbit and may not be differentiated from IIOP on imaging. It is characterized by an elevated serum titer of *IgG4* (13). Moreover, sarcoidosis and Wegener granulomatosis may also cause diffuse infiltration of orbital structures without any specific imaging features; however, there were no other associated signs of these diseases in our patient. The presentation of our patient with a frontotemporal region mass in which intraorbital extension was detected by imaging studies was quite unusual, and biopsy was required for the exact diagnosis which was shown as a non-OAML primary OL by immunohistochemical examination. In the treatment of non-MALT-lymphoma, as also in OAML a combination of two or more of surgical resection, radiotherapy, and chemotherapy are used. However, prognosis is less favorable than that of OAML.

CONCLUSION

Lymphomas are the most common primary orbital tumors in patients >60 years of age. Accurate diffferentiation of OL from benign lymphoproliferative disorders is crucial due to differences in their management. Imaging characteristics and clinical findings should be carefully evaluated in the differential diagnosis, and OL should be considered in cases of a slowly growing periorbital/orbital region mass in adults.

Ethics

Informed Consent: Informed consent was taken from the patient.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: B.E., E.K., H.Ö., Design: B.E., T.K., Data Collection or Processing: B.E., T.K., N.K., İ.M., Analysis or Interpretation: B.E., H.Ö., Literature Search: B.E., E.K., N.K., Writing: B.E., İ.M.

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