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Pathology

MARGINAL ZONE B CELL LYMPHOMA OF EYELID: A RARE CASE REPORT

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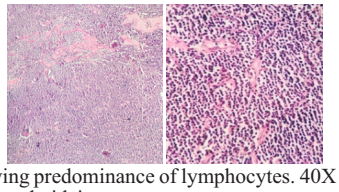
ABSTRACT Lymphomas are rare tumors of orbit representing malignant end of spectrum of lymphoproliferative orbital lesion. We report a case of 72-year-old female with chief complaints of ptosis, edema, mass on right upper eyelid. There was no diminution of vision but field of vision of right eye was restricted. Clinical examination and imaging lead to biopsy which revealed organized lymphoid tissue composed mainly of B cells. IHC was done and it was suggestive of MALT lymphoma.

KEYWORDS :

Introduction:
Lymphoma is the most common malignancy arising in the ocular adnexa, which includes conjunctiva, lacrimal gland, lacrimal sac, eyelids, orbit soft tissue and extraocular muscles. Ocular adnexal malignant lymphoma(OAML) accounts for 1-2% of non Hodgkin lymphoma and 5-15% of extranodal lymphoma.[1] Among various lymphoma variants that could arise in the ocular adnexa ,marginal zone B cell lymphoma(OA-MZL)is the most common one [2][3][4]. Other types of lymphoma arise in these anatomical sites; follicular lymphoma is the second most frequent histology, followed by diffuse large B cell lymphoma and mantle cell lymphoma.[5,6] MALT lymphoma originate from mature B cells in the marginal zone of MALT, is most prominent in gastric region (50%) but can also involve ocular adnexa referred as ocular adnexal marginal zone B cell lymphoma(OAML). Individuals in the fifth to seventh decade are mostly affected and a slight female preponderance is seen. Palpable mass, painless proptosis, diplopia are the common presenting features while the vision is rarely affected.

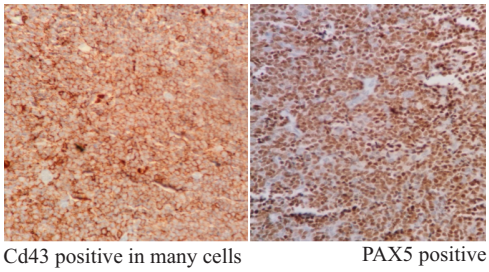
Case History:
A 73-year-old female presented in outpatient department with a painless swelling on right upper eyelid for one month, which was gradually increasing in size. On local examination it was solitary, firm to rubbery in consistency, non-tender and skin overlying was intact. There was no diminution of vision. The routine hematological parameters were normal. Biopsy was performed from the lesion and further IHC was done on the paraffin embedded tissue blocks.

Microscopic examination:
Biopsy showed tumor cells infiltrating into tissue of eyelid. The neoplastic cells were small to medium in size, slightly irregular nuclei with moderately dispersed chromatin and inconspicuous nucleoli, and moderate to abundant pale staining cytoplasm. Some tumor cells showed plasmacytic differentiation and few were small lymphocytes. Few large cells resembling centroblast or immunoblast were also present. These features favored possibility of lymphoid neoplasm. IHC was given.

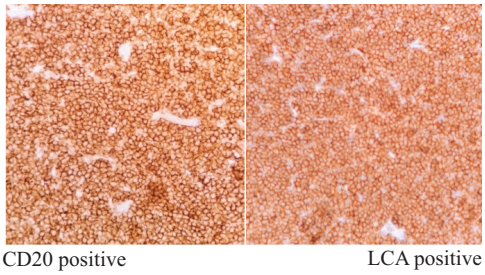


10X view showing predominance of lymphocytes. 40X view showing aggregates of lymphoid tissue

Immunohistochemistry results:
IHC was performed on the paraffin embedded blocks which showed LCA positive, CD20 positive, PAX 5 positive, and CD43 positive in many cells while negative markers were CD5 , CD23, CD3, Light chain kappa and lambda. Hence IHC was confirmatory of Non Hodgkin Lymphoma(NHL), B cell type, MALT lymphoma.



Cd43 positive in many cells PAX5 positive



CD20 positive LCA positive

Discussion:
Lymphomas can infiltrate any orbital and ocular adnexal tissue. The clinical picture of OAML depends on the tissue compromised. It is difficult to differentiate clinically OAML from other orbital diseases due to lack of specific signs or symptoms. Every histological type of malignant lymphoma can arise in the ocular adnexa with similar presenting symptoms and requires surgical biopsy for histopathological diagnosis; considering that treatment and prognosis remarkably vary among different lymphoma categories. Clinical manifestations usually consist of a slowly growing, painless mass that displaces the normal structures. Imaging techniques are supportive for distinguishing OAML from other orbital masses and for accurate staging.

Ocular adnexal MALT lymphoma displays the well known classical histopathology and immunophenotype profile of most MALT lymphomas. The characteristic marginal zone B cells have small to medium sized, slightly irregular nuclei with moderately dispersed chromatin and inconspicuous nucleoli, resembling centrocytes, and moderate to abundant pale cytoplasm. Alternatively, the marginal zone cells resemble small lymphocytes. Plasmacytic differentiation may be present. Small number of tumor cells resembling centroblast or immunoblast are usually present.[10]

The lymphoma cells infiltrate around reactive B cells follicles external to preserved mantle, in marginal zone distribution; and spread out to form larger confluent areas that eventually replaces most of follicles, leaving small remnants of germinal centers. The lymphoma cells may specifically colonize reactive germinal centers can lead to close resemblance to follicular lymphoma. Aggregates of lymphoid cells with distortion of epithelium may be seen around glandular structures of ocular adnexa.[10]

The neoplastic cells of MALT lymphoma are Cd20+, CD79a+, CD5-, CD23-, CD10-, BCL2-, BCL6-, cyclin D1-, CD43+/- and CD11c+/-. The tumor cells of MALT lymphoma typically express IgM heavy chain and less often IgA or IgG.

Differential diagnosis of OAML includes benign lymphoproliferative disorders such as lymphoid hyperplasia, pseudolymphoma and inflammatory pseudotumor which do not show light chain restriction. The other lymphomas in differential diagnosis are mantle cell lymphoma (Cd5+, CD23-, cyclin D1+), small lymphocytic B cell lymphoma (CD5+, CD23+) and follicular lymphoma (CD10+, BCL6+).

OAML shows a better prognosis in comparison to other lymphomas arising in the ocular adnexa. Most OAML patients displays good prognostic indicators like prolonged disease-free interval after local treatment. Involvement of other extranodal site or even bone marrow does not confer a worse prognosis.[10]

Conclusion:

OAML is a relatively rare tumor. Diagnosis is difficult because of its slow growth and many differential diagnoses which mimic OAML on histology. OAML displays some histopathologic and immunohistological peculiarities with respect to other MALT lymphomas, mostly regarding marked plasmacytic differentiation and altered expression of molecules regulating the cell cycle and apoptosis. Confirmatory diagnosis can be made on IHC. Recognition of this rare entity is important for making accurate diagnosis, proper treatment and better prognosis than other lymphoma categories.

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