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Case Report

A large primary orbital lymphoma with proptosis: A case report and review

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Abstract

Background: Primary orbital lymphomas are a rare subset of tumors constituting 1–2% of non-Hodgkin's lymphoma. They are mostly indolent B-cell lymphomas presenting with gradual progressive proptosis, decreased visual acuity, restricted ocular mobility, and diplopia. The role of surgery is mainly for obtaining a biopsy. Most of these tumors require multimodality treatment including chemotherapy, radiation, or both, which have major role.

Case Description: We report one such case of marginal zone lymphoma of the orbit in a female with significant proptosis who was treated with multimodality treatment, including surgical excision as a major treatment modality. Decompression of symptomatic proptosis was followed by chemotherapy and radiation.

Conclusion: Primary orbital lymphoma is a rare clinical entity with diverse clinical outcomes. It can be successfully managed with surgical excision for decompression of mechanical proptosis followed by chemotherapy, radiation, or both.

Key Words: Marginal zone lymphoma, orbital lymphoma, proptosis

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INTRODUCTION

Primary orbital lymphomas are a rare subset of non-Hodgkin's lymphomas. They are mostly indolent, slow-growing B cell lymphomas, usually presenting with progressive proptosis. The role of surgery is mainly for obtaining a biopsy, and the management of such tumours involves multimodality treatment. Here, we report the case of a marginal zone lymphoma of the orbit that was treated with surgical excision as a major treatment modality. Decompression of symptomatic proptosis was followed by chemotherapy and radiation.

CASE REPORT

A 47-year-old lady presented with a history of gradual progressive bulging of the right eye for 5 years. The proptosis was significant and restricting the right eye movement laterally [Figure 1]. It was painless but associated with visual blurring in the right eye. There was no history of any thyroid disorder or trauma to the right eye. On physical examination, the right eyeball was deviated downward and medially with significant proptosis and restricted right lateral eye movements. The proptosis was nonpulsatile without ocular bruit. Vision was 1/60 in the right eye and 6/6 in the left eye. There

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was no organomegaly or lymph node enlargement. Rest of the cranial nerves and neurological assessment was normal. Magnetic resonance imaging (MRI) of the orbit [Figures 2 and 3] revealed an intraconal 3 × 3 cm lesion, which was hypointense on T2 and uniformly enhancing on contrast. It showed significant diffusion restriction and was suggestive of lymphoma. The patient was taken up for surgery with a right frontal craniotomy and superior orbitotomy [Figure 4a] with removal of superior orbital rim, roof, and superior wall of the orbit. The tumor was exposed from the posterior aspect after complete exposure with meticulous dissection and separation of the periorbita [Figure 4b and c]. Tumor was firm, fibrous, and relatively less vascular with no distinct plane between the tumor and periorbital fat. There was no infiltration of the dura or extraocular muscles. Intraoperatively, frozen section suggested lymphoproliferative lesion. Gross total excision of the tumor was done with cranioplasty and reconstruction of the orbital rim with restoration of orbital contour. Postoperatively, there was significant reduction of proptosis with preserved right eye vision and no neurological worsening. A computed tomography (CT) scan of the brain done in the postoperative period showed adequate tumor clearance. Histopathology with immunohistochemistry reported an extranodal marginal zone lymphoma [Figure 5] with dominance of CD20/CD79a/Pax5, expressing B lymphoid cells with substantial populations of peripherally differentiating plasma cells. Ki67 index was 5%. The patient further received chemotherapy consisting of cyclophosphamide, Adriamycin, and vincristine (CHOP), and was planned for radiotherapy at a later date.

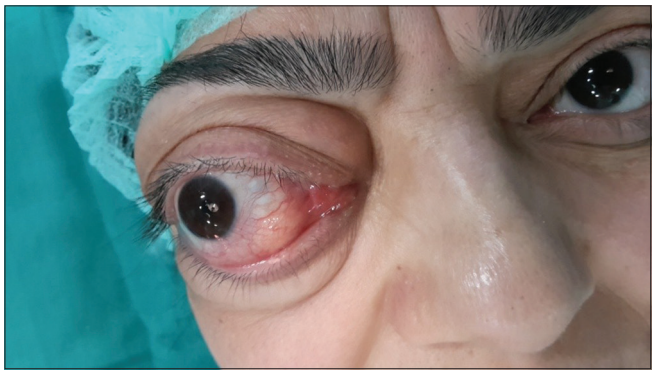

Figure 1: Clinical picture showing the extent of proptosis in the right eye

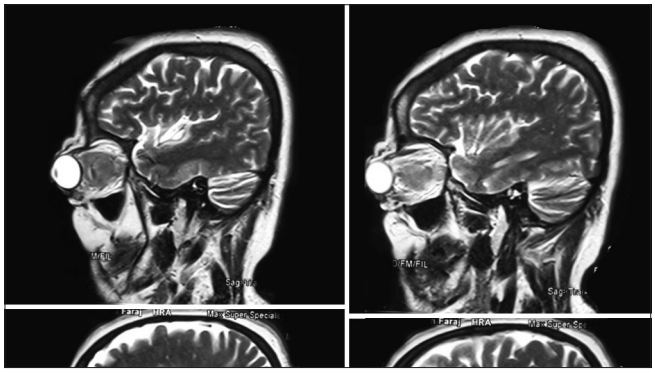

Figure 2: T2-weighted sagittal and coronal MRI images showing hypointense and intraorbital lesion

DISCUSSION

Orbital lymphoma is reported as the most common malignant tumor of ocular adnexa constituting 55% of all orbital tumors.[4] The occurrence of primary orbital lymphoma, on the other hand, is exceedingly rare and comprises approximately 1% of non-Hodgkin's lymphoma and 8% of extranodal lymphoma.[7] Such patients usually do not give any history of prior lymphoma and do not show any manifestation of systemic lymphoma. Majority of them are B-cell lymphomas, and marginal zone lymphomas

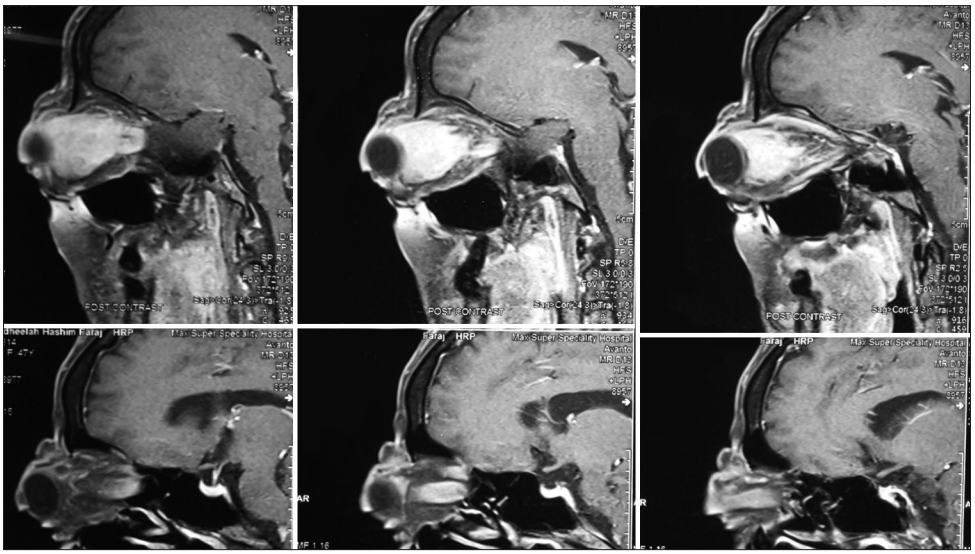

Figure 3: Contrast T1 images showing homogenously enhancing mass lesion 3 × 3 cm in the right orbit in the intraconal location

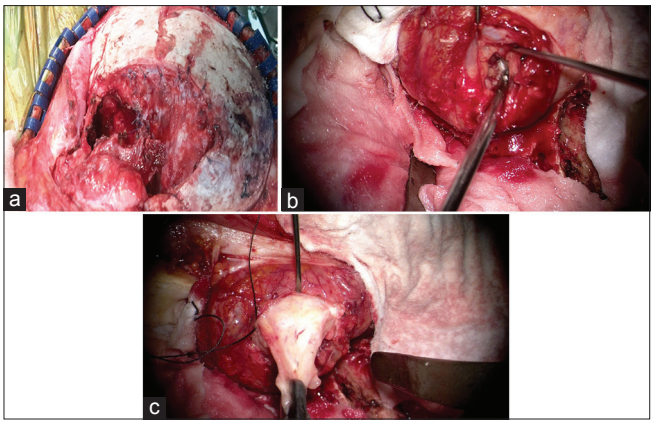

Figure 4: (a) Intraoperative image after right superior orbitotomy and removal of orbital rim (b) Approach to the tumor (periorbita exposed) (c) Intraoperative image of tumor dissection from the periorbita

represent the most common subtype.[7] The other histopathological subtypes include follicular lymphoma, diffuse large B-cell lymphoma, mantle cell lymphoma, small lymphocytic lymphoma, and lymphoplasmacytic lymphoma. Orbital lymphomas usually present in the age group of 50–70 years, with a slight female preponderance.[2] In our case, the patient was a 47-year-old female. Most tumors present as slowly growing, painless, orbital masses with increasing proptosis and follow a largely indolent course.[2] Progressive proptosis, decreased visual acuity, diplopia, and restricted ocular mobility are the usual presenting features.[2,5] Majority of the tumors are unilateral at presentation; however, bilateral involvement has been reported in 5% of the cases.[5] Ninety percent of the patients present with localized disease. The most frequent tumor location is the superior lateral quadrant, and the superior rectus is most commonly involved extraocular muscle.[5] In our patient, there was no involvement of any extraocular muscle. The patient had a long-standing history of proptosis over as long as 5 years. An interval MRI scan of 1 year did not show significant increase in size, but radiology was highly suggestive of lymphoma. A differential diagnosis of an orbital neurofibroma was also considered as per the clinical presentation and history. As there was diagnostic uncertainty, surgery was undertaken as a tool for establishing a diagnosis with histopathological confirmation, as well as for symptomatic decompression of proptosis, which was causing facial disfigurement. Cases with rapidly increasing painful masses with visual loss have also been reported.[6] This is seen in diffuse large B-cell lymphomas. Spontaneous regression of tumors has also been reported.[3]

Cases reported tumors with high vascularity where either wedge biopsy or total tumor clearance was done.[1,6] This is in striking contrast to our case where the tumor was relatively firm and avascular and did not pose a significant challenge for hemostasis.

Multimodality treatment including surgery, chemotherapy, and radiation or a combination have been advocated in the

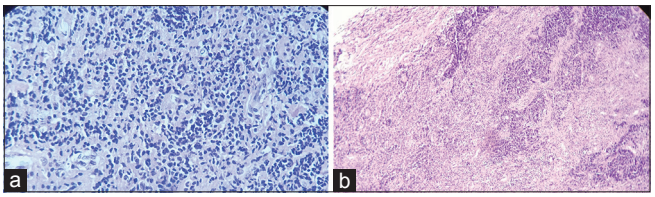

Figure 5: (a and b) H and E staining of the histopathological specimen showing typical lymphoplasmacytic cells, suggesting extranodal marginal zone lymphoma

management of orbital lymphomas. Staging of tumor is an important guide to selection of an appropriate treatment strategy. Surgery alone; however, is not recommended due to the high rate of local relapse. Surgery is usually utilized as a diagnostic tool. Complete remission without significant complications has been reported with radiation alone with a median dose of 3,060 cGy (centigrays).[8] Radiotherapy alone has been reported to provide excellent local control and survival in patients having localized mucosa-associated lymphoid tissue lymphoma.[8]

CONCLUSION

To conclude, primary orbital lymphoma is a rare clinical entity with diverse clinical course. It can be successfully treated with multimodality treatment after appropriate staging.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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