Reactive Lymphoid Hyperplasia – A Case Report

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We are reporting an unusual case of reactive lymphoid hyperplasia of choroid. The ancillary laboratory parameters, intraocular fluid analysis and whole body imaging scans ruled out any infectious etiology or any extraocular foci of primary malignancy. Due to no useful vision in the affected eye and increasing size of the lesion, the eye had to be enucleated. Histopathology, immunophenotyping and molecular study of the enucleated globe confirmed the diagnosis of reactive lymphoid hyperplasia of choroid.

Abstract

Reactive lymphoid hyperplasia of choroid (RLH) is an uncommon choroidal disease and is a diagnosis of exclusion. The most commonly affected site of RLH in ocular tissue is the conjunctiva and orbit. Till now, there has been no published report of RLH of uveal tissue from India. The lesions frequently present a diagnostic challenge, due to considerable overlap of the processes between a malignant and reactive condition.

Introduction

Imaging studies such as ultrasound scan, computerized tomography and magnetic resonance imaging will help in ruling out a metastatic lesion and in making the diagnosis. However, accurate diagnosis is possible only by performing the choroidal biopsy. Early diagnosis is essential as prompt treatment may help patients to regain useful vision.

Case History

A 50 year old Asian-Indian female from Chennai complained of gradual and painless diminution of vision in the right eye for last one month. The patient was a known case of diabetes and hypertension. Best corrected visual acuity of right eye was hand movement and <N36 while of left eye, it was 6/6 and N6. Intraocular pressure was 10 mmHg in both eyes with gonioscopy showing open angles. There was no proptosis noted with no restriction in extraocular movements. The right eye anterior chamber was quiet with anterior vitreous cells 1+. The fundus showed exudative retinal detachment with a flat sub-retinal amelanotic lesion situated temporal to the optic disc. In the left eye, the anterior and posterior segment findings were within normal limits. Ultrasonography scan of the right eye showed total retinal detachment, shifting fluid sign and a diffuse retinochoroidal mass lesion in the inferotemporal quadrant abutting the optic nerve with a maximum thickening of 4.1 mm, sub-tenon space widening and with peripapillary choroidal thickness 2.6 mm (Figure 1). Magnetic resonance imaging of brain and orbit showed an ill-defined soft tissue lesion in the inferotemporal quadrant of the right eye with scleral and optic nerve head involvement and extracranial extension. The findings suggested either an inflammatory or a metastatic pathology. The routine blood test with complete and differential leucocyte count was normal. The patient was investigated for Quantiferon-TB gold test, RPR, TPHA, HIV, hepatitis B and C all of which came out to be negative. Anterior chamber tap of the right eye for PCR and RT-PCR for tuberculosis was also negative. A 64-slice whole body PET scan ruled out any foci of systemic malignancy. Fine needle aspiration biopsy of the lesion showed numerous atypical cells along with numerous RBCs and scattered mixed inflammatory cells. Repeat B-scan of the right eye done after 20 days showed the lesion to be increasing in size. As by that time, the right eye had no perception of light, with the patient’s consent, enucleation of the right eye was done.

Histopathology Report of Enucleated Globe

Light microscopic examination of the specimen revealed dense collections of lymphoplasmocytic cells in uveal tissue starting from the choroid. Plasma cells showed Russell bodies. The choroid was seen to have extensive fibrosis and thickening with multiple focal collection of reactive lymphocyte follicles with germinal centres (Figure 2 a-b). Immunohistochemical staining with B-lymphocyte (anti-CD-20) and T-lymphocyte (anti-CD-3) markers showed heavy staining of the cells (Figure 2 c-d). Retinal detachment with sub-retinal fluid containing scattered macrophages and lymphocytes was seen. Sclera and adjacent orbital tissue showed thickening and extensive fibrosis with multiple lymphoid follicles. Focal collections of lymphocytes were also seen in extracranial muscle fibres. The cornea, iris and ciliary body, however appeared to normal. The light microscopic and immunohistochemical findings were...
consistent with a benign reactive process, and a diagnosis of reactive lymphoid hyperplasia of the choroid was made. Post-enucleation, the right socket was found to be healthy and implantation was done. The patient was found to be stable at 3 and 6 months follow-up.

**Case Report**

**50**

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immunohistochemical stain with CD3 (C) and CD20 (D) (magnification- 40X). (magnification- A: 10X ; B:400X).

**Figure 2:** (a,b): Histopathological morphology of choroidal lesion showing diffuse infiltration with lymphocytes under Haematoxylin and Eosin stain (magnification- A: 10X ; B:400X). (c,d): Choroidal biopsy lesion under immunohistochemical stain with CD3 (C) and CD20 (D) (magnification- 40X).

**Discussion**

RLH of choroid consists of a low-grade proliferation of lymphocytes and histiocytes that arises in and primarily involves the uveal tract. The disease was first described by Gass in 1967 and was called as “inflammatory pseudotumor of the uveal tract” in his article. Crookes and Mullaney recognized the low-grade nature of the proliferation. However, Ryan and co-workers reviewed 21 enucleated eyes from 19 patients and used the term “reactive lymphoid hyperplasia” for the lesions, a name that has persisted in the literature. RLH usually affects middle-aged patients, with a mean age of 55 years and is usually unilateral with no apparent racial or sexual predisposition. Patients complain of recurrent episodes of blurred vision and metamorphopsia secondary to serous fluid detachment of the macula. In cases with longer disease course, symptoms of secondary angle-closure glaucoma may happen due to the displacement of the iris and lens plane. The fundus may show subretinal solitary or multiple creamy yellow-orange lesions due to infiltration of choroid. Episceral extension of the lesion is a common and important finding. Ultrasonographic findings are especially helpful in the diagnosis of RLH in that they reveal diffuse choroidal thickening with low internal reflectivity, a lack of choroidal/scleral excavation, and the presence of extrascleral peripapillary nodules. Choroid biopsy helps to make accurate diagnosis. Histopathological characteristics of RLH is described as marked infiltration by plasma cells, plasmacytoid cells, lymphocytes and eosinophils. Lymphocytes are often seen to be well differentiated without having mitotic figures. Flow cytometry to check polyclonality of lymphocytes may be helpful to exclude lymphoma and confirm the diagnosis. A unicentric, polyclonal lymphoproliferation favors a diagnosis of RLH as compared with multicentricity and monoclonality, which are hallmarks of lymphomas. RLH usually responds to steroid therapy promptly. However, in recalcitrant cases, radiation therapy may be necessary. The response to radiation therapy has been seen to be dramatic and the regression of choroidal mass is found to be significant and rapid. Before the diagnosis of RLH, several diseases should be considered including diffuse choroidal melanoma, metastatic carcinoma, inflammatory diseases such as multifocal choroiditis, uveal effusion, and malignant lymphoma. The advent of ultrasonography and computed tomography in combination with refinement of the clinicopathologic definition has assisted in recognition and avoidance of enucleation. The diagnosis is not easy but important for making the correct treatment plan. Misdiagnosis may lead to unnecessary enucleations leading to severe legal problem.

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