Massive Retinal Gliosis: A Rare Entity

Massive retinal gliosis (MRG) is a benign disorder in which there is proliferation of glial cells. MRG may be associated with chronic inflammation, vascular disorders, trauma, glaucoma and congenital abnormalities. Here we describe a case of MRG in a 23 year old female. She presented with congenital microphthalmia of the left eye with loss of vision. Due to cosmetic reasons she underwent left enucleation. The specimen was received at our department for Histopathological evaluation. Upon histological examination the eye showed massive retinal gliosis and strong positivity for GFAP (Glial fibrillary acidic protein) protein on Immunohistochemistry application.

Key Words: Massive retinal gliosis, Congenital microphthalmia.

Introduction

Massive Retinal Gliosis can be defined as that benign proliferation of well-differentiated glial cells. In 1926 the term massive retinal gliosis was used by Friedenwald. In 1971 Yanoff defined three criteria for massive gliosis of the retina: \(^1\) segmental or total replacement of the retina by glial tissue; \(^2\) abnormal blood vessels within the glial mass; and \(^3\) obliteration of the normal retinal architecture by the proliferating glial tissue. Yanoff concluded that this lesion was a non-neoplastic tissue response to retinal injury. Muller cells are the cell of origin of MRG. MRG has been reported to be associated with retinopathy of prematurity and one case of bilateral gonorrheal ophthalmia. Histologically MRG is composed of proliferation of benign glial cells filling up the vitreous cavity.

We report a case of massive retinal gliosis in a microphthalmic eye removed for cosmetic reasons.

Case Report

A 23 year old female had a small left eye since birth. Initially she had some vision in this eye but as she grew up she became totally blind. There were no other positive clinical or ophthalmologic findings except for a left microphthalmic eye. Being a female the patient opted for artificial eye implant for cosmetic reasons. The left eye was enucleated and the specimen was sent to Armed forces Institute of Pathology (AFIP) Rawalpindi for Histopathological examination. On gross examination, the eye had anteroposterior diameter of 2.5 cm and horizontal diameter of 1.3 cm. The cornea was hazy and opaque. The cut surface of the eye showed haphazard and fragmented vitreous cavity with loss of architecture. The specimen was embedded in paraffin and 5 um sections were stained with Haematoxylin and eosin. No gross picture was taken. Histologically the sections revealed disorganized internal architecture and replacement of the retina with intervening bundles of spindle to oval shaped cells filling up the vitreous.( Figure I)

![Image](https://via.placeholder.com/150)

*Figure I: Inset A: Interweaving glial cells giving a eosinophilic appearance at low power due to the abundant fibrillary cytoplasm of glial cells. (H-E stain x 40)*

*Inset B: High power photomicrograph of bland appearing glial cells and few blood vessels. (H-E x 200)*
Individual cell had fibrillary cytoplasm and indistinct cell borders. Few thick walled hyalinized blood vessels were evident. No necrosis or mitosis was seen. No evidence of malignancy was found in the material examined. Immunohistochemistry was applied for determining the glial origin of the spindly cell. GFAP (Glial fibrillary acidic protein) was positive in the spindly glial cells. (Figure II) The morphological and immunohistochemical findings were consistent with Massive retinal gliosis.

Discussion

Massive retinal gliosis is a non-neoplastic proliferation of the glial cells. Both genders and all ages may be affected. MRG appears clinically as a single or multiple nodules, mostly in the peripheral retina but can occur anywhere. MRG can sometime mimic an intraocular tumor. In this case there was no such clinical suspicion and the eye was removed for cosmetic reasons. Newsome et al used phase contrast and electron microscope for investigating the cellular constituents in MRG. They found four major constituents Macrophages, pigmented epithelioid cells, glial cells, and fibroblastic cells. MRG is the retinal response to chronic injury. In our case the patient has congenital microphthalmia and progressively decreasing vision. The differential diagnosis of such an intraocular lesion includes uveal melanoma, astrocytic hamartoma, retinal hemangioblastomas, tumors of the retinal pigment epithelium, intraocular metastasis, and vasoproliferative tumors of the retina (VPTR). The distinction from VPTR is often difficult if the vascular proliferation is minimal or obscured by the glial component. MRG, VPTR and retinal hemangiomas may be in fact different phenotypes of the same spectrum. If the distinction between these entities is not possible than the term reactionary retinal gliangiosis may be used. Histologically Massive retinal gliosis is typically characterized by a nodular proliferation of spindle shaped cells have fibrillary cytoplasm. MRG involves the posterior pole and fills the vitreous cavity. The spindle cells have minimal atypia and their glial origin can be determined by application of immunohistochemistry for GFAP. VPTR and presumed acquired retinal hemangiomas usually have small lesions, located peripherally, anterior to the equator. The anatomical extent, morphological features and immunohistochemical findings of our case were that of MRG.

References