Massive retinal gliosis: An unusual case with immunohistochemical study

Sanjay D Deshmukh, Amrut V Ashturkar, Shridhar V Babanagare, Suvarna K Gokhale[1], Anand A Deshpande[1]

Massive retinal gliosis (MRG) is a rare, benign intraocular condition that results from the proliferation of well-differentiated glial cells. Immunohistochemically, these cells show positivity for glial fibrillary acid protein (GFAP), neuron specific enolase (NSE), and S-100 protein. We encountered a case of a 45-year-old female with loss of vision in the left eye. She had a history of trauma to that eye two years ago. Enucleation was carried out, because malignancy was suspected due to retinal calcification. On the basis of light microscopy and immunohistochemistry (IHC) performed on the enucleated eye, it was diagnosed as massive retinal gliosis.

Key words: Enucleation, immunohistochemistry, massive retinal gliosis

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Departments of Pathology and [1]Ophthalmology, Shrimati Kashibai Navale Medical College and General Hospital, Narhe, Pune-41, India

Correspondence to: Dr. Sanjay Deshmukh, 5- Gurudeep-2 Society, 62, Sahakarnagar, No. 2, Pune - 411 009, India. E-mail: drsanjay123in@yahoo.co.in

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Massive retinal gliosis (MRG) is a rare, benign intraocular condition resulting from the proliferation of well-differentiated glial cells.[1] The term massive retinal gliosis was first used by Friedenwald, in 1926.[2] It is a non-neoplastic tissue response to retinal injury that may develop in association with congenital malformations, trauma, glaucoma, intraocular neoplasms, vascular disorders, and chronic inflammatory conditions resulting in atrophic phthisis bulbi.[3] It may also occur due to an old retinal detachment, retinopathy of prematurity or as a complication of retinal detachment surgery.[4,5] This condition may be mistaken for intraocular tumors.[4] We are presenting here a rare case of MRG, diagnosed on the basis of routine histopathology and immunohistochemical studies.

Case Report

A 45-year-old female presented with loss of vision in the left eye, since two years. She gave a history of trauma to that eye two years ago. Clinically, the diagnosis was anterior staphyloma in left eye. The right eye was normal. On ophthalmoscopic examination, a large tumor-like mass was seen in the left eye. A computed tomogram (CT) scan of the orbit revealed that the left eyeball was deformed with stretching and thinning of the anterior sclero-uveal coats. Dense calcification was noted within the vitreous of the left eye ball. There was a homogenous haze within the left eye ball, associated with the atrophic optic

nerve [Fig. 1]. The right eyeball appeared normal. Based on these findings and clinical diagnosis of intraocular neoplasm, enucleation of the left eye was carried out.

On gross examination the eyeball was hard, shrunken, and deformed with an opaque cornea. An optic nerve measuring 5 mm length was also identified attached to the eyeball. The cut surface showed a vitreous cavity replaced by a solid, firm, homogenous gray-white mass, measuring 3.0 × 1.8 × 1.5 cm. On light microscopy, the retina was replaced by spindle-shaped glial cells arranged in interlacing bundles and whorls [Fig. 2]. Individual cells were elongated with abundant eosinophilic fibrillary cytoplasm [Fig. 3]. Thin-walled blood vessels with a thick hyaline layer were noted. Foci of calcifications were also identified. On immunohistochemical study, the spindle shaped cells showed intense positivity for glial fibrillary acidic protein (GFAP) [Fig. 3], neuron specific enolase (NSE), and S-100. Therefore, on the basis of light microscopy and immunohistochemistry study findings, a diagnosis of MRG was offered.

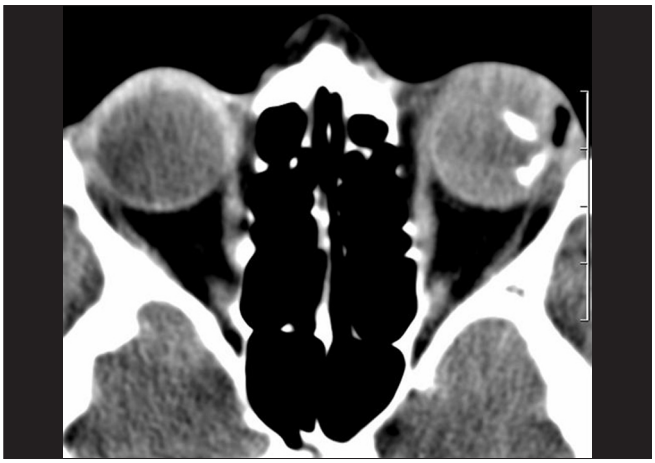

Figure 1: Computed tomogram-scan showing deformed left eyeball with homogenous haze and areas of calcification

Discussion

Massive retinal gliosis represents non-neoplastic proliferation of the retinal glia. We believe this is the first reported case from Indian literature with immunohistochemical confirmation. Regarding histogenesis, the nodule of MRG and the associated pre-retinal glial membrane, result from the proliferation and migration of Müller cells.[1] In MRG, both sexes and all ages may be affected in nearly equal frequency.[1] We encountered massive retinal gliosis in a middle-aged female patient.

Its onset often occurs ten or more years after a predisposing disorder such as chronic inflammation, vascular disorder, glaucoma, trauma, retinal detachment surgery or congenital abnormalities.[4,5] In our case, the patient had trauma to the affected eye two years back, which is a known predisposing factor for the development of massive retinal gliosis.

This lesion appears as a single or multiple, well-vascularized nodule, which has a predilection for the peripheral retina, but may occur anywhere.[6] 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).[7]

Yanoff and co-workers,[3] reported 38 cases of massive gliosis of the retina. The author defined three criteria for massive gliosis of the retina: (a) segmental or total replacement of the retina by glial tissue; (b) abnormal blood vessels within the glial mass; and (c) obliteration of the normal retinal architecture by the proliferating glial tissue. The current case fulfilled the criteria mentioned here; in addition it showed positivity for GFAP, NSE, and S-100 on immunohistochemical marker studies, confirming the glial origin.

This case is differentiated from these alternative diagnoses based on the histopathological features and immunohistochemistry of the lesions. However, the distinction between MRG and VPTR is more difficult, as both entities share similar histological features consisting mainly of glial and vascular proliferations. Even as the glial component

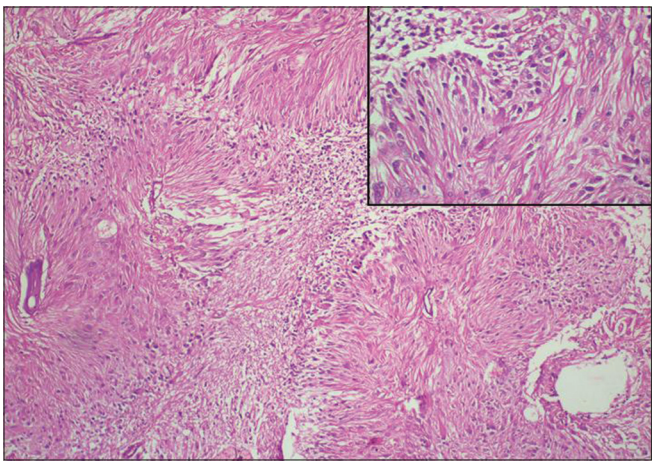

Figure 2: The retina is replaced by spindle-shaped glial cells arranged in interlacing bundles and whorls (H and E, ×100). Individual cells are elongated, with abundant eosinophilic fibrillary cytoplasm (Inset) (H and E, ×400)

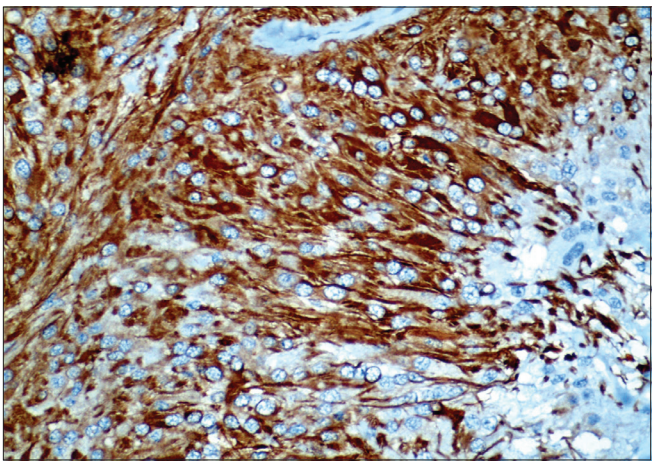

Figure 3: On an immunohistochemical study the spindle-shaped cells show intense immunoreactivity for glial fibrillary acidic protein (IHC)

predominates in MRG, in VPTR both the components are largely represented. This may explain the exudative features of VPTR, which are not usually seen in MRG.[8]

It must be mentioned that massive retinal gliosis is a very rare condition. The distinction of massive retinal gliosis from other intraocular neoplasms is clinically very difficult, so the eyeball may need to be enucleated to potentially save the patient's life; however, light microscopy and immunohistochemistry is mandatory for the final diagnosis. The clinicians should keep this in mind when they come across retinal calcification, especially in an adult patient. Trauma is a significant cause of blindness worldwide.[9] MRG is another manner in which trauma can cause loss of vision, long after the instigating event.

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