CHOROIDAL LEIOMYOMA: A CASE REPORT AND DISCUSSION OF ITS HISTOGENESIS

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Abstract - Leiomyoma is a very rare intraocular tumor which can occur in the iris, ciliary body and choroid. We report a case of choroidal leiomyoma in a 16-years old woman whose left eye was enucleated because of clinical suspicioun for melanoma. Using conventional light microscopy, tumor was intrepreted as spindle cell tumor, most probably of neurogenic orgin and after using immunohistochemical studies, smooth muscle origin of tumor was confirmed.

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INTRODUCTION

Leiomyoma, a benign smooth muscle tumor, occurs in various sites, although the commonest sites at which it develops is the subcutaneous soft tissue, the wall of the gastrointestinal tract and the wall of the uterus. Ocular leiomyomas are very rare and may occur in the iris and Ciliary body. Reviewing the literature up to the present, few documented cases of choroidal leiomyoma have been reported (1,2).

Apparently upto 1981, only two case of leiomyomas of choroid had been documented, one by Jakobiec and colloagues (3) and the other by Naumann (4). The next time, in 1997, another case of choroidal leiomyoma was reported by Sikic-J and colleagues (5).

Ocular leiomyomas have female predilection. The tumor tend to affect the ciliary body and anterior choroid, unlike melanoma which favors the posterior choroid (2). It is difficult to differentiate a leiomyoma from other spindle cell neoplasms, especially amelanotic spindle cell nevus and low grade melanoma, without using electron microscopy and immunohistochemical studies. Many cases perviously diagnosed as leiomyomas are probably melanocytic, rather than smooth muscle lesions(2).

In the immunohistochemical staining technique, leiomyomas are positive for muscle specific antigen, smooth muscle actin, and vimentin.

Report of a case

A 16-year old woman presented with a 1-month history of decreased vision and pain in the left eye. On retinoscopy of the left eye, total retinal detachment and grayish white mass behind the lens was detected. Orbital CT scan showed a mass behind the lens and inferior vitreous cavity. Patient's eye was enucleated because of the suspicion of melanoma.

HISTOPATHOLOGICAL FINDINGS

Macroscopic findings

The enucleation specimen was fixed in 10% formalin. The tumor mass was a well circumscribed white and firm tissue located near the ciliary body that projected into the vitreous cavity. Tumor had bean-shaped appearance and was 1 cm in diameter.

Microscopic findings

In the sections from the intraocular neoplasm, the tumor was composed of uniform spindle shaped cells arranged in fascicles and admixed with abundant fibrillary material.

In the histological examination, differential diagnosis of this spindle cell tumor, schwannoma, leiomyoma and less probably an amelanotic spindle cell melanocytic lesion were respectively suggested by conventional light microscopy (Fig. 1.2). Immunohistochemical studies showed positive reaction for monoclonal smooth muscle actin (Fig. 3), scattered weakly positive reaction for S-100 protein and negative reactions for epithelial membrane antigen (EMA), desmin and HMB+45 Because of these findings, smooth muscle origin of the neoplasm was confirmed. Although leiomyoma is a common neoplasm in some parts of the body, intraocular leiomyoma is a rare tumor and specially choroidal leiomyoma is very rare. Most cases of the intraocular leiomyomas occurred in the ciliary body or ciliary body and anterior choroid rather than choroid alone. Reviewing the literature few documented cases of this tumor has been reported (1,2,5).

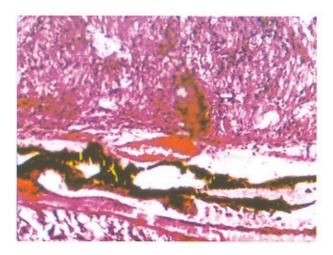


Fig. 1. Detached retina and choroidal tumor near the ciliary body. H and E. stain (x100)

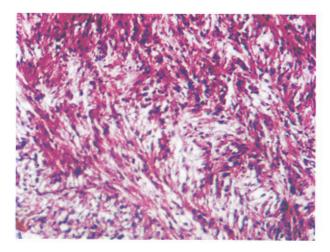


Fig. 2. Leiomyoma of choroid tumor is composed of uniform spindle cells arranged in fascicles and admixed with abundunt fibrillary material. H and E stain, (×100)

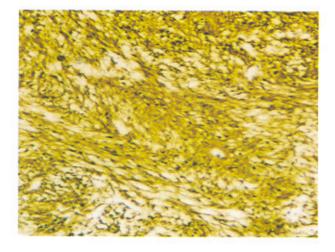


Fig. 3. Immunohistochemical (IHC) stain for monocolonal smooth muscle actin shows positive reaction in cytoplasm of neoplastic cells. IHC stain, (×100)

DISCUSSION

Some hypotheses have been suggested for histogenesis of ocular leiomyomas. Although most investigations suggested vascular origin for these lesions using electron microscopic studies (6) in a few cases of benign ciliary body spindle cell tumor, mesectodermal origin was proposed, as the tumor origin (2).

The cells of the neural crest that contribute to the formation of bone cartilage, connective tissue and smooth muscle in the region of the head and neck, have been called mesectoderm. Embryoligic experiments in chicks have demonstrated that the ocular and orital supporting tissues are of neural crest origin (7-9). The investigators postulated the neural features of some of ocular leiomyoma may be explaind on the basis of the embryologic origin of the smooth muscles of the eyes.

Mesectodermal leiomyma is a rare variant of leiomyoma composed of cells with both myogenic and neurogenic features in light and electron microscopy Most of the reported instances of this tumor microscopically resembles a neurogenic tumor more than a myogenic tumor (1). According to microscopic findings in this unique case we suggest that the neural appearance of this tumor using conventional light microscopy scattered and weakly positive immunoreactivity for S-[00 protein immunohistochemical staining technique, may be a reflection of it's probable origin from mesectodermal smooth muscle of the choroid.

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