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Intraocular Malignant Teratoid Medulloepithelioma of Ciliary Body in an Adult: A Case Presentation

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Abstract

Purpose: To report a case of intraocular malignant teratoid medulloepithelioma, an embryonal tumor with extremely rare presentation in adults.

Method: The case of a 36-year-old male with intraocular malignant teratoid medulloepithelioma, primarily diagnosed as intraocular choroidal melanoma 6 year back, is described and the literature on this subsect is reviewed.

Result: The patient presented with blurring of vision in right eye on both distance and near vision of two-month duration. In association to this he has also a history of itching and pain of the same duration. Six years back he was diagnosed as intra ocular choroidal melanoma with histopathological examination.

Keywords: ***Malignant teratoid Medulloeptheloima; Ciliary body; Pigmented***

1. Introduction

Malignant teratoid medulloepithelioma in the eye is an extremely rare embryonal tumor of neuroepithelial origin. The majority of these tumors originate in the ciliary body epithelium and rarely from iris, retina and the optic nerve [1].

Usually presenting as unilateral tumors, they do not have any predilection for particular, sex, race or laterality. Intraocular medulloepithelioma is a rare and potentially malignant tumor. It primarily occurs in children in the first decade of life and is extremely rare in adults [2].

Histologically, intraocular medulloepithelioma is classified into teratoid and non-teratoid, the latter being the most frequent. They are also classified into benign and malignant [3].

It presents as slow growing and is locally invasive, it is typically characterized by presenting a cyst or mass in iris, ciliary body, or anterior chamber with decreased visual acuity, epiphora, pain, leukocoria, exophthalmos, strabismus, iridis rubeosis, cataract, subluxation of the lens and generally tends to be unilateral, predominantly in the right eye [4].

Zimmerman [1] hypothesized that medulloepithelioma in children is derived from incompletely differentiated embryonic ciliary epithelium (congenital medulloepithelioma). In rare cases in adults, medulloepithelioma is thought to arise from fully differentiated ciliary epithelium after undergoing non neoplastic reactive hyperplasia or pseudoadenomatous hyperplasia. Fuchs [5] believed that adult medulloepitheliomas represent neoplastic transformation of hyperplastic ciliary epithelium, triggered by inflammation or trauma.

Medulloepithelioma diagnosed in adults could represent delayed transformation of a preexistent retinal anlage. Most cases in adults are histopathologically malignant, suggesting a delayed malignant transformation of a preexisting lesion. Benign medulloepithelioma in adults is detected following decades of gradual growth, eventually manifesting clinically. Medulloepithelioma can rarely arise from the optic disc, iris, and retinal stalk [3,4].

2. Case Report

In December 2020, a 30-year-old man presented with 2-month history of blurring of vision on right eye both distance and near vision. In association to this he has a history of Intermittent itching sensation and tenderness. For this complaint he was admitted to the department of ophthalmology. On clinical examination he had hyperemic tarsal conjunctiva with tortuous vessel. On funduscopic examination, he had retinal detachment with secondary angle closure glaucoma to rule out choroidal melanoma. For this the plan was to investigate him with CBC, organ function test, orbital ultrasound and CT scan of the head. Right Orbital ultrasound finding was 17 mm by 17 mm right ocular retinal nodule at the medial retinal surface with exophytic growth to the vitreous having scanty Doppler flow within it. There was vitreous hemorrhage but no retinal detachment.

With this he was diagnosed with Right eye retinal nodule secondary to rule out retinal melanoma and CT scan of head was recommended (FIG.1). CBC and organ function test was normal. But the patient left against medical advice. After four year he returned with the complaint of right eye swelling and pain with total loss of vision of four-month duration. On physical examination there was right globe fumigating mass. For this he was investigated with CBC, organ function test, orbital ultrasound, chest radiography, abdominal ultrasound and brain CT scan including orbital protocol. On CT scan of the head, there was a heterogeneous hyperdense exophytically growing mass in the right posterior globe having post-contrast enhancement. The mass has infiltrated the sclera. There was a biconvex shaped hyperdensity seen in the globe suggesting choroidal detachment. There was an area of soft tissue air foci and pre-septal soft tissue thickening and increased density. The

optic nerve outline was normal. With this finding he was diagnosed with a Right globe mass secondary to likely choroidal melanoma with choroidal detachment and scleral invasion. For this Right eye debulking surgery with post enucleation was done. Tissue sample was sent for pathology. The biopsy result came back reported as a Right eye pigmented malignant teratoid medulloepithelioma.

Microscopic description: Sections show stratified immature neuroepithelial cells forming undulating cord like structures with pseudopapillary, pseudoglandular and fish net like appearance embedded inside a myxoid material frequent multi layered rossette like structures are seen along with frequent mitosis and extensive necrosis. Areas of rhabdoid and adrenal medullary epithelium differentiation are seen. Focal areas of pigmented neoplastic neuroepithelial cells are seen.

The plan was to start the chemotherapy. But the patient left against medical advice again. After two years, he presented with a history of right eye swelling of four-month duration. On physical examination there was right eye fungating mass from the surgical site after removal of the orbital implant. (FIG. 3)

For this, brain CT scan was done, it showed 5.5 cm × 4.5 cm × 3.3 cm relatively well defined heterogeneously hyper dense soft tissue attenuating mass having post contrast heterogeneous enhancement with invasion of optic nerve occupying most of right globe and extension to anterior pre septal soft tissue. The mass had no intracranial extension (FIG. 2).

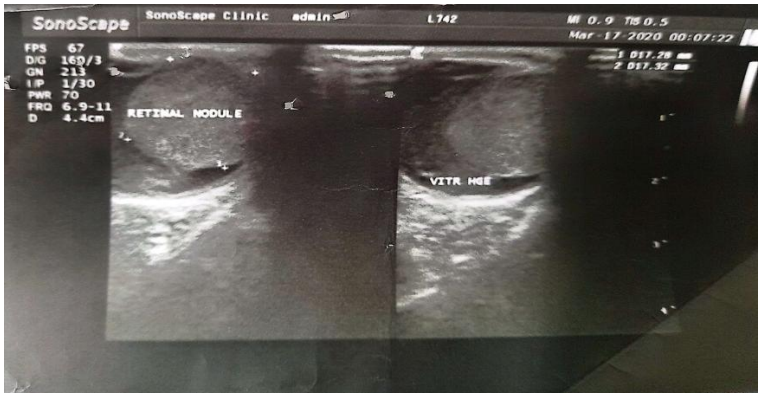

FIG. 1. **Orbital ultrasound, 2020.**

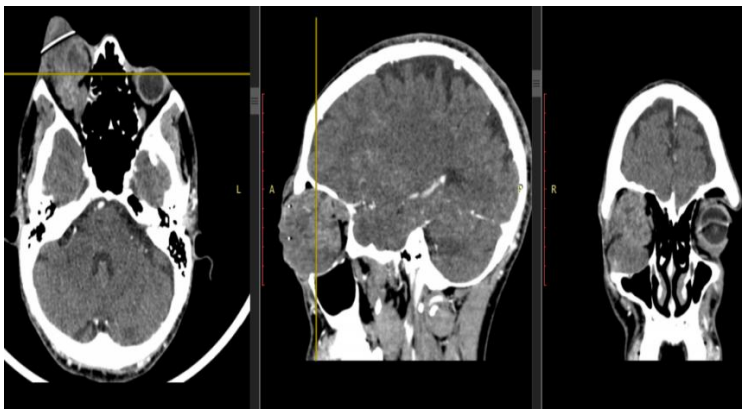

FIG. 2. **Pre and post contrast brain CT scan, 2025.**

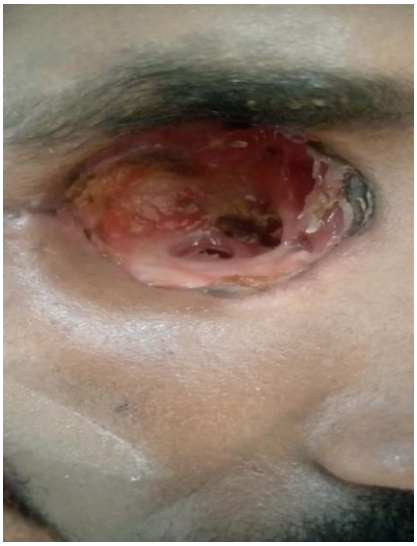

FIG. 3. **Post-encucleation image, 2025.**

3. Discussion

Intraocular medulloepitheliomas are embryonal tumors that are extremely rare in adult. In the radiologic imaging base reported case, the initial diagnosis was intraocular melanoma. Even most of the time the initial histopathologic diagnosis is intraocular teratoma with the unsatisfactory response to chemotherapy led us to revise pathological diagnosis to malignant teratoid medulloepithelioma [6].

The classic features of medulloepithelioma consists of a ciliary body mass with intratumoral cysts. Ultrasound B-scan shows heterogeneous, highly echogenic mass localized to the ciliary body and specifically within the epithelium, demonstrating intratumoral, irregular cystic areas. This tumor may or may not demonstrate calcification, depending if there is cartilage within the tumor, and this further complicates differentiation of intraocular medulloepithelioma from retinoblastoma.

Anterior segment optical coherence tomography (AS-OCT) and ultrasound biomicroscopy (UBM) delineate the exact radial and circumferential extent and the height of the ciliary body mass and demonstrate intratumoral cysts, neoplastic cyclitic membrane, anterior chamber cysts and sequelae, such as lens subluxation. UBM, due to its higher resolution, is a good modality to delineate the tumor and document the dimensions for serial follow-up of the patient.

Fluorescein angiography shows large, branching arteries and veins arising from the ciliary body tumor peripherally and haphazardly crisscrossing within the retro lenticular tissue. One study on fluorescein angiography demonstrated a vascular stalk extending to the tumor from an adjacent detached retina [3].

Magnetic resonance imaging (MRI) is the standard preoperative investigative modality. The tumor appears as a heterogeneous mass composed of both solid and cystic components arising from a lateral retrolental location. The tumor is usually isointense to mildly hyperintense to vitreous on T1-weighted images on MRI, with avid enhancement with contrast. Linear or irregular soft tissue that extends posteriorly along the ciliary body region with abnormal signal intensity of the vitreous body, and the

presence of intratumoral cysts are all features that can be identified at MRI. Scleral invasion, extraocular and optic nerve extension identifiable on MRI can help in the management and in predicting possibility of metastatic disease [3].

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