**Case Report**

# Teratoid Medulloepithelioma -A Rare Ocular Tumor

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## ABSTRACT

A 7 years old child presented with gradually increasing loss of vision along with swelling and redness of his left eye for 2 years. Enucleation revealed a whitish tumor mass in the anterior chamber. Histopathologically, epithelial tubular structures and rosettes were present with lobules of cartilage and glial tissue. medulloepithelioma should be considered in the differential diagnosis of a ciliary body mass especially in a child. (Rawal Med J 2008;33:126-127).

Key Words: Medulloepithelioma, ciliary body, retinoblastoma.

## INTRODUCTION

Medulloepitheliomas of the ciliary body are rare primitive embryonic tumors which arise from the ciliary body epithelium.[1] Those with heterologous elements such as cartilage and neural tissue are called teratoid medulloepitheliomas.[2] These usually occur in the 1st decade of life but may be seen at birth and in adults.[3] We present a case of this rare childhood tumor.

## CASE REPORT

A 5 years old child, resident of Afghanistan presented with gradually increasing loss of vision along with swelling and redness of his left eye for 2 years. On examination, left eye showed protrusion with corneal necrosis. Right eye was normal. Systemic examination did not reveal any abnormalities. A provisional diagnosis of retinoblastoma was made and enucleation was performed. At gross examination, cut surface showed a whitish mass of 1.5x0.8cm size lying

in the anterior chamber (Fig. 1). Microscopic examination revealed a tumor composed of sheets of cells having hyperchromatic nuclei, forming tubular structures and rosettes (Fig. 2).

**Fig 1. Photomicrograph of the gross specimen showing a grey white tumor lying in anterior chamber.**

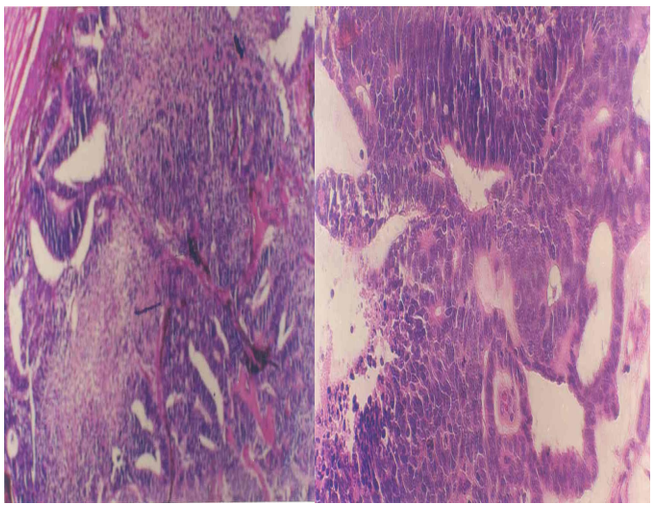
Pigmented cells along with lobules of cartilage and glial tissue were also present within the tumor. Mitotic figures were frequent. The tumor was arising from the ciliary body and involving the anterior surface of lens as well as posterior surfaces of the cornea and conjunctiva. It was not extending outside the eyeball.

**DISCUSSION**

Tumors arising from the ciliary epithelium are the rarest of ocular neoplasms.[4] They contain elements that closely resemble the medullary epithelium and may contain structures resembling those derived from the optic cup, retinal pigment epitheliulm, vitreous and neuroglia. They arise from the non-pigmented epithelial layer of the ciliary body[4] and are said to be counter parts of retinoblastomas. However, in contrast to retinoblastomas, they are never bilateral.[5] They occur in the first decade with an average age of five years. All the reported cases have arisen from ciliary body although iris can also be the site of origin.[3] Some that occur in infancy have a conspicuous network of epithelial bands leading to the name diktyoma given by Fuchs[5] in 1908.

**Fig 2. Photomicrographs showing tumor forming tubules and rosettes with glial tissue**

**(Heamatoxylin and Eosin stain at x 200 magnification).**



Grossly, the tumor develops as a white flat lesion arising from the ciliary body and growing over the iris, posterior surface of cornea and anterior surface of the lens. Since it grows like a membrane, it may occlude the angle and produce glaucoma along with distension of the eye ball.[7] Microscopically, the tumor appears as sheets of nuclei with indistinct cytoplasmic borders resembling embryonic retina. It may also grow as sheets of cells forming rosettes or pseudo- rosettes resembling retinoblastoma.[4] Characteristically diktyomas lack stroma. Various authors have reported focal areas of pigment epithelium along with cartilage and neuroglial tissue. Diktyomas showing elements such as hyaline cartilage, brain tissue or rhabdomyoblasts are called teratoid medulloepitheliomas.[8]

One study on cytogenetic analysis showed primary abnormality of der (16)t (1;16) chromosome, del (6q) and monosomy15.[9] The malignant nature is manifested by adverse histological features, local infiltration and destructive features. Zimmerman and Broughton[9] found histological evidence of malignancy in 66% of cases. Enucleation is the best treatment for advanced cases. Local resection may be attempted for smaller circumscribed lesions; though such treatment is followed by local recurrence.[3] The tumor has a slow growth rate, invades the surrounding structures, may recur locally but rarely metastasizes.

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