

Bilateral medulloepithelioma of the ciliary body: a case report

Ya-Guang Hu¹, Jin-Jin Wang², Xin Ge², Cheng Pei¹, Jian-Min Ma²

¹Department of Ophthalmology, the First Affiliated Hospital of Xi'an Jiaotong University, Xi'an 710061, Shaanxi Province, China

²Beijing Ophthalmology and Visual Sciences Key Laboratory, Beijing Tongren Eye Center, Beijing Tongren Hospital, Capital Medical University, Beijing 100005, China

Correspondence to: Jian-Min Ma. Beijing Ophthalmology and Visual Sciences Key Laboratory, Beijing Tongren Eye Center, Beijing Tongren Hospital, Capital Medical University, Beijing 100005, China. jmma@sina.com; Cheng Pei. Department of Ophthalmology, the First Affiliated Hospital of Xi'an Jiaotong University, Xi'an 710061, Shaanxi Province, China. peich71@163.com

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Dear Editor,

We present an exceptionally rare case about bilateral medulloepithelioma of the ciliary body. This case was approved by Ethics Committee of Capital Medical University. Written informed consent was obtained from the patients. Discovering leukocoria in both eyes and proptosis in right eye for 2mo by parents in a 4.5-month-old boy. He had undergone no treatment elsewhere except magnetic resonance imaging examination before coming to Beijing Tongren Hospital, Capital Medical University on November 2021. His bilateral vision was aberrant, as he was unable to follow the movement of light or an object. Intraocular pressure was measured by digital tonometry and ocular hypertension was found in the right eye. Proptosis, conjunctival hyperemia and slightly corneal edema were seen in the right eye. In both eyes, there was apparent leukocoria and a delayed pupillary light reaction (Figure 1).

Accessory examinations were performed to further explore the disease. Ret-cam fundus examination found leukocoria in pupil and invisible fundus. Ocular ultrasound showed abnormal echo

and retinal detachment in both eyes (Figure 2). Furthermore, orbital computed tomography scan displayed bilateral space-occupying lesion, retinal detachment (Figure 3).

All evidences suggest that it might be retinoblastoma. On October 27th 2021, enucleation and hydroxyapatite implantation was performed in right eye. The volume of right eye was increased, without thickening of optic nerve. At the same time, bone marrow and cerebro-spinal fluid was obtained by puncture, which was used for flow cytometry to assist diagnosis. Flow cytometry examination showed there was no retinoblastoma cell.

Histopathological observation is the gold standard for diagnosis. And it revealed that it was medulloepithelioma of the ciliary body, malignancy, well-differentiated, non-teratoid (Figure 4A, 4B). The tissue of choroid, sclera, optic disc and nerve were not involved. Immunohistochemical result showed neuron-specific enolase (+), synapsin (+), CD56 (+), S-100 (+), chromogranin A (-), glial fibrillary acidic protein (+), vimentin (+), cytokeratin (+), epithelial membrane antigen (-), leukocyte common antigen (-), CD99 (+), Ki-67 (-).

After 7mo of follow up, without no chemotherapy or radiotherapy, orbital computed tomography scan showed the volume of left eye was decreased (Figure 5). On June 24 2022, enucleation and hydroxyapatite implantation was performed in left eye. The result of histopathology examination showed that it was also medulloepithelioma of the ciliary body, malignancy, well-differentiated, non-teratoid (Figure 4C). The tissue of choroid, sclera, optic disc and nerve were not involved. Immunohistochemical result showed neuron-specific enolase (+), synapsin (+), CD56 (+), S-100 (-), chromogranin A (-), glial fibrillary acidic protein (-), vimentin (+), leukocyte common antigen (-), CD99 (-), Ki-67(-). After surgery, the patient did not receive radiation and chemotherapy. And he was asked to follow up by outpatient visit every month.

Medulloepithelioma of ciliary body is a rare intraocular tumor arising from the nonpigmented ciliary epithelium of pars plicata and only nearly 120 cases were reported all over the world^[1-6]. The tumor is believed to be nonhereditary, but it mainly occurs in infant and children, at a median age of 2 to 5y^[1,5,7-11]. Moreover, after reviewed the literatures, we do not find out bilateral medulloepithelioma of ciliary body. In

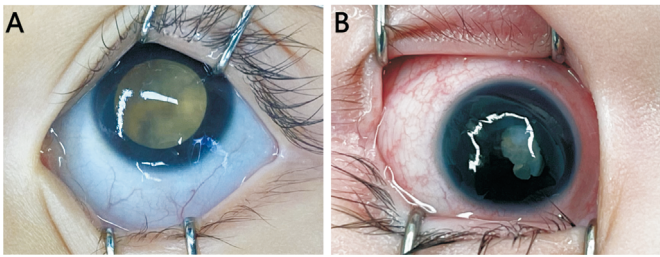


Figure 1 Image of right eye (A) and left eye (B).

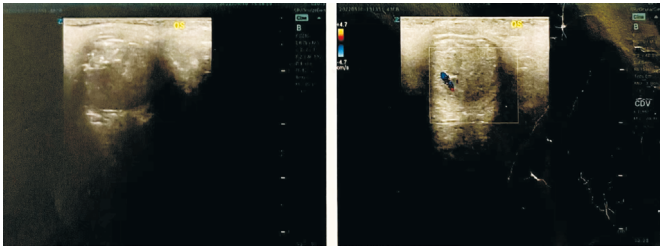


Figure 2 Result of ocular B-ultrasound.

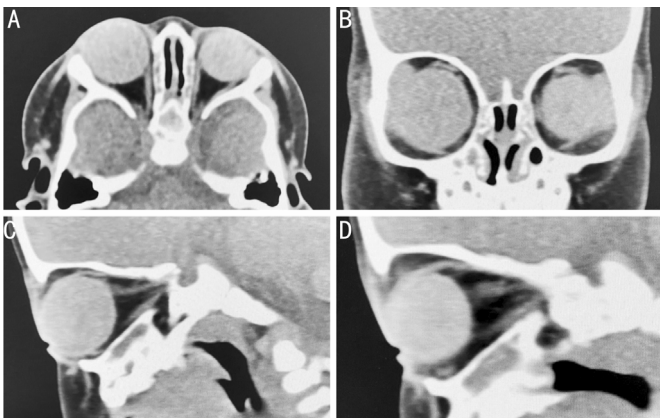


Figure 3 Result of orbital computed tomography scan (October, 2021).

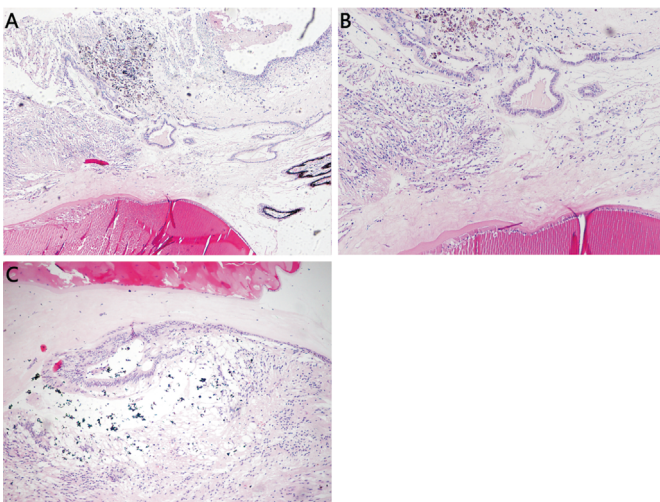


Figure 4 Result of histopathology examinations of right eye (A-B) and left eye (C).

our reported case, it is exceptionally rare that a 4-month boy suffered bilateral medulloepithelioma of the ciliary body, which was initially misdiagnosed as retinoblastoma. Thus it could be seen that the exceptionally rare occurrence of this tumor should be kept in mind while dealing with leukocoria, which was suspected as retinoblastoma.

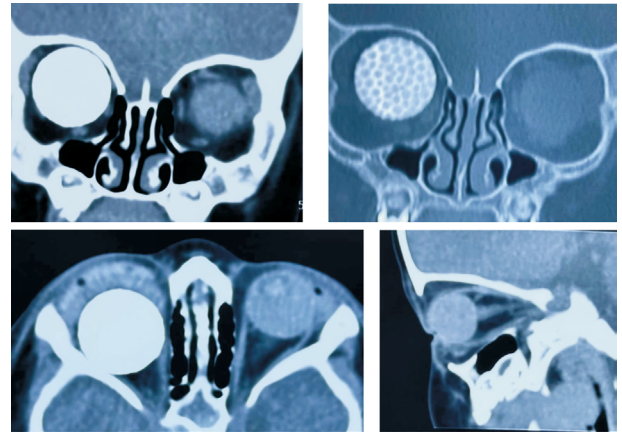


Figure 5 Result of orbital computed tomography scan (June, 2022).

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