

Adenocarcinoma of the nonpigmented ciliary epithelium manifested as an anterior chamber mass

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Dear Editor,

Adenocarcinoma of the nonpigmented ciliary epithelium (NPCE) is a rare malignant tumor which is not easy to be found in the early stage^[1]. Herein, we report a case of adenocarcinoma of the NPCE in a Chinese boy and discuss its clinical features.

A 17-year-old boy complaining of anterior chamber mass recurrence in his right eye for 8mo presented to our Department of Ophthalmology. There was a history of ocular trauma a year ago, then a mass in his right anterior chamber was observed accidentally after 20d. It was diagnosed as iris ciliary body cyst and surgically removed in the local hospital, the pathological examination result was unknown. Three months later, the tumor reappeared in the same section and continued to grow. Four months after recurrence, there was a grayish pink heterogeneous tumor in the anterior chamber, with the size of 5×4×2 mm around (Figure 1A). Previous history of tumor was denied.

On examination, the visual acuity was 0.2 in the right eye and 1.0 in the left eye. Intraocular pressure reading was 18.8 mm Hg in the right eye and 11.8 mm Hg in the left eye. Slit-lamp examination revealed a yellowish white solid mass located in the inferior anterior chamber, with uneven surface, relatively clear boundary, rich vessels and minor pigmentation. The size of the mass was 7×5×3 mm

approximately. The iris was compressed and displaced posteriorly, with pupil transformation as reniform. There were mild conjunctival congestion, hyphema, anterior chamber flare, iris neovascularization and posterior synechia, without visible cataract (Figure 1B). Anterior segment of the left eye and bilateral fundus were normal.

Color Doppler ultrasound image (CDI) showed an acoustically solid mass with dotted blood flow signal in the ciliary body of the right eye (Figure 2A). Ultrasound biomicroscopy (UBM) demonstrated a moderately irregular acoustically solid in 4:00-7:30 clock meridians in the anterior chamber of the right eye, measuring 6.05×7.78×5.29 mm, next to the peripheral cornea. The demarcation between the lesion and iris ciliary body was unclear (Figure 2B). Additionally, there were multiple echo free zones in the bilateral ciliary body, which indicated ciliary body cysts. Positron emission tomography/computed tomography (PET/CT) examination discovered no other tumors except the lesion in the right eye.

The patient underwent local iridocyclectomy of the tumor in general anesthesia (Figure 1C). Histopathologically, the tumor was diagnosed as adenocarcinoma of the NPCE. H&E staining showed that tumor cells varied in size and shape from cell to cell, arraying irregularly and forming gland-like structures in some regions. The nuclear-cytoplasmic ratio was relatively high, with obvious nucleolus and occasional mitotic figure (Figure 3A). Brisk cell growth was observed. The tumor tissue was positive for periodic acid-schiff stain (PAS) (Figure 3B). Immunohistochemical examinations showed that tumor cells were strongly positive for vimentin (Figure 3C), moderately positive for cytokeratin (CK) (Figure 3D), S-100, neuron specific enolase (NSE), and negative for epithelial membrane antigen (EMA), smooth muscle actin (SMA), glial fibrillary acidic protein (GFAP), HMB-45, Melanin-A, B-cell lymphoma-2 (Bcl-2). There was no evidence of recurrence or metastasis at the 19-month follow-up after surgery. The visual acuity of the right eye has recovered to 0.9.

The NPCE as the extension of the retinal neurosensory layer, can undergo abnormal hyperplasia due to some pathogenic factors, such as ocular trauma or endophthalmitis. Adenocarcinoma of the NPCE is extremely rare, manifested as a single, slow-growing and nonpigmented neoplasm in the posterior chamber. It mostly occurs in middle aged and elderly people, occasionally in children with previous penetrating trauma^[2].

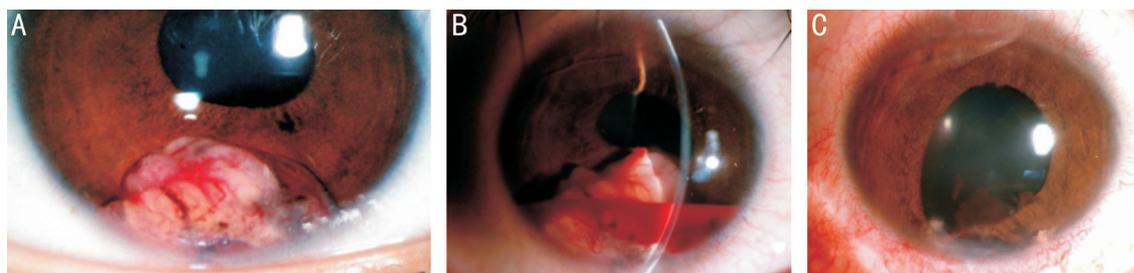


Figure 1 External eye images A: A grayish pink tumor with uneven surface and rich neovascularization in the anterior chamber (this photo was taken in the local hospital); B: The mass grew rapidly and compress the iris posteriorly, with visible pupil transformation and hyphema; C: Local iridocoloboma and posterior synechia after the operation.

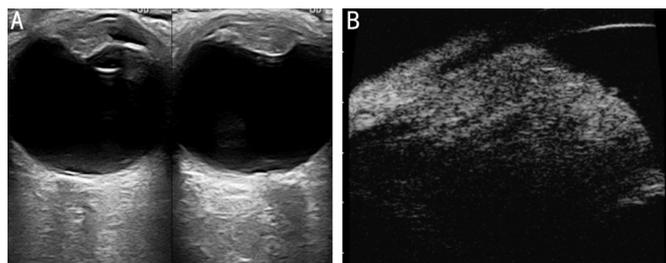


Figure 2 Imaging examination A: Ultrasonography showed an acoustically solid mass in the ciliary body; B: Ultrasound biomicroscopy demonstrated a moderately irregular acoustically solid in the anterior chamber.

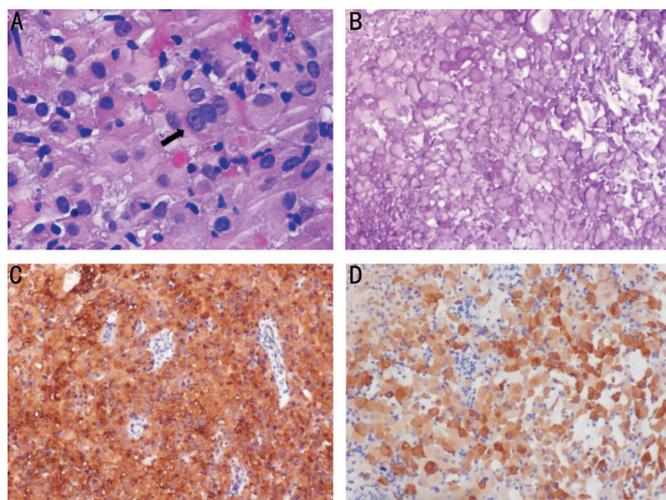


Figure 3 Pathological features A: H&E staining showed that tumor cells which varied in size and shape arrayed irregularly, with high nuclear-cytoplasmic ratio and obvious nucleolus ($\times 400$); B: PAS staining positive ($\times 100$); C: Vimentin staining positive ($\times 100$); D: CK staining positive ($\times 100$).

Minor tumor is barely detectable in the early stage, large tumor oppressing the lens may lead to secondary cataract or lens subluxation, iris bombe or neovascularization blocking the angle can result in secondary glaucoma^[3]. Then the patients complained of decreased vision or eye discomfort. In some cases, posterior segment could be involved due to large tumor or diffuse tumor, then the patients may develop secondary retinal detachment and complain of severe vision loss^[4]. Unlike most other cases, the tumor in our patient grew forward rapidly, perforated the iris and continued growing

in the anterior chamber, which may be related to the trauma or surgical history. Ultrasonographically, adenocarcinoma of the NPCE shows medium to high internal reflectivity and abrupt margin with acoustic solidity^[5]. A solid lesion with heterogenous internal echo was detected in this case.

However, the special clinical features of our patient, especially the yellowish white solid tumor on the anterior surface of iris and rapid growth rate, were easy to be confused with metastatic tumor. Metastatic tumor in the iris and ciliary body, single or multiple, mono-ocular or binocular, can be the initial sign of tumor in other sites^[6]. Although our patient had no tumor history, PET/CT examination was necessary to exclude the metastatic possibility.

Histopathological diagnosis of the tumor is the golden standard. Tumors of the NPCE have been divided into solid, papillary, and pleomorphic types, the majority are mixed type^[5]. A significant feature is its positive reaction for PAS. The tumor cells show negative reactivity to melanoma-specific antigen HMB-45, positive reactivity to CK, S-100, and vimentin in varying degrees. Adenocarcinoma of the NPCE can be distinguished from adenoma by cytologic features and extent of invasion. It is worth noting that adenoma cells that proliferate actively can sometimes show malignancy of adenocarcinoma.

In most cases, this tumor can be managed successfully by iridocyclectomy. Recurrence or metastasis is rarely seen. Good visual recovery can be expected when the tumor is localized. Enucleation may be considered in case of diffuse tumor and poor visual prognosis. The effectiveness of radiotherapy is still uncertain.

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