Pol J Pathol 2003, 54, 3, 187-190 PL ISSN 1233-9687

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Adenoma of the Iris and Ciliary Body. Case Report

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A case of rare tumor of the iris and ciliary body in a 24-year old woman is presented, which was diagnosed as adenoma of the nonpigmented ciliary body epithelium. The diagnosis was confirmed immunohistochemically.

Introduction

Iris and ciliary body tumors can be asymptomatic, larger tumors may cause subluxation of the lens, segmental cataract or secondary glaucoma. In differential diagnosis amelanotic malignant melanoma, foreign body, metastatic disease, granuloma, epithelial cyst, haemangioma, schwannoma, leiomyoma and tumor of ciliary epithelium should be considered. Adenomas of nonpigmented ciliary epithelium (NPCE) are rare tumors, that may be suspected on clinical examination. The histologic features of these tumors have been documented, also the immunohistochemical properties have been described [1 - 6, 9]. We report a characteristic cytomorphology of a case of the nonpigmented ciliary epithelium adenoma (NPCE).

A Case Description

We present a woman, aged 24, with asymptomatic, nonpigmented iris tumor of the right eye, 0.5x0.5mm in diameter, located 8-o'clock, with sentinel vessel (Fig. 1). Gonioscopy showed that the lesion did not obscured a view of the angle structures. The mass did not block transillumination. Ultrasound examination showed high internal reflectivity in the mass, with partial involvement of ciliary body and clear margins (Fig. 2). The clinical features of the lesion suggested an intraocular melanoma of the iris and ciliary body. Because of small size of the tumor and very good visual acuity we suggested to continue to follow up the patient, but the young patient did not want to have any intraocular tumor. Irydocyclectomy with surgical excision of the tumor was performed (Fig. 3). There were no complications during surgery. During follow up patient was examined 4 times. Sixteen months after surgery the patient's visual acuity was 20/20 in both eyes, intraocular pressure was normal and no evidence of recurrence of the tumor was found

The 2mmx2mmx3mm fragment of iris, sclera and ciliary body containing well separated white tumor 1.5mm in diameter was received in formalin. Histologically the tumor consisted of loosely arranged round and polygonal well-differentiated epithelial cuboid cells forming tubuloacinar pattern. The stroma of the tumor contained mucoid material. Tumor cells had eosinophilic cytoplasm and bland round to oval nuclei (Figs. 4 and 5). Cell cytoplasm and matrix were paS-positive and in some areas positive alcian blue staining was observed (Fig. 6). No mitotic figures were found in the tumor. Necrosis was absent. Only a few capillaries were visible. The growth of the neoplasm was expansive without invasion of the stroma and surroundings. The surgical margins of resection were evaluated and it was determined that the tumor had been completely excised. Immunohistochemically tumor cells were positive for vimentin (Fig.7) and for S-100 protein (Fig. 8) and negative for pancytokeratin, cytokeratin 7 (CK7) and HMB-45.

Histological, histochemical and immunohistochemical results of the examination gave the base to diagnose the tumor as adenoma of nonpigmented ciliary body epithelium, extending through the iris stroma into the anterior chamber.

Discussion

Adenoma of the nonpigmented ciliary body epithelium is clinically indistinguishable from amelanotic malignant melanoma of the ciliary body or metastatic carcinoma. The retroiridic location, anterior distortion and thinning of the iris due to tumors of NPCE have been described before [2, 3]. Location of the tumor in the anterior chamber is due to extension of the tumor mass through the iris stroma. A failure of the adenoma to block transilumination and high internal reflectivity in the ultrasound examination may be helpful. feature to distinguish it from primary melanoma.

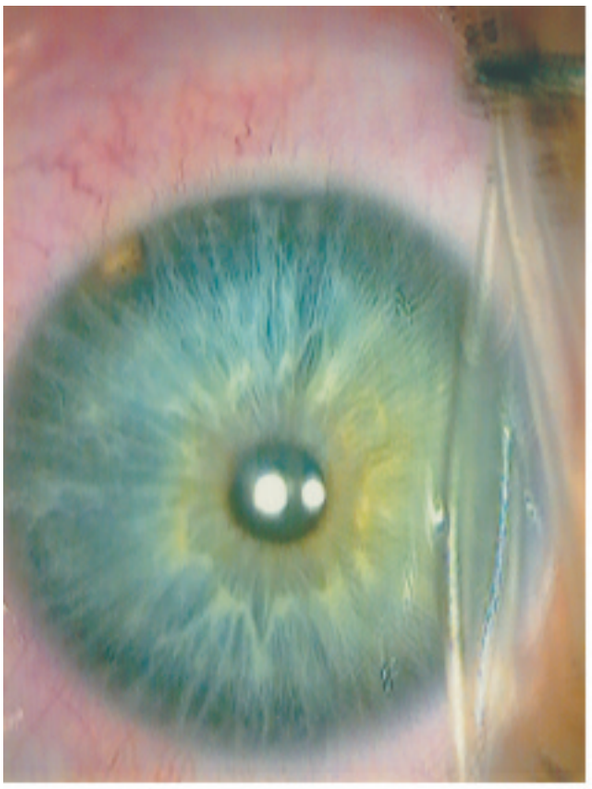

Fig. 1. Tumor of iris and ciliary body.

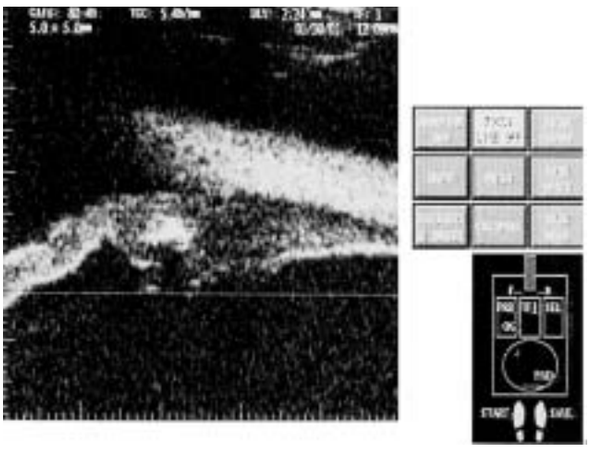

Fig. 2. Ultrasound (UBM) of ciliary body tumor.

The differential diagnosis in this case should include melanoma, pseudoepitheliomatous hyperplasia, adenoma and adenocarcinoma. The immunohistochemical characteristics: protein S-100 and vimentin positivity, pancytokeratin, cytokeratin 7 (CK7) and HMB-45 negativity showed that the tumor was composed of nonpigmented ciliary body epithelium. Lack of staining for cytokeratin 7 and HMB-45 and positive staining for S-100 protein and vimentin confirm neural origin of nonpig-

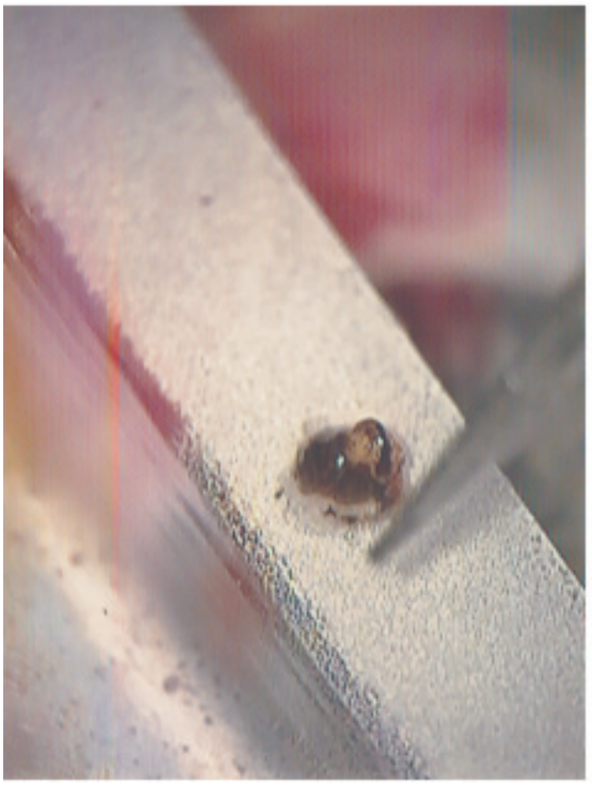

Fig. 3. Tumor of iris and ciliary body after iridocyclectomy.

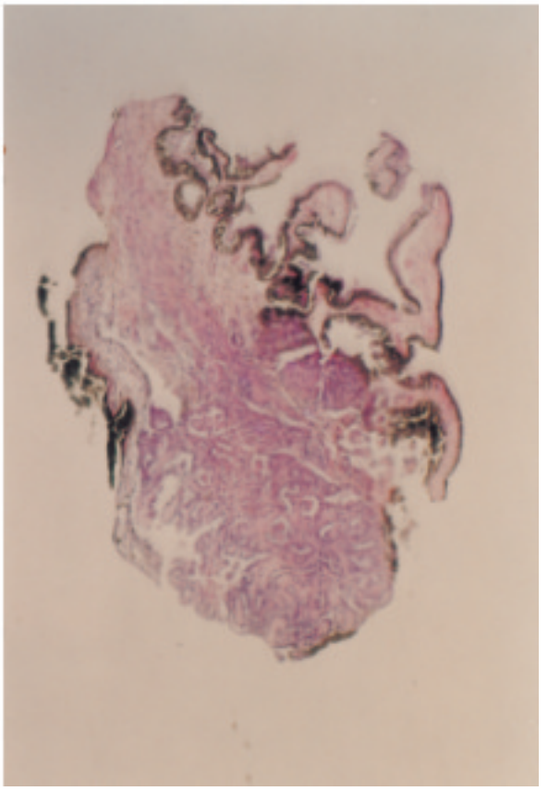

Fig. 4. Cross-section of the ciliary body and iris tumor. HE.

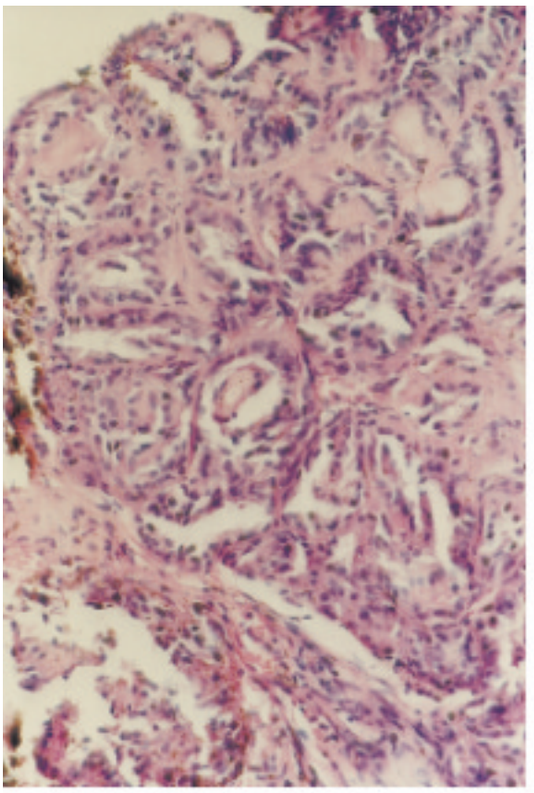

Fig. 5. The structure of adenoma of NPCE - the eosinophilic cytoplasm, well visible nuclei, no mitotic activity, no necrosis. HE.

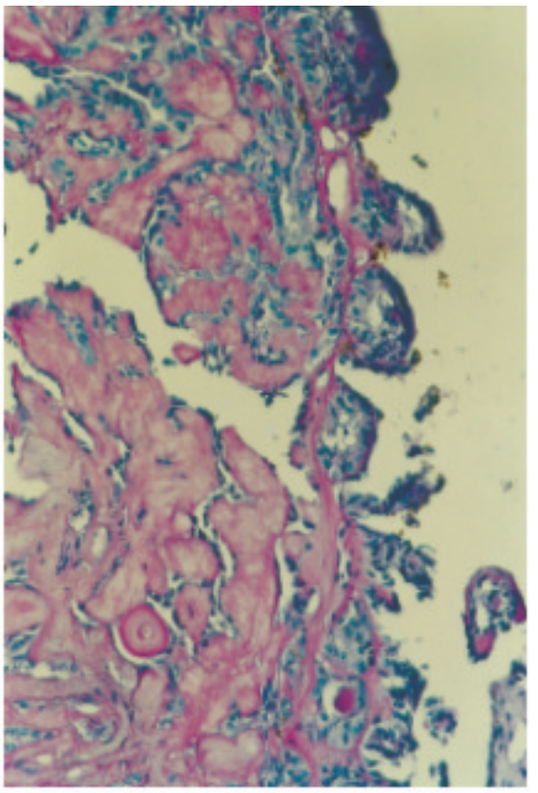

Fig. 6. Adenoma of NPCE. Alcian-blue-paS.

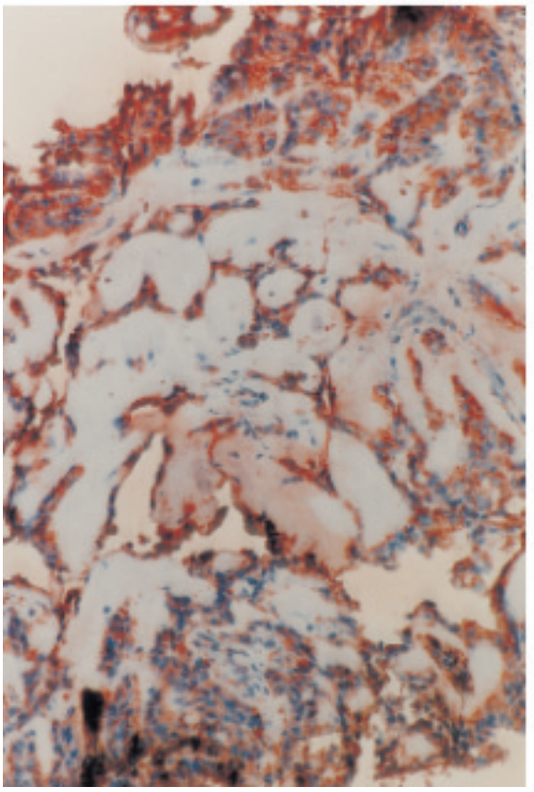

Fig. 7. Vimentin staining - positive.

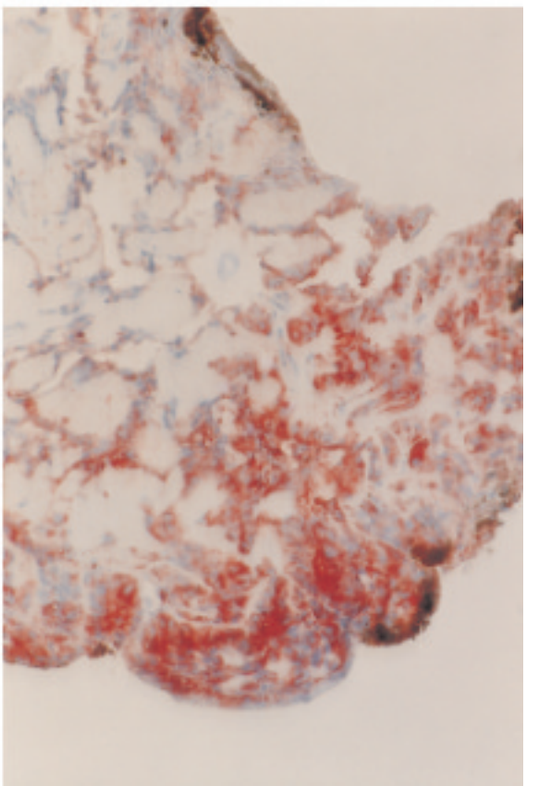

Fig. 8. S-100 protein staining - positive.

mented ciliary body epithelium. It is a part of so called "blind retina" and does not contain any pigment. It is composed of Mueller's cells organized in a single layer of glial support cells.

Primary tumors of the ciliary epithelium have been classified as congenital or acquired [2, 7 - 9]. Acquired tumors include pseudoepitheliomatous hyperplasia (Fuchs adenoma) and ciliary body nonpigmented, pigmented or mixed epithelial tumors - adenoma and adenocarcinoma, congenital - glioneuroma and medulloepithelioma [2].

The cytologic features of the glandular component of the tumor in our case are similar to that illustrated in other reports [1 - 3].

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