Ciliary body adenoma of non-pigmented epithelium

S Mansoor, A Qureshi

CASE REPORT

Primary tumours of the non-pigmented ciliary epithelium are rare. Zimmerman1 proposed dividing these tumours into two classes—congenital and acquired. Congenital tumours arise in the embryonic or early postnatal period, and include medulloepithelioma and glioneuromas. The acquired tumours are seen in adulthood and include Fuch's adenoma (hyperplasia of the NPCE, pseudoadenomatous hyperplasia, and coronal adenoma),1–3 adenoma, and adenocarcinoma of the non-pigmented ciliary epithelium.

Acquired tumours of the ciliary body have a wide spectrum of clinical and histopathological features and biological behaviour. Many are rare, and most studies have consisted of single case reports.4–6 Classification is based on the presence or absence of pigmentation, the cellular pattern, and malignant behaviour. Acquired neoplasms of the NPCE include Fuchs adenoma, adenoma, and carcinoma of the NPCE.15

“Although benign, adenoma of the non-pigmented ciliary epithelium can behave aggressively locally, causing cataract and vitreous haemorrhage”

Our patient developed a true adenoma of the NPCE, a benign tumour with variable histopathological features. The immunohistochemical results were consistent with an origin from the NPCE: S-100 and vimentin were positive. Immunohistochemistry for cytokeratin was also positive—variable staining for cytokeratin has been described in the literature.7 Although benign, adenoma of the NPCE can behave aggressively locally, causing cataract and vitreous haemorrhage.

Figure 1 Photomicrograph of the tumour showing cords of non-pigmented epithelium separated by mucoid fibrillary material; haematoxylin and eosin stain.

Abbreviations: NPCE, non-pigmented ciliary epithelium
Adenomas are differentiated from adenocarcinoma of the NPCE by the absence of local infiltrative behaviour and a moderate degree of cellular pleomorphism with nuclear atypia. Rare mitoses are found. This tumour is seen in adulthood, with an age range of 24–70 years (mean, 45), and no sex predilection. The most common presenting symptom is visual loss. A ciliary body adenoma with smooth muscle differentiation has also been reported.

Histopathological differential diagnosis of ciliary body adenoma includes ciliary body adenocarcinoma, adenoma of pigment epithelium, medulloepithelioma, in addition to lesions that occur in the deeper uveal tissue, such as melanoma, melanocytoma, metastatic carcinoma, leiomyoma, and neurilemoma.

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REFERENCES

Take home messages
- We report a case of ciliary body adenoma of the non-pigmented ciliary epithelium in a 27 year old Pakistani man—the first report of this entity in the Pakistani population to date.
- The histological and immunohistochemical profiles were typical of the adenomas described in the literature—immunohistochemistry for S-100, vimentin, and cytokeratin was positive but the HMB45 antibody was negative.

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Figure 2  Photomicrograph of the tumour showing positive immunohistochemical staining for cytokeratin.
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