

cystoid macular edema in patients with uveitis. *Ophthalmology* 2000;**107**:593–9.

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EDUCATION

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Answers

From questions on page 1582

(1) Describe the ultrasound biomicroscopic findings

Ultrasound biomicroscopy (UBM) revealed an elongated dome shaped irido ciliary mass that extended from the posterior aspect of the peripheral iris to the ciliary body (asterisk (*)) in fig 1B). The mass was centred at 7 o'clock meridian and measured 8.5 mm circumferentially, 5.0 mm anteroposteriorly and 2.4 mm in height. An incidental iris pigment epithelial cyst was also observed (fig 1B arrow). Note anterior displacement of the peripheral iris and amorphous deposits in the angle.

(2) What is the diagnosis based on fine needle aspiration biopsy sample?

The cells are large and columnar with prominent nucleoli. The cytological appearance is consistent with adenocarcinoma. (Papanicolaou stain, 100x). No particular site of origin of the tumour can be identified by cell morphology in this sample. The cytological sample was insufficient to perform immunoperoxidase stains. (B) Histopathology of the enucleated globe. Adenocarcinoma in the angle and ciliary body (haematoxylin-eosin stain, 20x). The tumour cells were weakly positive for CK 20 and strongly positive for CK7, suggesting adenocarcinoma arising in the gastrointestinal tract, lung or bladder. Metastatic ciliary body adenocarcinoma is the most likely diagnosis.

(3) How would you manage this patient?

The approach to management of this patient is twofold: detection and treatment of the primary cancer and management of the intraocular lesion. Further workup by his primary oncologists, including serum tumour markers and whole body PET/CT scans, failed to demonstrate recurrence or metastasis of the previously treated testicular tumour at other sites or additional primary cancer. The patient underwent radiation to the right eye (40 Gy in 16 fractions).

One month after completion of radiation therapy, the patient complained of significant pain, vision had reduced to 20/200, there was neovascular glaucoma with an intraocular pressure of 28 mm Hg and the tumour had progressed. The right eye was enucleated.

Histopathologically, adenocarcinoma involved the ciliary body, anterior surface of the iris (with associated neovascularisation), trabecular meshwork and posterior cornea (fig 2). Histological features of germ cell tumours were not present. Immunoperoxidase stains, performed to identify the possible primary site of this tumour, showed positivity for cytokeratins AE1/3, CK7 and CK20 with negative staining for prostatic specific antigen and thyroid transcription factor-1 suggesting a primary adenocarcinoma in the gastrointestinal tract (colon, pancreas, stomach, appendix), lung or bladder.¹

Discussion

Our case demonstrates the importance of considering intraocular malignancy in the differential diagnosis of a patient with persistent intraocular inflammation who does not respond to appropriate treatment, with a negative laboratory and imaging

workup for an underlying inflammatory disease. Meticulous examination is critical in making the correct diagnosis. In our patient, minimal anterior bowing of the peripheral iris and observation of yellow gelatinous deposits in the angle on the slit lamp examination led to a suspicion of an underlying mass lesion which was confirmed by UBM (fig 1).^{2,3}

Intraocular biopsy becomes a necessity when the systemic evaluation fails to offer alternative accessible sites for fine needle aspiration biopsy, such as the lymph nodes, lungs and liver.⁵

Uncommonly, scleritis can be the presenting ocular finding in patients with uveal metastasis.⁶ Other diagnostic possibilities include primary adenocarcinoma of the ciliary body that arises in the pigmented and non-pigmented ciliary epithelium.⁷ Clinically, these tumours mimic melanomas because they appear as slowly growing pigmented circumscribed mass.⁷ Histopathologically, the tumour cells are arranged in a glandular pattern and exhibit prominent basement membrane.⁷

The recommended approach to the management of patients with suspected or proven uveal metastasis is to investigate for the primary tumour as the nature of the primary tumour has direct implications on the diagnosis, treatment and overall prognosis.⁸ In our patient with a history of smoking, adenocarcinoma of the lung is the most likely source of the metastasis. Other sources of primary adenocarcinoma such as gastrointestinal tract or bladder are also being considered. Despite all these efforts, in about 3–5% of metastatic lesions, the primary tumour may not be identified.⁹ Teratocarcinoma of the testis is a non-seminomatous germ cell tumour that spreads by lymphatics to the retroperitoneal nodes and is not known to metastasise to the uvea.¹⁰

Final diagnosis

Iridociliary metastatic adenocarcinoma. Primary tumour undetected.

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