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Phthisis with a threat: imaging and histopathology of a ciliary body adenocarcinoma

Sai Priyalatha Mariapan[1,2], **Gowri** Supramaniam[1], **Wan Mohd Hafidz** Wan Abdul Rahman[1], **Bastion** Mae-Lynn Catherine[2]

[1]Department of Ophthalmology, Hospital Tuanku Ja'afar Seremban, Negeri Sembilan, Malaysia; [2]Department of Ophthalmology, Pusat Perubatan Universiti Kebangsaan Malaysia, Cheras, Kuala Lumpur, Malaysia

Abstract

Background: Adenocarcinoma is a rare malignancy derived from the pigmented or non-pigmented epithelium of the ciliary body and/or iris.
Case presentation: A 35-year-old male with childhood left eye (LE) blindness developed throbbing pain and redness of LE for a few months. On examination, the LE showed conjunctival injection, chemosis, opaque cornea, and raised intraocular pressure. Computed tomography (CT) revealed LE phthisis bulbi and orbital cellulitis. Histopathology indicated a malignant epithelial tumour, likely adenocarcinoma of the ciliary body, with positive pancytokeratin immunohistochemistry. Contrast-enhanced CT showed an ocular mass without distant metastasis. Four months later, magnetic resonance imaging revealed residual tumour with local infiltration. The patient underwent exenteration and recovered well with a prosthesis.
Conclusion: A high index of suspicion for malignancy, along with appropriate laboratory tests, histopathological evaluation, imaging, and surgical intervention are essential to alleviate symptoms and preserve life.

Keywords: adenocarcinoma, ciliary body, exenteration, ocular malignancy, phthisical eye

Correspondence: Dr Sai Priyalatha Mariapan, Medical Officer, Pusat Perubatan Universiti Kebangsaan Malaysia, Jalan Yaacob Latif Kuala Lumpur, Bandar Tun Razak, 56000 Cheras, Wilayah Persekutuan Kuala Lumpur, Malaysia.

Tajuk: Pthisis dengan ancaman: pengimejan dan histopatologi adenokarsinoma badan siliari

Abstrak

Latar belakang: Adenokarsinoma adalah malignansi jarang berlaku yang berasal daripada epitel pigmen atau tidak pigmen pada badan siliari dan/atau iris.
Pembentangan kes: Seorang lelaki berusia 35 tahun dengan kebutaan mata kiri (LE) sejak kecil mengalami kesakitan berdenyut dan kemerahan pada mata kiri selama beberapa bulan. Pada pemeriksaan, mata kiri menunjukkan jangkitan konjuntiva, kemosis, kornea legap, dan tekanan intraokular yang tinggi. Tomografi berkomputer (CT) menunjukkan phthisis bulbi dan selulitis orbital. Histopatologi menunjukkan tumor epithelia yang malignan, kemungkinan adenokarsinoma badan siliari, dengan imunohistokimia positif. CT yang dipertingkatkan kontras menunjukkan jisim okular tanpa metastasis jauh. Empat bulan kemudian, pengimejan resonans magnetik mendedahkan tumor sisa dengan infiltrasi tempatan. Pesakit menjalani eksenterasi dan sedang pulih dengan prostesis.
Kesimpulan: Indeks kecurigaan yang tinggi terhadap malignansi, bersama ujian makmal yang sesuai, penilaian histopatologi, pengimejan, dan intervensi pembedahan adalah penting untuk mengurangkan gejala dan menyelamatkan nyawa.

Kata kunci: adenokarsinoma, badan siliari, eksenterasi, malignansi okular, mata phthisical

Introduction

Ciliary body tumours are rare, making up approximately 6% of all intraocular tumours.[1] Ciliary body adenocarcinoma (adenoCA) that arises from the uvea has an incidence rate of less than 3% of all ciliary body tumours, making it extremely rare.[2] They can either be melanotic or amelanotic, without sex predilection, and are often associated with a phthisical eye or underlying chronic inflammation.[3] In this case report, we describe the clinical presentation and histopathological features of a case of ciliary body adenoCA.

Case presentation

A 35-year-old man with childhood left eye (LE) blindness presented to the eye clinic with increasing LE throbbing pain and redness for a few months. He was otherwise healthy. There was no family history of malignancy and the cause of blindness of the LE was unknown. He denied recent trauma to the eye. LE examination revealed periorbital swelling, injected conjunctiva with chemosis, an opaque bulging cornea, and raised intraocular pressure (IOP) of 60 mmHg. Vision was confirmed to be no light perception by 2 specialists. There was no view of the anterior chamber, pupil, iris, or fundus. B-scan ultrasonography of the LE showed a clear vitreous, flat retina, and no T-sign or loculations. Right eye (RE) and systemic review were normal. There was Grade 2 reverse relative afferent pupillary defect elicited in his RE. Extraocular muscle movement was limited in the LE by 10% in all the gazes.

The patient was admitted for urgent imaging. Computed tomography (CT) of the orbits showed the left globe wall was thickened with calcification, suggesting underlying phthisis bulbi. Additionally, the CT showed a posterolateral collection, suggestive of globe rupture. The patient was also started on pain relief and IOP-lowering medications. Despite the medications, pain control was poor, indicating that the pain could be due to an ongoing inflammation. Topical and oral antibiotics were administered to prevent or treat possible infection.

We then proceeded with LE evisceration as the definitive management for diagnosis and symptom relief for this patient. The eviscerated tissue was sent for histopathological examination (HPE). The HPE findings of the eviscerated tissue sample revealed malignant epithelial tumour with features suggestive of ciliary body adenoCA. Immunohistochemical study showed tumour cells diffusely positive for pancytokeratin and focal positive for cytokeratin. Contrast-enhanced axial CT (CECT) (Fig. 1, left) of the orbits showed a lobulated mass at the left orbital region. CECT of the thorax, abdomen, and pelvis showed no evidence of distant metastasis. Tumour markers carbohydrate antigen 19.9, carcinoembryogenic antigen, alpha fetoprotein, and prostate-specific antigen all came back normal.

This patient subsequently developed left eyelid swelling 4 months after evisceration. Magnetic resonance imaging of the orbits revealed a residual tumour with local infiltration and a bulky left lacrimal gland (Fig. 1, right). He was then referred to the oculoplastic team and underwent exenteration. He recovered well and had a prosthesis inserted 6 months after exenteration.

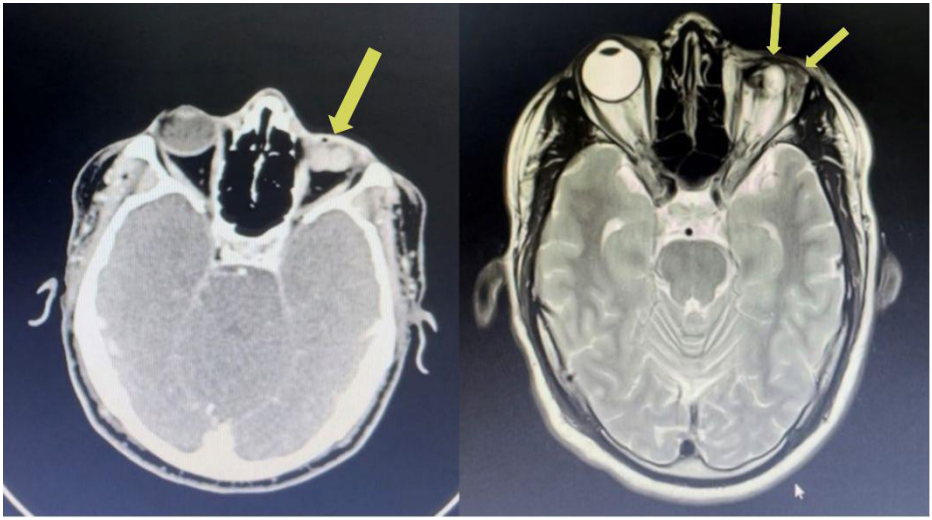

Fig. 1. (Left) CECT axial view of the orbits showing a lobulated heterogeneously enhanced mass (yellow arrow) at the left orbital region and thickening of overlying skin. *(Right)* T2W MRI axial view of the orbits showing loss of normal left globe configuration and lobulated mass measuring 1.5 x 1.1 x 1.0 cm, and a bulky left lacrimal gland (yellow arrows).

Discussion

Adenocarcinoma is a rare malignancy derived from the pigmented or non-pigmented epithelium of the ciliary body and/or iris, associated with local invasion and cellular differentiation, which may result in distant metastasis.[2] Poorly differentiated tumours such as this are commonly found in phthisical eyes with or without a history of trauma or ocular inflammation.[4]

According to the largest series on ciliary body adenoCAs to date, involving a total of 12 eyes, 9 out of 12 (75%) tumours were noted to have occurred in phthisical eyes in adults.[3] Even patients with longstanding phthisis bulbi, such as our patient, may present with new onset of proptosis, intractable pain, inflammation and swelling. Floaters, decreased vision, or increased IOP may also be one of the symptoms.[5] Some of the ocular signs that may be seen in these type of cases are as intraocular calcification, haemorrhage, secondary cataract formation, and/or subluxation of the lens. Nevertheless, when a patient presents in such fashion as our case, the first step in the presence of new symptoms of acute onset is to consider whether infection is involved; in this case, orbital infection due to the presence of chemosis. When the history is uncertain, it is appropriate to institute antibiotic prophylaxis with broad-spectrum systemic antibiotics upon admission, as even scleral buckles have been missed previously in patients with unknown history.[6] A thorough

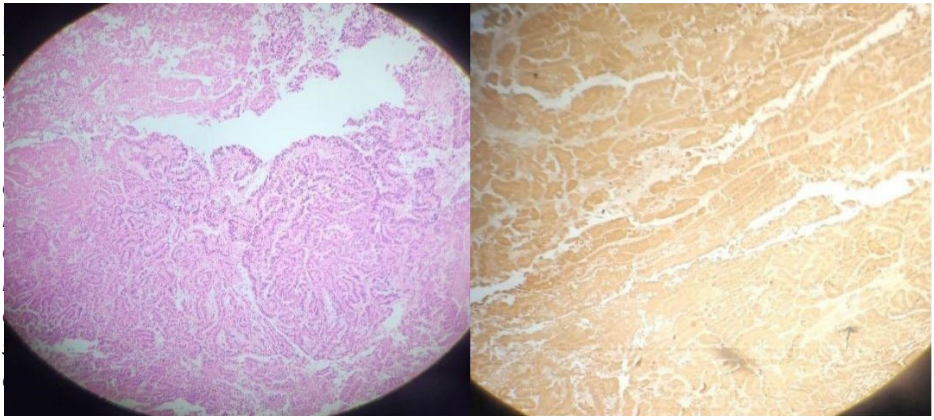

Fig. 2. (Left) Tumour cells arranged in a complex glandular pattern with prominent basement membrane. *(Right)* Tumour cells with diffuse pancytokeratin.

Indeed, renal cell carcinoma can also present for the first time with metastasis to the choroid, as has described by Feendi et al. in the absence of a primary.[9] Another type of cancer that can involve the eye and present with ocular signs and symptoms first includes leukaemia or lymphoma.[10] Hence the importance of performing systemic examination and screening for this patient to exclude a primary tumour elsewhere.

Histopathologically, primary ciliary body epithelial or retinal pigment epithelial adenocarcinomas are characterised by a prominent basement membrane, a feature also observed in the HPE of the eviscerated tissue from our patient (Fig. 2, left). This explains the inclusion of HPE findings in our report. The presence of prominent basement membrane is never a feature of metastatic carcinomas to the globe.[3] Studies have also shown that tumour cells near the iris are arranged in papillary and tubular fashion. The singular cells are round to oval in shape with hyperchromatic nuclei, high nuclear cytoplasmic ratio, and scanty cytoplasm.[5] Ciliary body adenoCAs have been described to have either one of 4 basic patterns: glandular or papillary; pleomorphic of low grade; pleomorphic with hyaline stroma; or anaplastic.[4] In our patient, the tumour cells were arranged in a complex glandular pattern (Fig. 2, left), with pleomorphic cells displaying vesicular nuclei. Microscopic examination of the eviscerated tissue from our patient revealed extensive necrosis.

Immunohistochemically, the tumour cells typically express pancytokeratins (Fig. 2, right) and cytokeratins specific to ciliary body adenoCA; these were positive in our patient as well.

Evisceration is an established treatment for the management of a painful blind eye,[3] which was the immediate management for our patient. Enucleation is typically considered when an intraocular tumour is suspected to be confined to the globe. However, our patient presented with symptoms following the initial evisceration. Fortunately, there was no evidence of tumour spread beyond the globe, aside from

extension into the adjacent orbit. The defect in the eyewall seen on the preoperative CECT scan provided an important clue, suggesting that the tumour had already extended into the orbit prior to surgery. In retrospect, exenteration would have been more prudent from this perspective. In contrast to evisceration and enucleation, exenteration is a more invasive procedure with a higher risk of disfigurement, particularly for a young man in his productive working years. If the less invasive option is pursued, then close monitoring, such as in our case, with patient education is mandatory to reduce recurrence and distant spread. Surveillance for this condition is lifelong. Our patient was also referred to the oncology team for surveillance, and an MRI 1 year after the exenteration showed no evidence of recurrence or tumour metastasis. Oncologists recommend considering systemic therapy with palliative chemotherapy to slow the progression of the disease and improve symptom control in metastatic cases. The choice between initial systemic chemotherapy and local radiotherapy depends on factors such as the patient's overall fitness, the extent of symptoms, previous treatments, and the patient's preferences.[3] Prosthesis implantation helps restore some degree of cosmesis for the patient, which goes a long way towards his self-esteem. It also has the advantage of retaining the socket shape and size, thus ensuring the eye socket and the remaining eye anatomy to function properly.

Conclusion

Ciliary body adenoCA should be suspected in patients with new symptoms and signs in their phthisical eye. Good health and well-being can be achieved through early detection and management of ocular malignancies, including the role of imaging and histopathology in diagnosis and treatment.

Declarations

Informed consent
This case report was written using the clinical data of the patient in 2021. Verbal consent was obtained in which the patient agrees with the research use of images of the eye, clinical records, and data with anonymization.

Competing interests
The authors declare that there is no conflict of interest with respect to the publication of this article.

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