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Michelle M. Hui

Rohan Merani

Fiona Bonar
The University of Notre Dame Australia, fiona.bonar1@nd.edu.au

Angela M. Hong

Adrian T. Fung

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Case report

Presumed choroidal metastasis from soft tissue myoepithelial carcinoma

Michelle M. Hui (BMed, MD, MMed)[a], Rohan Merani (MBBS, MMed, FRANZCO)[b,c,d], Fiona Bonar (MB, FRCPI, FRCPath (Lond), FRCPA)[d,e,f], Angela M. Hong (MBBS, MMed, PhD, FRANZCR)[g,h], Adrian T. Fung (MBBS, MMed, FRANZCO)[c,d,i,*]

[a] Royal Prince Alfred Hospital, Sydney, New South Wales, Australia
[b] Concord Repatriation General Hospital, Sydney, New South Wales, Australia
[c] Save Sight Institute, University of Sydney, Sydney, New South Wales, Australia
[d] Faculty of Medicine and Health Sciences, Macquarie University, Sydney, New South Wales, Australia
[e] Douglass Hanly Moir Pathology, Macquarie Park, Sydney, New South Wales, Australia
[f] School of Medicine, Notre Dame University, Sydney, New South Wales, Australia
[g] Central Clinical School, University of Sydney, Sydney, New South Wales, Australia
[h] Genesis Cancer Care, Radiation Oncology, Mater Hospital, New South Wales, Australia
[i] Westmead Hospital, Sydney, New South Wales, Australia

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ABSTRACT

Purpose: To report a case of presumed choroidal metastasis from soft tissue myoepithelial carcinoma and highlight challenges in its diagnosis.

Observations: A 52-year-old man was referred with a two-week history of photopsia in his left eye. His background medical history included known soft tissue myoepithelial carcinoma metastatic to his bone, lung, liver and chest wall. A large, raised, yellow choroidal lesion was identified nasal to and abutting the optic disc. This lesion demonstrated growth 1 month after presentation. The patient died with widespread metastatic disease 5 months after initial presentation.

Conclusion and importance: Soft tissue myoepithelial carcinoma can rarely metastasise to the choroid and present as a rapidly-growing, yellow, echodense tumour with serous retinal detachment. MRI brain can assist in tumour evaluation and monitoring progression, while immunoperoxidase stains and molecular testing can assist with diagnosis. The condition has an aggressive natural history and poor prognosis.

Introduction

Metastases to the choroid are an uncommon presentation of metastatic malignancy, generally associated with advanced disease. The most common primary sites are the lung and breast. We describe a rare case of presumed choroidal metastasis from soft tissue myoepithelial carcinoma, and highlight features required to make this challenging diagnosis.

Report of case

A 52-year-old Caucasian male was referred with a six-week history of noticing flickering lights in his left eye. Visual acuities were 20/20 in both eyes. Anterior segment examination was normal, and the vitreous was quiet. A large (10mm diameter) raised yellow lesion was observed nasal to and abutting the left optic disc (Fig. 1A). The right fundus was normal.

Two weeks after initial presentation, he noticed a temporal shadow in his left eye. The lesion documented rapid growth by 4mm at 2 weeks (Fig. 1B) and 6mm at 4 weeks (Fig. 1C), when it was associated with an inferior serous retinal detachment. Although the lesion was isoautofluorescent, the areas of subretinal fluid were hyperautofluorescent (Optos PLC, Dunfermine, UK Fig. 1D). Optical coherence tomography (CIRRUS™ 5000 HD-OCT, Carl Zeiss Meditec AG, Jena, Germany) demonstrated subretinal fluid over the lesion and at the macula (Fig. 1E). Brightness-scan (B-scan) ultrasonography (Eye Cubed™, Ellex, Mawson Lakes, Australia) demonstrated an echodense lesion measuring 4.0mm in height with overlying and inferior subretinal fluid but no extraocular extension (Fig. 1F). Magnetic resonance imaging (MRI) scan of the brain (Sequences used: Three plane T1, T2, diffusion and susceptibility

* Corresponding author. 1A/12 Central Road, Miranda, NSW, 2228, Australia.
E-mail address: adrian.fung@sydney.edu.au (A.T. Fung).

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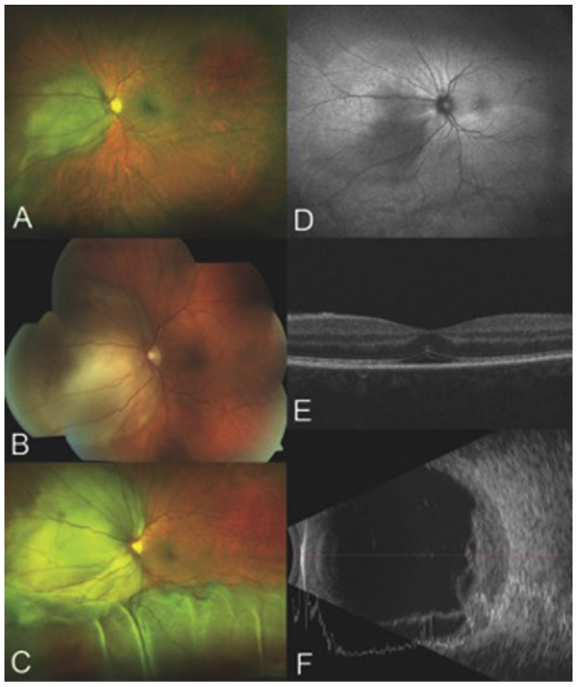

Fig. 1. A large raised yellow choroidal lesion is present nasal to and abutting the left optic disc (A). The lesion grew by 4mm (B) and 6mm (C) in diameter 2 and 4 weeks after presentation respectively. The lesion is isoautofluorescent but the areas of subretinal fluid are hyperautofluorescent (D). Optical coherence tomography demonstrates a small amount of subretinal fluid at the macula (E). B-scan ultrasonography shows an echodense tumour measuring 4.0mm in height with overlying and inferior subretinal fluid but no extraocular extension (F).

weighted and post-contrast volumetric fluid attentuated inversion recovery) showed thickening of the choroid in the medial aspect of the left globe with no evidence of intracranial metastases.

The patient had no significant ocular history. However, he was recently commenced on an immunotherapy trial (durvalumab and temelimumab) for widespread metastatic poorly differentiated soft tissue myoepithelial carcinoma, which initially presented to his right knee three years ago. Subsequently, he developed metastases in his bone, liver lung and chest wall. He underwent numerous cycles of multimodal treatment including surgery, stereotactic radiotherapy to the liver and chemotherapy, but the results were unfruitful.

Given the rapid progression of symptoms and growth of the tumour, he received radiotherapy to his left eye using via volumetric modulated arc therapy technique (30Gy in 10 fractions) using stereotactic radiosurgery setup. His vision remained stable during the treatment.[1] Unfortunately, no choroidal biopsy or further ocular follow-up was possible as the patient's condition rapidly deteriorated; he developed two levels of spinal cord compression from metastases in his thoracic and lumbar spine. Sadly, he passed away in palliative care five months after the initial ophthalmic presentation.

Discussion

The diagnosis of choroidal lesions is primarily based on clinical examination and investigations. In our case, the patient presented with a rapidly growing pale lesion, associated with serous retinal detachment, hyper-autofluorescence associated with subretinal fluid and echodensity on B-scan ultrasonography. These features in conjunction with a known history of biopsy proven metastatic soft tissue myoepithelial carcinoma make choroidal metastasis from this tumour by far the most likely diagnosis.

Metastatic soft tissue myoepithelial carcinoma to the choroid is exceedingly rare. A search of PubMed, MEDLINE, and the Cochrane Library with keywords “myoepithelial carcinoma”, “soft tissue myoepithelioma”, “metastasis” and “choroid” revealed no previous cases. There has been one report of soft tissue myoepithelial carcinoma metastatic to the orbit of an 11-year-old boy from his left thigh, who died from widespread metastases.[2] Soft tissue myoepithelial carcinoma should be recognised as a potential primary site that can cause metastases to the choroids since 34% of patients presenting with choroidal metastases have no known primary at the time of ocular presentation, and in half of these, a primary site is never found.[3]

Differential diagnoses considered included amelanotic choroidal melanoma and choroidal metastasis from another primary. Amelanotic melanoma is less likely given the rapid growth of the tumour and echodensity on B-scan ultrasound. The mean time for choroidal melanoma to double in size is 128.2–291.6 days for mixed and spindle cell types respectively,[4] compared to only 31 days in our patient. Most choroidal melanomas are echolucent on B-scan ultrasound. No other potential primary neoplasm was detected on systemic investigations, making this also unlikely.

The classification of myoepithelial tumours of soft tissue is still evolving, and the specific histopathology has not been fully elucidated.[5] It affects male and females equally with the peak incidence in the third to fifth decades.[6,7] Typically it presents as a painless or painful mass, commonly found in the extremities, especially in the lower limbs, followed by the neck, head and trunk.[6,7] The morphology and immunophenotype are similar to their counterparts in the salivary gland. They are composed of epithelial and/or myoepithelial cellular elements present in varying proportions usually in a hyalinised to chondromyxoid stroma and may show foci of ductal differentiation. They show overlap with mixed tumour of skin and soft tissue. Current nomenclature includes that of soft tissue myoepithelioma, which have bland “benign” appearing morphology and morphologically malignant forms which are called myoepithelial carcinoma.[7–9]

Although the vast majority of such neoplasms behave benignly, a minority of cases may recur locally and metastasise. Currently, no morphologic features reliably predict behaviour, however cytological atypia represents the most reliable prognostic parameter.[6] Soft tissue myoepithelial carcinoma behaves aggressively with a 39% recurrence rate, and 52% of patients may develop distant metastases.[7]

In our case, the patient initially presented with a soft tissue lesion medial to the right patellar tendon. Histopathology revealed a malignant tumour, the morphology of which was reminiscent of a poorly differentiated large cell carcinoma in which irregular islands of cells were present in small sheets and strands, the latter lying in the hyalinised stroma. Immunoperoxidase stains identified expression of a variety of keratins including cytokeratin (CK) 8/18 and CK 7 with focal expression of CK AE1/3, CK 5/6 and CK 19. Polyclonal carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA) was also positive, and co-expression of S100 protein was identified in areas. CK 14 and p63 were also negative as were a variety of other markers including endothelial and myoid markers. Integrase interactor 1 (INI1) nuclear staining was retained (Fig. 2). The differential diagnoses included metastatic carcinoma, poorly differentiated synovial sarcoma and, given the location in the tibia, an unusual form of adamantinoma like Ewing sarcoma. Fluorescence in situ hybridization (FISH) testing for SS18 and Ewing sarcoma breakpoint regional 1 (EWSR1) were negative, excluding Ewing sarcoma and synovial sarcoma.

The diagnosis of poorly differentiated soft tissue myoepithelial carcinoma can be challenging as observed in this case. Immunoperoxidase stains are often used to assist with diagnosis, usually associated with co-expression of keratins, S-100, p63 and myoid markers such as actin, calponin, caldesmon and desmin.[8,9] Molecular findings such as rearrangements in EWSR1 on FISH testing can also assist in diagnosis but is only present in 50% of cases and not mandatory for diagnosis.[10–12] The morphological features and co-expression of cytokeratins and S100 protein with the absence of a primary tumour

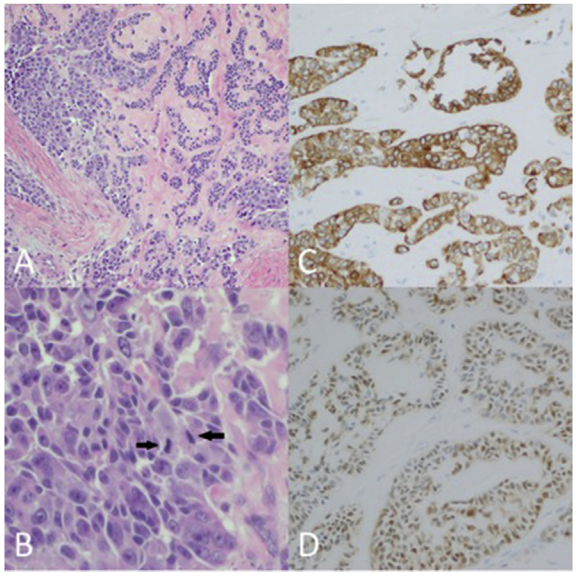

Fig. 2. Sheets and strands of tumour cells with epithelial morphology in a focally hyalinised stroma (A). Higher power shows highly atypical cells with vescicular nuclei, prominent nucleoli and several mitoses (arrow) (B). Cytokeratin 8/18 expression (C) and co-expression of S100 protein (D) supporting a myoepithelial tumour.

elsewhere were consistent with of a poorly differentiated soft tissue myoepithelial carcinoma in this case.

The prognosis of soft tissue myoepithelial carcinoma is poor. Current literature of patients treated with radiation and chemotherapy in recurrent and metastatic of soft tissue myoepithelial carcinoma have been reported to be unsuccessful.

Conclusion

In summary, we report a rare case of presumed choroidal metastasis from soft tissue myoepithelial carcinoma that presented as a rapidly-growing, yellow, echo-dense choroidal tumour with serous retinal detachment. The lesion is isoautofluorescent, but hyperautofluorescence can be seen in areas of subretinal fluid. MRI brain can assist in tumour evaluation and monitoring progression, while immunoperoxidase stains and molecular testing can assist with confirming this difficult to diagnose condition. It is important to include this condition in the differential diagnosis of choroidal metastases because one-third of choroidal metastases have no known primary at the time of presentation. Soft tissue myoepithelial carcinoma has a highly aggressive natural history, is resistant to current anti-neoplastic treatments and has a poor prognosis.

Patient consent

Patient consent: Patient provided both orally and written consent for the information gathered and the publication of his case. This report does not contain any personal information that could lead to identification of the patient.

Acknowledgements and Disclosures

Conflict of interest

There are no conflicts of interest and the following authors have no financial disclosures: MH, RM, FB, AH, AF.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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