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Case Report

Just another metastatic carcinoid tumour to the uveal tract

Ivette Hernández-Ayuso [a,*]; Abelardo A. Rodríguez-Reyes [b]; Dolores Ríos y Valles-Valles [b]; P. Ayumi Kawakami-Campos [c]; Sharon L. Herrera Cifuentes [d]

Abstract

We describe the clinic, image, and histopathologic features of a well differentiated neuroendocrine carcinoma (carcinoid tumour) metastatic to choroid and ciliary body in a 52-year-old Mexican Mestizo man. The ophthalmologic examination showed an inferior choroidal mass accompanied by exudative retinal detachment. Ultrasound B-Scan study revealed a diffuse thickened choroid with overlying serous retinal detachment, ultrasound A-Scan revealed a high internal reflectivity solid lesion. Ultrasound biomicroscopy (UBM) evidenced a dome shaped ciliary body mass, presumptive diagnosis was uveal tract metastatic disease. Scleral flap choroidal incisional biopsy was performed. Microscopic evaluation demonstrated a hypercellular lesion replacing choroid, composed by cohesive oval-round cells with finely granular chromatin arranged in organoid pattern. Immunohistochemical reactions were Pankeratin AE1/AE3 (+), Cytokeratin CK5/6 (+), Chromogranin A (+), Ki67 (20%), typical well differentiated neuroendocrine carcinoma (carcinoid tumour) was diagnosed. Patient had a mediastinal carcinoid diagnosed 3 years earlier. Metastatic cancer to the eye is perhaps the leading cause of intraocular tumour, despite this fact metastases are rarely seen by the ophthalmologist while the patient is alive. Intraocular metastasis should be considered in the presence of ciliary body or/and choroidal amelanotic or pigmented mass and serous retinal detachment in a patient with history of carcinoid tumor, althought its low frequency (2.2%).

Keywords: Mediastinum, Choroidal metastasis, Carcinoid tumour, Immunohistochemistry, Chromogranin A

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Introduction

Neuroendocrine tumours arise from enterochromaffin cells that are producers of biologically active amines, intestinal peptides and serotonin. They are diffusely localized in the gastrointestinal and bronchopulmonary systems.[1] In the classification of neuroendocrine tumours, carcinoid tumour corresponds to a typical well differentiated neuroendocrine carcinoma, atypical carcinoid makes reference to a moderately differentiated neuroendocrine carcinoma and finally the poorly differentiated neuroendocrine carcinoma.[2] The majority of carcinoids are found within the gastrointestinal tract (55%) and bronchopulmonary system (30%). Less common primary sites include breast, larynx, thymus, and gallbladder.[3] Metastatic disease affects liver, bone and lymph nodes. Ocular metastases are uncommon but when they

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[a] Ophthalmic Pathology Service, Ophthalmologist, Ophthalmic Pathologist, Asociación para Evitar la Ceguera en México I.A.P ''Hospital Dr. Luis Sánchez Bulnes'', Mexico City, Mexico
[b] Ophthalmic Pathology Service, Anatomic Pathologist, Ophthalmic Pathologist, Asociación para Evitar la Ceguera en México I.A.P ''Hospital Dr. Luis Sánchez Bulnes'', Mexico City, Mexico
[c] Retina Service, Ophthalmologist, Vitreo-Retinal Surgeon, Asociación para Evitar la Ceguera en México I.A.P ''Hospital Dr. Luis Sánchez Bulnes'', Mexico City, Mexico
[d] Pathology Service, Anatomic Pathologist, Ophthalmic Pathologist, ''Hospital Español de México'', Mexico City, Mexico

* Corresponding author at: Asociación para Evitar la Ceguera en México, ''Hospital Dr. Luis Sánchez Bulnes'', Vicente García Torres No. 46, Barrio San Lucas Coyoacán, Ciudad de México 04030, Mexico. Fax: +52 (55) 10841400x1210.
e-mail address: varitahernandez@hotmail.com (I. Hernández-Ayuso).

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appear uveal tract is usually affected, rarely the orbit.[4] Primary typical well differentiated neuroendocrine carcinoma (carcinoid tumour) represents 2.2% of uveal tract metastases.[5] The combination of positive immunohistochemistry epithelial markers such as Pankeratin (AE1/AE3), Cytokeratin (CK5/6) and Epithelial Membrane Antigen (EMA) as well as neuroendocrine markers Chromogranin A and Synaptophysin (Syn) confirms the diagnosis. In difficult cases electron transmission microscopy is useful in the identification of neuroendocrine granules.

Case report

A 52-years-old Mexican-Mestizo male complained of a 1 month history of sudden onset superior scotoma. The patient had a mediastinal mass resection 3 years earlier, receiving radiotherapy and chemotherapy as complementary treatment. The best corrected visual acuity of right and left eye were 20/25 and 20/40 respectively. Ocular examination revealed an inferior serous retinal detachment in association with an inferior choroidal pigmented mass in the left eye. B-scan ultrasound showed nodular choroidal thickening with retinal detachment. Uveal tract metastasis was suspected, scleral flap incisional biopsy was performed in the 5 O'clock meridian at 4 mm from limbus (Fig. 1). Histopathologic study showed a portion of normal sclera, the adjacent choroid was moderately infiltrated by cohesive oval-round cells with finely granular chromatine arranged in an organoid pattern. Immunohistochemistry reactions were Pankeratin AE1/AE3 (+), Cytokeratin 5/6 (+), Chromogranin A (+) (Fig. 2), Synaptophysin and CD56 were non contributory, p63 and CD5 (−) Ki67 20%. Typical well differentiated neuroendocrine tumour (carcinoid tumour) metastatic to uveal tract (ciliary body and choroid) was diagnosed. Later on, the patient obtained his mediastinal tumour histopathologic study with the diagnosis of typical well differentiated neuroendocrine tumour (carcinoid tumour) of the anterior mediastinum, confirming the metastatic origin of the choroidal neoplasm.

Discussion

Metastatic cancer to the eye is perhaps the leading cause of intraocular tumours, despite this fact metastases are rarely seen by the ophthalmologist while the patient is alive. A study performed in an eye bank population from cancer deceased patients, proved metastatic disease in 4.7% to 12.6% of cases.[6] Intraocular metastases are as 12 times more frequent than uveal melanoma, they are usually located in the posterior choroid arising from a distant primary carcinoma.[7] Most of the patients presenting with ocular metastases have previous history of cancer treatment. Clinical manifestations vary according to metastasis location. Choroidal affection is found as a single or multiple, pigmented or amelanotic perhaps slightly orange mass accompanied by considerable subretinal fluid leading to serous retinal detachment causing visual lose or localized scotoma.[8]The reported prevalence of clinically evident uveal metastases in carcinoma patients ranges from 2% to 9% with breast and lung cancer together accounting for between 71% and 92% of cases.[9]

In conclusion, neuroendocrine carcinomas represent about 0.5% of all newly diagnosed malignancies. Metastatic disease of neuroendocrine carcinoma accounts for 12–22%.[10] Primary mediastinal neuroendocrine carcinoma is *per se* unusual, representing less than 5% of all the anterior mediastinum neoplasms.[11] Mediastinal typical well differentiated neuroendocrine carcinoma (carcinoid tumor) biological behaviour is similar to carcinoids elsewhere.

The most frequent primary localization of the carcinoid tumour metastatic to the uveal tract is the bronchoalveolar system. Intraocular metastases from a mediastinal carcinoid are uncommon. Shields and cols. reported that 2.2% of the uveal tract metastases arise from carcinoid tumours.[12]

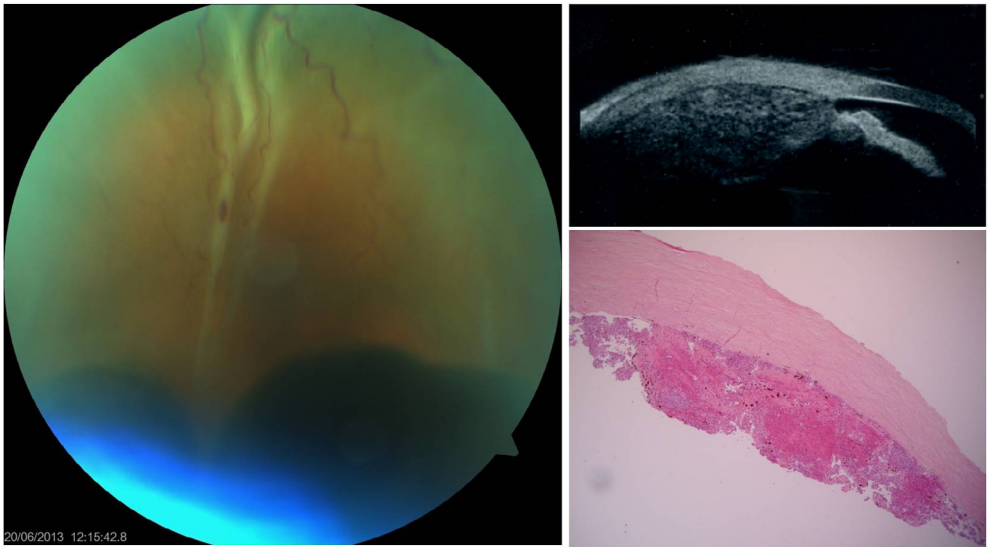

Fig. 1. Funduscupy revealing an inferior choroidal pigmented mass with serous retinal detachment. UBM showing a dome shaped ciliary body mass. Microscopic view of the incisional biopsy (slceral flap), with hypercellular lesion replacing the choroid.

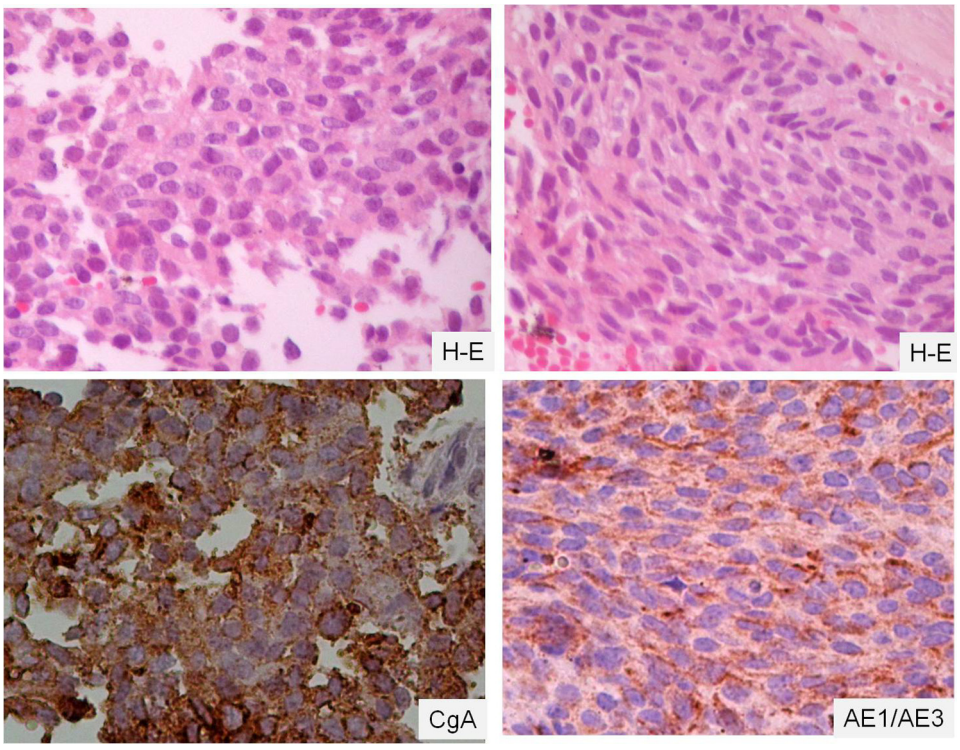

Fig. 2. Histopathological examination showed cohesive oval-round cells with fine granular chromatine, disposed in an organoid pattern. Immunohistochemical reactions demonstrated metastatic carcinoid tumour cells postive for CgA and AE1/AE3.

Intraocular metastasis should be considered in the presence of ciliary body or/and choroidal amelanotic or pigmented mass with serous retinal detachment in a patient with history of carcinoid tumor regardless primary location.

Conflict of interest

The authors declare that they have no conflict of interest.

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