

Carcinoid Tumor of the Ovary with Bilateral Infiltration of the Extraocular Muscles: A Case Report and a Brief Review of the Literature

*Ali Sadeghi-Tari, MD,¹ Mohammad-Taher Rajabi, MD²
Fahimeh Asadi-Amoli, MD,³ Shahram AghaRokh, MD⁴*

Abstract

Purpose: We report an unusual case of trabecular carcinoid of the ovary with orbital involvement.

Methods: We encountered a 49-year-old woman with a complaint of bilateral proptosis. Ocular examination revealed severe bilateral proptosis and signs of corneal exposure. Orbital CT scan showed massive enlargement of extraocular muscles and optic nerve compression. The patient medical history showed that she have had an ovarian mass for which she was underwent right oophorectomy followed by left oophorectomy and total abdominal hysterectomy.

Results: Histological examination showed trabecular carcinoid in both ovaries. Fine needle aspiration from extraocular muscles showed trabecular carcinoid tumor compatible with the primary tumor of the ovary.

Conclusion: Ocular metastases from carcinoid tumors are considered rare. They can be the primary presentation of a carcinoid tumor or develop during the course of the disease. The extent of distant metastases from carcinoid tumors correlates with poor prognosis and survival; early detection of metastasis may change the overall management.

Keywords: carcinoid tumor, extraocular muscles, proptosis

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Introduction

Carcinoid tumors are slow growing, low-grade malignant neoplasms that are believed to originate from neuroendocrine cells, usually in the gastrointestinal mucosa.¹⁻³ The most common sites of origin are appendix (40%), jejunioileum (27%), rectum (13%) and bronchus (11.5%). Some rare primary locations are thymus, thyroid, biliary system, pancreas, and ovaries.³ Metastasis of

carcinoid tumor to the orbit is a rare occurrence. When metastasis does occur, the choroid is the most common ocular structure involved.¹ Carcinoid tumor metastatic to the orbit accounts for only 4% to 5% of orbital metastatic disease.⁴ It typically is seen as a unilateral orbital mass inducing proptosis and diplopia.^{5,6}

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1. Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
 2. Ophthalmologist, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
 3. Associate Prof. of Pathology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
 4. Resident in Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences

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Correspondence to:
Mohammad-Taher Rajabi, MD
Eye Research Center
Farabi Eye Hospital
Tel: 55414941-6
Email: mt_rajabi@yahoo.com

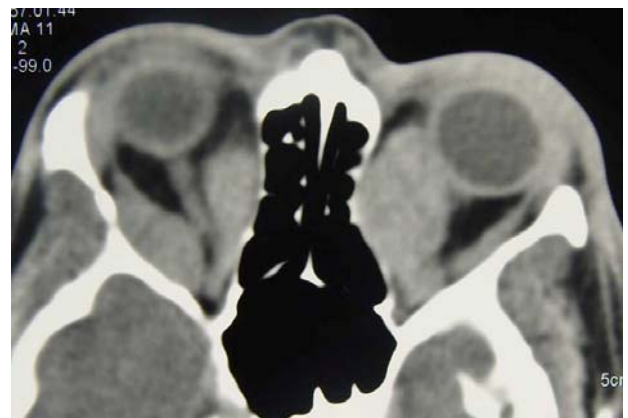
On anatomical imaging studies, such as CT scanning and magnetic resonance imaging, carcinoid tumor metastases to the orbit are characteristically focal, round, sharply delineated, nonspecific tumor masses, and demonstrate an affinity for the extraocular muscles.⁷ Orbital inflammation is an infrequent presentation for patients with this tumor.²

The authors report a rare case of metastatic carcinoid tumor with involvement of extraocular muscles who had had one self-limiting episode of acute orbital inflammation in the past.

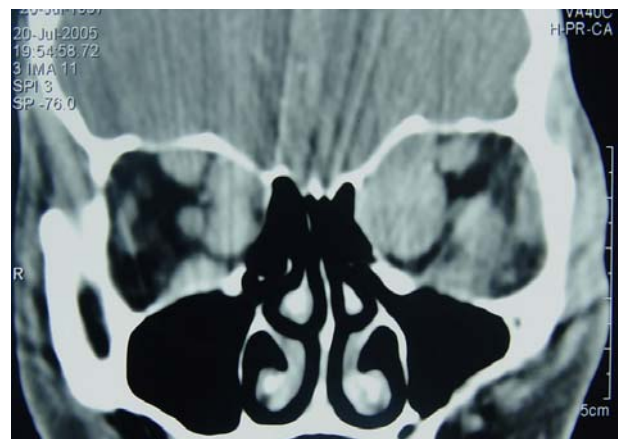
Case Report

We encountered a 49-year-old woman with a complaint of bilateral proptosis, periorbital swelling, and decreased vision of 1-month duration. She had injection of both eyes from five months before. She described a similar episode three years earlier that spontaneously subsided over three months. He could not recall antecedent trauma. Her medical history was notable for epitheloid hemangioendothelioma of common bile duct 12 years ago that had been undergone surgery. He also had history of trabecular carcinoid tumor of the ovaries that first underwent right oophorectomy and then left oophorectomy and total abdominal hysterectomy 5 years ago.

Visual acuity was $\frac{8}{10}$ OD and $\frac{6}{10}$ OS. There was marked upper and lower lid edema on both eyes, and 3 mm of proptosis. In both eyes conjunctiva showed marked vascular injection. Motility of the right eye was globally reduced but on the left eye adduction and abduction were severely reduced. Fundus examination of both eyes was normal. The patient was afebrile and without leukocytosis. A computed tomography scan of the orbits showed proptosis of both globes, massive enlargement of extraocular muscles particularly widely involving the medial rectus of the left eye and optic nerve compression (Figure 1). There was no invasion of the optic nerve or bone. The patient underwent Fine needle aspiration (FNA) from extraocular muscles. Histological examination confirmed the diagnosis of metastatic carcinoid. It was composed of many isolated small clusters or sheets of monomorphous cells with relatively large hyperchromatic nuclei, inconspicuous nucleoli and scant cytoplasm (Figure 2).



A



B

Figure 1. A,B; computed tomography scans of the orbits show proptosis of both globes, massive enlargement of extraocular muscles particularly the medial rectus of the left eye and optic nerve compression. Optic nerves and bony walls are spared..

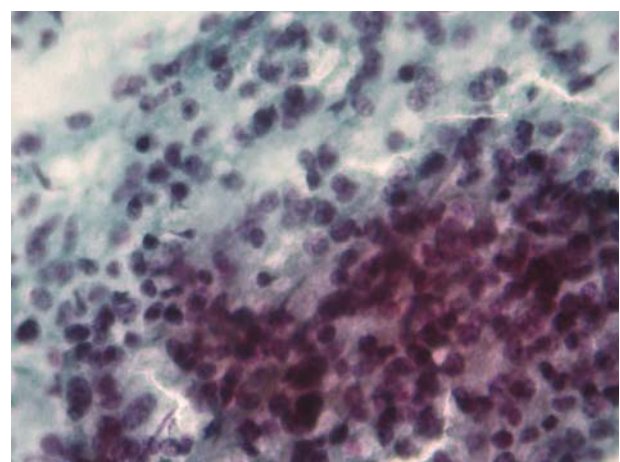


Figure 2. Histopathologic evaluation of specimen from fine needle aspiration with Papanicolaou staining showed frequent isolated small clusters or sheets of monomorphous cells with round nuclei and scant cytoplasm in the background of granular material.

Despite systemic chemotherapy, her tumor progressed. By the time of ocular presentation, her carcinoid tumor had metastasized to the liver. Unfortunately, 3 months later the patient died due to massive liver metastases.

Discussion

Metastatic tumor is one of several etiologies of space-occupying masses in the orbit that accounts for 1%-13% of all orbital masses.⁸ In the adult patient population, breast cancer is the most common tumor to metastasize to the orbit followed by metastases from the lung, prostate and gastrointestinal tract. The most common site for carcinoid metastases is the liver.¹ It is rare for carcinoid tumors to metastasize to the eye or to the orbit. Metastatic potential is mainly correlated to the tumor size and the site of origin, although histological atypia is also associated with a greater risk of metastatic spread. Tumors larger than 1 cm are likely to metastasize.⁹

In a study of 227 patients with metastatic carcinoma to the eye or orbit only two had a metastatic carcinoid.¹⁰ Nevertheless, several cases of carcinoid metastatic to the orbit were reported after the first large study in 1980 including 15 patients.^{5,11} To the best of our knowledge, there are only 11 documented reports of carcinoid metastases to the extraocular orbital muscles.^{2,3,6,7,12-14,16} In only one report, all of the extraocular muscles were involved¹¹, and just two cases involved the inferior rectus only.^{3,15} Our patient developed orbital metastasis nearly 24 months after the primary operation for ovarian carcinoid. This is a significantly shorter period of time than the average of 62.7 months from the diagnosis of systemic disease to the eye involvement previously reported in a group of 21 patients.¹² Presentation of orbital carcinoid tumor as orbital inflammation is rare.^{2,5,6,17} In a clinicopathologic series of patients with choroidal and orbital carcinoid tumor, *Riddle et al*⁵ described seven patients with orbital metastasis. Of the six cases whose clinical data were included, only one patient had chronic orbital inflammation.⁵ In a case series of three orbital carcinoid tumors, *Fan et al*⁶ described a single patient who presented with proptosis, chemosis, and dysmotility. *Skibell et al*¹⁷ described a solitary case of orbital carcinoid tumor masquerading as acute orbital

cellulitis. Ocular manifestations of the systemic carcinoid syndrome include proptosis, periorbital edema, tearing, conjunctival injection, decreased central retinal artery pressure, intraretinal vascular sludge, and perivascular retinal pigment clumping.¹⁸

The inflammation manifested by our patient during the first orbital compliant 2 years after oophorectomy was self-limited. The rapid clinical resolution without antimicrobial therapy at that time refutes an infectious cause of the inflammation. The tumor location, lack of superior ophthalmic vein distention, and spontaneous resolution do not support tumor-induced orbital apex obstruction. *Knox et al*² explained this clinical manifestation as a unique form of the systemic carcinoid syndrome, and believe this presentation to be due to the spontaneous release of inflammatory mediators intrinsic to the orbital tumor.

Pathological examination, including imprint, frozen section and histology studied with immunohistochemistry, is invaluable in the diagnosis of orbital metastasis. Neither clinical signs nor radiological features are diagnostic for orbital carcinoid. Different radiopharmaceuticals such as [123] meta-iodobenzylguanidine and [111] In-DTPA-octreotide have been used for the identification of primary and secondary carcinoid tumors.^{19,20} However, no single radiopharmaceutical gives 100% sensitivity. Therefore only histopathological examination including histology and immunohistochemistry may provide a definitive diagnosis of carcinoid tumors of the orbit. The histopathological diagnosis is particularly important in the extremely rare cases of primary carcinoid tumor of the orbit, or in unusual cases where the identification of orbital carcinoid precedes the diagnosis of primary tumor.^{6,21}

Our patient metastasis to extraocular muscles was diagnosed with FNA. Although FNA has been reported in literature as a good tool for diagnosis of orbital or eye metastasis but we did not find any report about this tumor and it seems that it is the first report of diagnosis of carcinoid metastasis to orbit that has been diagnosed with the use of FNA.

Conclusion

This case was illustrative of several points; The orbital presentation of carcinoid tumor is rare. The atypical orbital presentation of the tumor in a patient with history of carcinoid tumor should alert the physician to metastasis.

Early biopsy rather than observation in such cases is essential for improved prognosis. We saw that FNA can be used as a good tool for diagnosis of orbital metastasis of carcinoid tumor.

References

- Wein FB, Perry JD, Miller NR, Brinker DA, et al. Pulmonary carcinoid tumor presenting with simultaneous orbital and intracranial metastases: value of transnasal endoscopic orbital biopsy and decompression. *Orbit* 1999 Dec; 18(4):267-272.
- Knox RJ, Gigantelli JW, Arthurs BP. Recurrent orbital inflammation from metastatic orbital carcinoid tumor. *Ophthal Plast Reconstr Surg*. 2001 Mar;17(2):137-9.
- Borota OC, Kloster R, Lindal S. Carcinoid tumour metastatic to the orbit with infiltration to the extraocular orbital muscle. *APMIS*. 2005 Feb;113(2):135-9.
- Goldberg RA, Rootman J, Cline RA. Tumors metastatic to the orbit: a changing picture. *Surv Ophthalmol*. 1990 Jul-Aug;35(1):1-24.
- Riddle PJ, Font RL, Zimmerman LE. Carcinoid tumors of the eye and orbit: a clinicopathologic study of 15 cases, with histochemical and electron microscopic observations. *Hum Pathol*. 1982 May;13(5):459-69.
- Fan JT, Buettner H, Bartley GB, Bolling JP. Clinical features and treatment of seven patients with carcinoid tumor metastatic to the eye and orbit. *Am J Ophthalmol*. 1995 Feb;119(2):211-8.
- Shields CL, Shields JA, Eagle RC Jr, Peyster RG, et al. Orbital metastasis from a carcinoid tumor. Computed tomography, magnetic resonance imaging, and electron microscopic findings. *Arch Ophthalmol*. 1987 Jul;105(7):968-71.
- Hanson MW, Schneider AM, Enterline DS, Feldman JM, et al. Iodine-131-metaiodobenzylguanidine uptake in metastatic carcinoid tumor to the orbit. *J Nucl Med*. 1998 Apr;39(4):647-50.
- Jensen RT, Doherty GM. Carcinoid tumors and the carcinoid syndrome, in *Cancer: Principles & Practice of Oncology*, V.T. De Vita, S. Hellman, and S.A. Rosenberg, Editors. 2001, Lippincott Williams & Wilkins: Philadelphia, PA.1813-1833.
- Ferry AP, Font RL. Carcinoma metastatic to the eye and orbit. I. A clinicopathologic study of 227 cases. *Arch Ophthalmol*. 1974 Oct;92(4):276-86.
- Shetlar DJ, Font RL, Ordonez N, el-Naggar A, et al. A clinicopathologic study of three carcinoid tumors metastatic to the orbit. Immunohistochemical, ultrastructural, and DNA flow cytometric studies. *Ophthalmology*. 1990 Feb;97(2):257-64.
- Couch DA, O'Halloran HS, Hainsworth KM, Cowen DE, et al. Carcinoid metastasis to extraocular muscles: case reports and review of the literature. *Orbit*. 2000 Dec;19(4):263-269.
- Rush JA, Waller RR, Campbell RJ. Orbital carcinoid tumor metastatic from the colon. *Am J Ophthalmol*. 1980 May;89(5):636-40.
- Krohel GB, Perry S, Hepler RS. Acute hypertension with orbital carcinoid tumor. *Arch Ophthalmol*. 1982 Jan;100(1):106-8.
- Harris AL, Montgomery A. Orbital carcinoid tumor. *Am J Ophthalmol*. 1980 Dec;90(6):875-7.
- Khaw P, Ball D, Duchesne G. Carcinoid tumour of the orbital muscles: A rare occurrence. *Australas Radiol*. 2001 May;45(2):179-81.
- Skibell BC, Patrinely JR, Font RL. Metastatic carcinoid presenting as orbital cellulitis. *Ophthalmic Pract* 1998;16:88-90.
- Wong VG, Melmon KL. Ophthalmic manifestations of the carcinoid flush. *N Engl J Med*. 1967 Aug 24;277(8):406-9.
- Isidori AM, Kaltsas G, Frajese V, Kola B, et al. Ocular metastases secondary to carcinoid tumors: the utility of imaging with [(123)I]meta-iodobenzylguanidine and [(111)In]DTPA pentetretotide. *J Clin Endocrinol Metab*. 2002 Apr;87(4):1627-33.
- Carnaille B, Nocaudie M, Pattou F, Huglo D, et al. Scintiscans and carcinoid tumors. *Surgery*. 1994 Dec;116(6):1118-21; discussion 1121-2.
- Zimmerman LE, Stangl R, Riddle PJ. Primary carcinoid tumor of the orbit. A clinicopathologic study with histochemical and electron microscopic observations. *Arch Ophthalmol*. 1983 Sep;101(9):1395-8.