

Case Report

Presentation of Metastatic Cholangiocellular Cancer with Orbital Metastasis

Metastatik Kolanjiöselüler Kanserin Orbital Metastaz ile Prezantasyonu

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ABSTRACT

Cholangiocarcinomas are cancers arising from epithelial cells of the intrahepatic and extrahepatic bile ducts. Most are locally advanced lesions when present. They usually present with jaundice, abdominal pain, and abnormal liver biochemical tests.

Proptosis developed in the right eye of a 49-year-old female patient who was diagnosed with carcinoma metastasis in the right femur and we examined for the primary one. Lacrimal gland biopsy of the patient was found to be compatible with cholangiocarcinoma metastasis. Neuroophthalmologic complications in cholangiocarcinoma are very rare and 10 cases have been reported in the literature.

Keywords: orbital metastasis, CNS metastasis, cholangiocarcinoma, exophthalmos

ÖZET

Kolanjiokarsinomlar intrahepatik ve ekstrahepatik safra kanallarının epitel hücrelerinden kaynaklanan kanserlerdir. Çoğu prezente olduğunda lokal olarak ilerlemiş lezyonlardır. Genellikle sarılık, abdominal ağrı ve anormal karaciğer biyokimyasal testleri ile prezente olurlar.

Sağ femurda karsinom metastazı saptanıp primerine yönelik tetkik ettiğimiz 49 yaşındaki kadın hastanın sağ gözünde proptozis gelişti. Hastanın lakrimal bez biyopsisi kolanjiokarsinom metastazı ile uyumlu saptandı. Kolanjiokarsinomda nörooftalmolojik komplikasyonlar oldukça nadir olup literatürde 10 vaka bildirmiştir.

Anahtar Kelimeler: orbital metastaz, SSS metastazı, kolanjiokarsinom, ekzoftalmus

Introduction

Metastaticorbital masses make up less than 5% of orbital tumors [1]. Most frequent metastatic cancers of the orbital are breast (52%), prostate (12%) and lung cancers (8%) [2]. Only ten cases of neuro-ophthalmologic and ocular presentations of cholangio-

carcinoma have been reported in the literature [3-6].

Case Presentation

A 49-year-old female patient who admitted to orthopedics with low back and hip pain underwent a biopsy of the right femoral proximal after the bilateral T1A and T2A

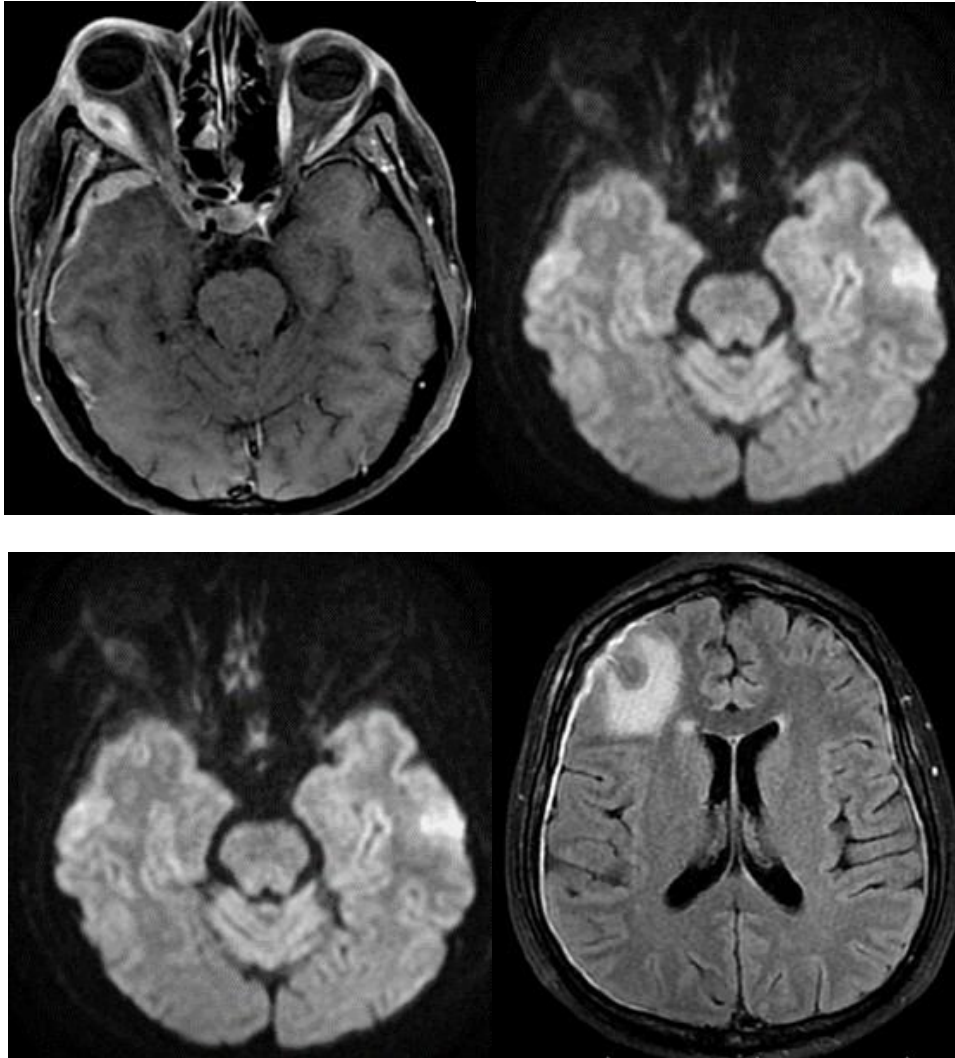


Figure 1a. In contrast T1 axial images, mass lesion in the right lateral rectus muscle, dural thickening and contrast involvement in the adjacent intracranial space and contrasting in subcutaneous plans are observed. 1b. Diffusion b 1000 MR images do not show diffusion limitation in the mass in the lateral rectus. 1c. In contrast axial FLAIR (c) and T1 weighted images (d), bilateral dural involvement as well as parenchymal extension and cerebral edema are seen to develop in the right frontal adjacent to dural involvement. 1d. In contrast axial FLAIR (c) and T1 weighted images (d), bilateral dural involvement as well as parenchymal extension and cerebral edema are seen to develop in the right frontal adjacent to dural involvement.

images in the patient's hip MRI showed diffused lesions suspected of metastasis on bone surfaces. As a result of the biopsy which revealed carcinoma metastasis, the patient admitted to the oncology outpatient clinic. In the computer tomography scans of the patient multiple metastatic mass lesions of 24x11 mm in size were detected in the liver parenchyma at the level of segment 7. A biopsy of the liver

was performed on the patient. While waiting for the biopsy result, proptosis and a limitation of eye movement developed in the right eye of the patient. An orbital MR was performed on the patient. The patient's cranial and orbital MRI images are shown in figure 1a-1b-1c-1d. In the orbital and brain MRI images before and after contrast, soft tissue mass lesions in the lateral vicinity of the right orbita in the

right lateral rectus muscle which erase extraconal periorbital fat plans, with their central showing necrotic heterogeneous contrast involvement were observed. Exophthalmos was present in the ipsilateral glob. Pathologic contrast involvement was also seen in the subcutaneous soft tissue muscle plans in orbital neighborhood. In addition, irregular dural thickening and contrast were observed in the intracranial area adjacent to the right orbit, which was evaluated as dural involvement. In T1 and FLAIR images after contrast, dural involvement was observed not only in orbital proximity but also in the opposite cerebral hemisphere. Diffusion MRI images showed no restriction of diffusion in the lesions. With these findings, the lesions were primarily considered in favor of metastatic involvements. But in MRI images, because of ventilation losses in the right paranasal sinuses compatible with sinusitis and rapid progression of the patient's MRI findings, it was thought in the differential diagnosis, that the infection should be excluded and biopsy from the lacrimal gland was performed on the patient. While waiting for the biopsy result, empirical vancomycin, meropenem and amphotericin B were initiated.

The result of the liver biopsy of the patient was reported as cholangiocellular carcinoma.

The result of biopsy from the lacrimal gland was consistent with cholangiocellular carcinoma metastasis. Radiotherapy was planned for the eye and the whole brain of the patient. After radiotherapy, it was decided to start gemcitabine and cisplatin.

Discussion

The presentation of our case was not compatible with typical presentation of cholangiocarcinoma. Initially bone metastases were detected and metastasis of orbital and central nervous system developed during the examination process. CNS metastasis and orbital metastasis are rather rare in cholangiocarcinoma [7-9]. There are 26 CNS metastasis cases in the literature. The brain was reported as the site of metastasis in 1.6% of patients with stage IV intrahepatic cholangiocarcinoma in a 2010-2015 review of US population. Orbital metastasis is also very rare and our case is the eleventh case in the literature. One case of a clival mass and sixth cranial nerve palsy, one case of metastasis to the medial rectus muscle and diplopia, two cases of metastasis to the occipital lobe and homonymous hemianopia, and one case of a hypercoagulable state-related stroke and homonymous hemianopia make up the five previously reported neuro-ophthalmologic presentations of cholangiocarcinoma [3]. A combination hepatocellular carcinoma/ cholangiocarcinoma that metastasized to the retina and vitreous was reported in one case, and there have also been two instances of cholangiocarcinoma that metastasized to the orbit and caused eye pain [5]. While, a case presenting with skin and orbital metastasis was reported in 2020 in Japan [10], in the same year metastatic cholangiocarcinoma presenting with 6th cranial nerve paralysis was reported from Miami [6].

REFERENCES

1. Babu A, Babu G, Sajeed A. Orbital Metastasis as The Initial Manifestation of Small

Cell Lung Cancer-A Case Report. Journal of Cancer Research & Therapeutics. 2017; 13.

2. Şerifoğlu İ, Bilgin M, Özdemir H. Metastatik küçük hücre dışı akciğer kanserinin nadir formu: orbital kitle. Düzce Üniversitesi Sağlık Bilimleri Enstitüsü Dergisi. 2011; 1(3): 35-7.

3. Rico G, Smith S, Siddiqui Y, Whyte A, Gombos D, Lee A. Neuro-ophthalmologic manifestations of cholangiocarcinoma: A caseseries. *Eye*. 2017; 31(8): 1245-8.
4. Fujimoto K, Kuroda J-i, Makino K, Hasegawa Y, Kuratsu J-i. Skull metastasis from intra hepatic cholangiocarcinoma: report of 3 cases and review of the literature. *Neurologia medico-chirurgica*. 2013; 53(10): 717-21.
5. Praidou A, Jacob S, Irion L, Sivaraj R, Groenewald C, Coupland SE, et al. Retinal and vitreous metastases from hepatocholangiocarcinoma. *BMC cancer*. 2017; 17(1): 1-3.
6. Fowler BJ, Lam BL. Sixth Cranial Nerve Palsy as the Presenting Sign of Metastatic Cholangiocarcinoma. *International medical case reports journal*. 2020; 13: 667.
7. Novegno F, Umana G, Granaroli P, Borri F, Orlandi A, Lunardi P. Current management of central nervous system metastasis from cholangiocarcinoma: the neurosurgical perspective. *Literature review. British Journal of Neurosurgery*. 2020; 34(5): 575-83.
8. Altay T, Krisht KM, Couldwell WT. Sellar and parasellar metastatic tumors. *International Journal of Surgical Oncology*. 2012; 2012.
9. Tan SK, Luther E, Eichberg D, Shah A, Khan K, Jamshidi A, et al. Complete regression of a solitary cholangiocarcinoma brain metastasis following laser interstitial thermal therapy. *World neurosurgery*. 2020; 144: 94-8.
10. Kawabe K, Urabe K, Momosaki S. Cutaneous and Orbital Metastases of Intrahepatic Cholangiocellular Carcinoma. *Clinical Gastroenterology and Hepatology*. 2020; 18(9): A23-A4.

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