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Metastatic breast carcinoma involving the optic disc

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ABSTRACT

Purpose: To report a rare case of unilateral optic nerve metastasis without choroidal involvement in a patient with known metastatic carcinoma of the breast.

Observations: A 65-year old female with a history of metastatic breast carcinoma presented to our clinic with a one-month history of blurred vision and floaters of the left eye. Fundoscopy of the left eye revealed a flesh colored nodule with white lobules demonstrating an endophytic growth pattern from the optic nerve. Magnetic resonance imaging of the brain and orbits revealed multiple intraparenchymal enhancing lesions, leptomeningeal enhancement, and enhancement of the left optic disc and proximal optic nerve all consistent with metastatic disease. The diffuse cranial disease was treated with whole brain radiation leading to regression of the optic nerve lesion and stabilization of visual acuity.

Conclusions and Importance: Intraocular metastases are the most common malignant tumor of the eye. Prompt identification and treatment of intraocular metastases is vital to prevent severe visual loss. Here, we document an unusual case of optic disc metastasis with unique fundoscopic features.

1. Introduction

Intraocular metastasis is the most common type of malignant tumor in the eye. However, metastatic disease to the optic disc accounts for only 4.5% of intraocular metastases.[1] These tumors are usually associated with a contiguous choroid lesion. Metastases to the optic disc without regional choroidal involvement disease are far less common, and therefore clinicians must remain vigilant for systemic disease when patients present with abnormal optic nerve lesions.[1] Herein, we present a rare case of unilateral optic disc metastasis without choroidal involvement in a patient with known metastatic carcinoma of the breast.

2. Case report

A 65-year-old African-American female presented to our clinic for evaluation of a one-month history of blurred vision and increased floaters in her left eye. Her past medical history was significant for estrogen and progesterone receptor mutation positive, Her2 mutation negative, invasive mammary carcinoma diagnosed four years prior to presentation. She had undergone mastectomy followed by anti-estrogen chemotherapy. One year prior to presentation, she was found to have lung and bone metastases that were treated with palliative radiation. Her ocular history was significant for remote retinal detachment in the right eye that was repaired by a scleral buckling procedure. On presentation, visual acuity of counting fingers at 3 feet in her right eye and 20/50 in the left. The anterior segment was unremarkable except for mild nuclear cataracts in both eyes. Fundus biomicroscopy revealed an attached retina with full scleral buckle and peripheral chorioretinal scarring in the right eye, consistent with her known history of retinal detachment. In the left eye, a flesh colored nodule with white lobules demonstrating an endophytic growth pattern was seen on the optic nerve (Fig. 1a). A broad area of hypopigmentation with pigment clumping was observed nasal to the fovea without evidence of elevation or mass lesion. The optic nerve lesion exhibited no hyperautofluorescence (Fig. 1b). Spectral domain optical coherence tomography (SD-OCT) of the left eye revealed an elevated disc lesion with a nodular appearance, a single cystoid space nasal to the fovea, and nodular excrescences at the level of the RPE and mild increased reflectivity of the choroid in the area nasal to the optic disc (Fig. 2a). Due to our high index of suspicion for metastatic disease, an MRI of the brain and orbits with and without contrast was obtained urgently. Post-contrast imaging identified multiple intraparenchymal enhancing lesions, nodular enhancement of the cerebellum, and linear enhancement in the subarachnoid space of the bilateral frontal lobes consistent with diffuse intracranial metastasis. Subarachnoid spread to the left optic nerve sheath was observed with enhancement of the left proximal optic nerve

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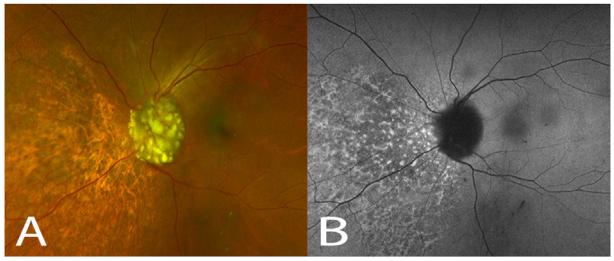

Fig. 1. A) Initial color fundus photograph of the left eye demonstrating a lobular, fleshy mass of the optic nerve head. An area of hypopigmentation with pigment clumping can be observed nasal to the fovea without the appearance of elevation or a mass lesion. B) Fundus autofluorescence imaging shows a hypoautofluorescent optic nerve head lesion. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

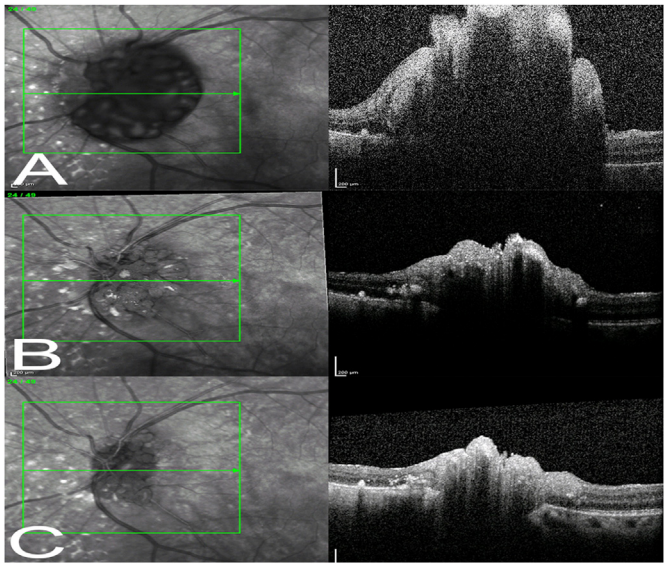

Fig. 2. A) Spectral domain optical coherence tomography (SD-OCT) of the left eye at presentation revealing an elevated, nodular lesion of the optic disc. Nodular excrescences at the level of the RPE and mild increased reflectivity of the choroid in the area nasal to the optic disc are present, however there is no thickening of the choroid that would suggest choroidal metastasis. Subsequent SD-OCT images taken six weeks (B) and fourteen weeks (C) after stereotactic radiation demonstrate a reduction in size of the optic disc lesion.

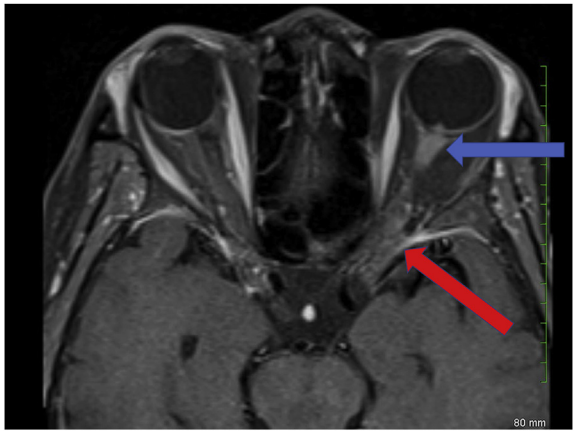

Fig. 3. Post-contrast MRI of the brain and orbits taken within a week of initial presentation demonstrating subarachnoid spread of tumor in the bifrontal regions and left optic sheath (red arrow) as well as proximal enhancement of the left optic nerve and optic disc (blue arrow).

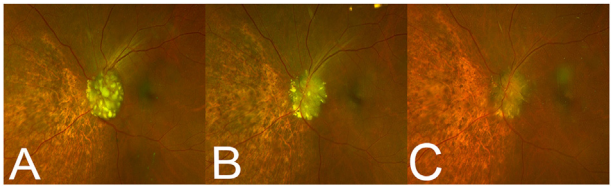

Fig. 4. Color fundus photographs of the left eye on presentation (A), and then at six weeks (B) and fourteen weeks (C) after stereotactic radiation demonstrating progressive reduction in size of the disc lesion, mild optic atrophy and calcium deposition. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

and optic disc (Fig. 3). Diagnostic lumbar puncture was not performed as the patient had elected to receive palliative measures only. After discussion with her oncology team, a plan was made for treatment of the brain and optic nerve lesions with stereotactic radiation. Follow-up color fundus photos taken six and fourteen weeks after therapy revealed significant regression in size of the lesion (Fig. 4b–c) and follow-up SD-OCT imaging demonstrated reduction in size of the optic disc lesion (Fig. 2b–c). The patient's visual acuity remained stable at 20/50 three months after therapy.

3. Discussion

Ocular metastases have an estimated incidence of 20,000 cases per year.[2] Most intraocular metastases occur in the uveal tract, with > 90% involving the posterior aspect of the choroid.[3] In comparison, optic disc metastases account for only 4.5% of intraocular metastases.[1] Breast carcinoma has been identified as the most common primary source of optic disc metastases (43%), followed by lung carcinoma (27%).[1]

Clinically, optic nerve metastases tend to present with unilateral loss of visual acuity as the main symptom.[4] Orbital pain and exophthalmos are also possible ocular manifestations.[5] The appearance of optic disc metastases on fundoscopic exam varies widely. In a comprehensive review by Shields et al.,[1] 16% of optic disc metastases presented as a distinct optic nerve head nodule, similar to our patient, while the remainder appeared as a diffuse thickening of the optic disc. Most masses were located centrally on the optic disc, and most were yellow or white in color, with only 16% sharing the fleshy-pink color of the mass observed in our case. Flame-shaped disk hemorrhages were found to be present in 42% of affected eyes. Fluorescein angiography may show hyperfluorescence of the mass in the late venous phase.[1]

Optic disc metastases are generally the result of direct spread from juxtapapillary choroidal metastasis or hematogenous spread via the posterior ciliary arteries supplying the optic nerve head.[4] An estimated 74% of metastases to the optic disc are contiguous with a choroidal metastasis due to the rich vascular supply in the choroid of the posterior pole[1]. In our case, no thickening of the choroid was observed that would suggest choroidal infiltration contiguous with the optic nerve lesion. Mild increased reflectivity of the choroid was noted on OCT imaging, however this most likely represented resolved subretinal fluid exudation.

Given the presence of subarachnoid spread of tumor in the bifrontal regions, extension into the left optic nerve sheath on MRI and enhancement of the optic disc and proximal left optic nerve, the optic disc metastasis in this case is more likely due to direct infiltration of the optic nerve by leptomeningeal carcinomatosis (LC). Leptomeningeal metastases may occur via hematogenous seeding of the meninges from a distant source or by direct subarachnoid invasion by a primary or metastatic intraparenchymal brain tumor.[6] An estimated 5% of all patients with breast cancer develop LC.[7] The diagnosis of LC is most often made by the combination of enhancing subarachnoid mass lesions on radiologic imaging and positive CSF cytology. However, in patients with known systemic disease and radiologic findings highly suggestive

of LC, such as our patient with known metastatic breast cancer, intraparenchymal brain metastases and multiple enhancing masses within the subarachnoid space on post-contrast MRI, imaging alone can be virtually diagnostic for LC.[6] Vision loss is the most common ocular symptom of LC, occurring in up to 44% of cases, and may present as the initial complaint.[8,9] The mechanism by which visual loss occurs in LC is not clear, but current theories include optic nerve sheath cuffing, vascular compromise, and neoplastic infiltration of the optic nerve.[9–11] Distant hematogenous dissemination of malignant cells from the primary source to the optic disc is also a possibility, but less likely given the radiologic evidence of LC involving the optic nerve sheath.

An optic nerve metastasis can easily be mistaken for another lesion on funduscopic examination. Retinal astrocytic hamartomas have a very similar appearance to the lesion in our case. They may appear as a raised multilobulated “mulberry” lesion but typically exhibit a characteristic hyperautofluorescence. Optic disc drusen can mimic this presentation if the calcific bodies are robust in number or demonstrate extrusion. However, disc drusen tend to have an eccentric location without disc swelling.

Treatment for leptomeningeal carcinomatosis is usually palliative and may include intrathecal chemotherapy or local radiation therapy for symptomatic management.[6] In regard to ocular metastases, fractionated external beam radiation of 35–40 gray is the first line choice for local treatment.[1,4] Tumor response to radiotherapy is variable, with 50% of patients reporting stabilization of vision and 36% reporting improvement in visual acuity.[12] Unfortunately, the prognosis for patients with leptomeningeal carcinomatosis is very poor, with a median survival of duration of 2–3 months.[13]

4. Conclusions

Optic disc metastasis is a rare presentation of metastatic carcinoma and may present a diagnostic challenge due to its heterogeneity on fundoscopic exam and nonspecific appearance on radiographic imaging. Clinicians should have a high index of suspicion for ocular metastases in patients with visual complaints and a past history of cancer, particularly breast or lung carcinoma. Prompt recognition can facilitate the initiation of local and systemic therapy, prevent severe vision loss and improve overall prognosis.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

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

Declaration of competing interest

None.

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