Case Report

Proptosis: A Rare Sole Presentation of Metastatic Disease

Abstract

Orbital metastasis is identified clinically in very few cases having metastatic disease. We report a case of 62-year-old female having breast cancer for 4 years with sudden proptosis. She was further investigated to have multiple metastases. As these patients have dismal prognosis, the treatment can only be palliative. This case report impresses upon the need of thorough systemic evaluation in a known patient of cancer with proptosis because it can be the sole presentation of potentially alarming multiple metastases.

Keywords: *Breast cancer, metastasis, proptosis*

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Introduction

Orbital metastasis is rare, accounting for only 1%–13% of all orbital tumors. In about 12%–31% of the affected patients with breast cancer, eye metastases are the first sign of metastatic spread.[1] Once the diagnosis has been established, treatment is mainly palliative focusing on symptomatic relief and improvement of orbital function as long as possible.[2] In these patients, extensive orbital surgery to remove metastasis is not recommended as this is not curative and leads to significant ocular morbidity.[3] Although orbital metastasis has poor prognosis and treatment is usually palliative, an ophthalmologist still plays a significant role in the detection and management of these lesions. This case report impresses upon the need of careful assessment of the eye complaints and detailed ocular and systemic evaluation in patients with previous history of cancer considering metastatic spread.

Case Report

We report a case of a 62-year-old female who came to the eye outpatient department with complaints of pain and protrusion of the right eye for 3 weeks and sudden loss of vision in the right eye for 1 week. The pain was sudden in onset, moderate to severe in intensity accompanied by progressive protrusion of the right eye. The patient also gave a history of nausea and loss of appetite for the past 4 years. Her history and old records revealed that she had been diagnosed with duct cell carcinoma right breast (stage T2N1M0) 4 years back.

On ocular examination, the patient denied perception of light in the right eye and the visual acuity of the left eye was 6/9. In the right eye, on inspection, there was lid retraction of approximately 3 mm, severe conjunctival chemosis, and axial proptosis (which was appreciated better on Naffziger's view with no change in size on Valsalva maneuver). On palpation the proptosis was irreducible with resistance to retropulsion and there was no thrill. Corneal sensations were normal with no infraorbital or supraorbital anesthesia. There was no preauricular lymphadenopathy. There was no bruit on auscultation. The ocular movements were restricted in all gazes. The right eye pupil showed relative afferent pupillary defect. On slit-lamp examination, the anterior segment was normal. The fundus examination revealed central retinal artery occlusion (CRAO) with sparing of cilioretinal artery [Figure 1]. The anterior and posterior segments of the left eye were within normal limits [Figure 2].

Her blood investigations showed raised glucose 211 mg/dl (70–110 mg/dl) and raised aspartate aminotransferase 42 μ/l (8–37 μ/l) with normal thyroid function test. On peripheral blood smear, there was lymphogranulocytosis.

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Magnetic resonance imaging (MRI) brain with orbits were planned which showed altered signal intensity lesion of size 27 mm × 22 mm in the right orbit, mainly intraconal region involving medial rectus and optic nerve leading to proptosis [Figure 3]. Similar enhancing lesion of the size 17 mm × 16 mm was seen in the head of caudate nucleus and left basifrontal region showing signal enhancement. Contrast-enhanced computed tomography (CECT) chest showed multiple small isodense lesions in the parenchyma suggestive of secondaries. Right breast showed spiculated lesion of size 14 mm × 18 mm with a nodule of size 8 mm × 10 mm. On CECT abdomen there was a hypodense lesion in the head and uncinate process of pancreas with multiple hepatic metastasis. There were peritoneal deposits in perirenal and hepatorenal region. Subcutaneous and intramuscular deposits were also seen [Figure 4]. The fine-needle aspiration cytology (FNAC) from the right breast revealed pleomorphic cells with hyperchromatic and pleomorphic nuclei with increased mitotic figures in solid pattern, suggestive of duct cell carcinoma. FNAC of subcutaneous lesion showed metastatic secondaries in the left shoulder and left cervical and right gluteal region with immunohistochemistry positive for estrogen receptor, progesterone receptor, and human epidermal growth factor receptor-2/neu receptors.

The poor visual prognosis due to CRAO was explained to the patient. The patient was planned lateral canthotomy for orbital compartment syndrome, but she refused for any surgical intervention.

A combined chemotherapy treatment with day 1 injection cyclophosphamide 800 mg intravenously (IV), injection adriamycin 90 mg IV, and injection fluorouracil 75 mg IV and day 2 injection granulocyte colony-stimulating factor subcutaneously and tablet mammazole 2.5 mg OD was started. In addition, the orbital mass was irradiated with a total dose of 30 Gy in 10 fractions over 2 weeks, which led to a significant reduction in the symptoms of the patient.

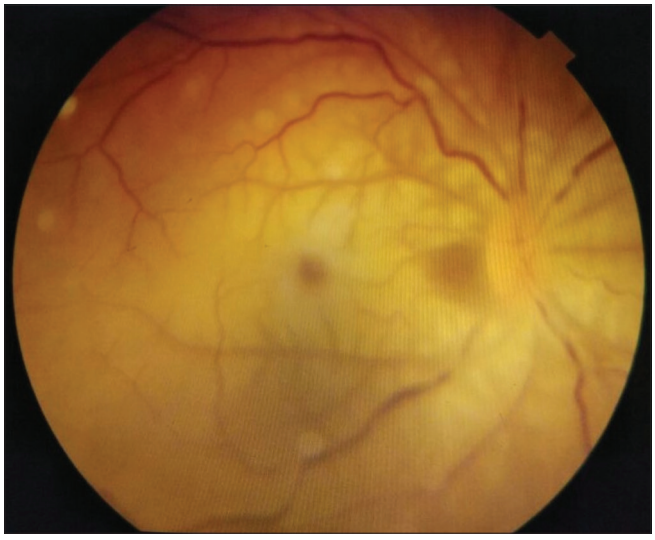

Figure 1: The fundus photograph of right eye showing central retinal artery occlusion with sparing of cilioretinal artery

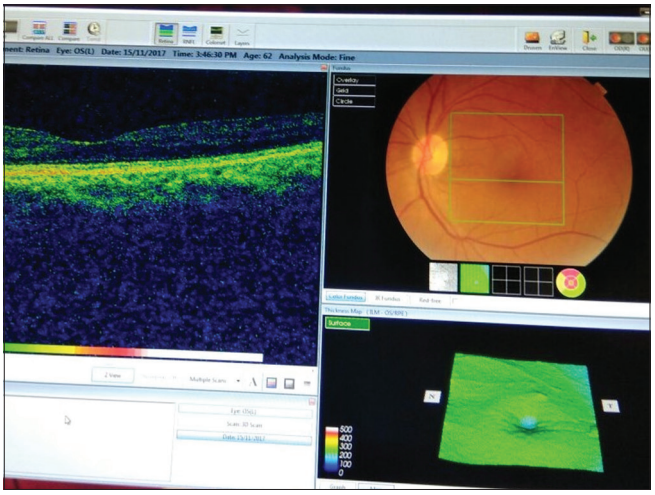

Figure 2: Optical coherence tomography image of the left eye showing normal macula and posterior pole

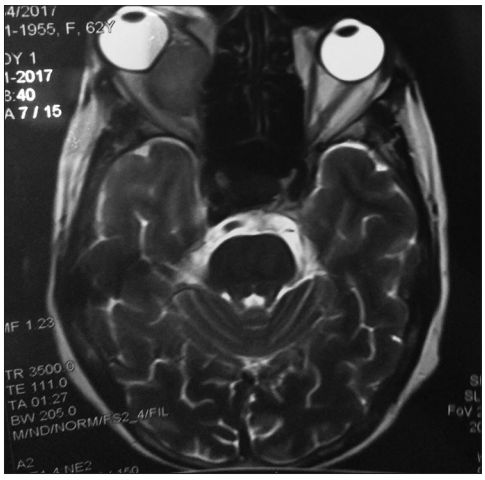

Figure 3: T2-weighted axial image showing well-defined hypointense mass in the right intraconal space causing proptosis

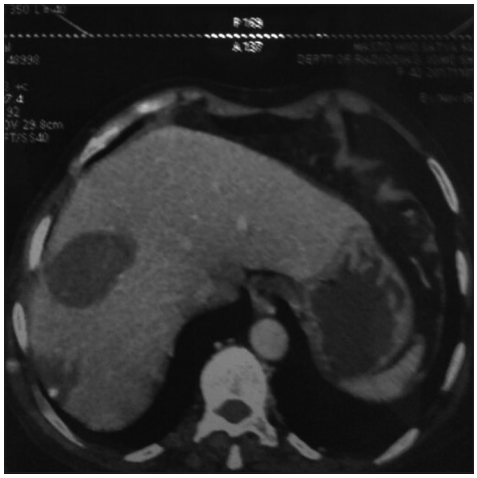

Figure 4: T2-weighted axial image showing liver secondary

Discussion

Orbital metastases are of rare occurrence accounting for 1%–13%.[4] Breast carcinoma is reported to be the most common cause of orbital metastasis ranging from 28.6% to 51% followed by lung (6%–21%), prostate (13%–17%), gastrointestinal (6%–7.1%), kidney (3%), and skin (melanoma) cancers (3%).[5-7] The most common symptoms of orbital involvement in metastases are diplopia (48%), pain (42%), visual loss (30%), and a palpable mass. Proptosis is the most common presenting sign accounting for 63%–67.9% of cases followed by motility disturbance (57.1%), strabismus, and visual loss. Motility disturbance and diplopia are due to either direct muscle invasion or mass effect of the tumor.[3] The perineural invasion of the malignancy explains the pain in these patients. In the present case, the patient presented with axial proptosis due to intraconal tumor growth and sudden vision loss due to CRAO. There were relative afferent pupillary defect due to CRAO and also the infiltration of optic nerve by the tumor.

The management of these patients involves imaging studies such as whole-body positron emission tomographic scan or CT scan/MRI whenever metastatic disease is suspected. In CT in most cases, soft tissue attenuating material is identified, located in the extraconal compartment with variable morphology ranging from well-defined rounded lesions to diffusely infiltrating lesions with general enhancement.[8] Bony destruction may be seen. MRI appearances are similar to CT, but it has greater contrast resolution with T1-weighted images isointense to muscle and T2-weighted images hyperintense to muscle and hypointense to fat. Although the diagnosis is established by FNAC of the lesion or biopsy wherever possible, it may be avoided if there is a strong clinical and imaging suspicion for metastatic disease. It should only be done in patients with no previous history of cancer and in patients with orbit as the only site of suspected metastasis in whom definite diagnosis would change the overall management.[9]

The patients with orbital metastasis have poor prognosis with the mean survival of 31 months (ranging 1–116 months).[10] The treatment of orbital metastases is generally palliative. In few selected patients, it may involve radiotherapy, systemic chemotherapy, hormonal therapy, or surgery. The external beam radiotherapy is applied to control tumor growth, restore visual function, and decrease mass size, thereby reducing proptosis and exposure keratopathy. Orbital surgery to remove the metastasis is not recommended generally as this is not curative and may be associated with significant ocular morbidity.

The patients with known primary cancer having ocular complaints should undergo complete ocular and systemic evaluation for orbital metastasis and multiple subclinical secondaries. Although these patients have limited survival, they can be provided a better quality of life by palliation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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