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Global Journal of Health Sciences and Research

Case Report

Secondaries from retromolar trigone carcinoma leading to orbital apex disorders

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Received : 28 November 2022
Accepted : 30 December 2022
Published : 20 February 2023

DOI
10.25259/GJHSR_11_2022

Quick Response Code:

ABSTRACT

Orbital apex disorders (OADs) are uncommon and they include – orbital apex syndrome (OAS), superior orbital fissure syndrome (SOFS), and cavernous sinus syndrome (CSS). They all share similar etiologies, diagnostic evaluation, and management strategies. These syndromes can occur isolated or combined – where SOFS progresses and develops into OAS or CSS. Combined presentation has been reported with infective etiologies, but no literature is available for neoplasms. OAD secondary to head and neck cancer is exceptionally rare. A 44-year-old male post left hemi-mandibulectomy with modified neck dissection, radiotherapy, and chemotherapy for left retromolar trigone carcinoma presented with complaints of inability to open left eye (LE) and defective vision in the same eye. On examination of LE, visual acuity was 6/24 NIP with complete ptosis, extraocular movements were restricted in all gaze, corneal sensation was diminished, and pupil was 5 mm and not reacting to light. LE fundus examination showed blurring of disk margins. The primary diagnosis was made as OAS. Contrast enhanced-MRI neck showed features suggestive of recurrence with intracranial extend involving left cavernous sinus, orbital apex, and left orbit. Later considering the extend of spread in MRI and the clinical presentation, diagnosis was made to be OAD. The patient was given third cycle of chemotherapy – injection paclitaxel, injection cisplatin, and injection fluorouracil, along with injection dexamethasone. The patient was provided temporary eye crutch glasses and asked to review after 3 weeks for repeat assessment. Although individual syndromes of OAD are rare, they are closely related entity. Hence, it is important to know that SOFS can progress to OAS causing visual dysfunction or to CSS, leading to systemic ailments. Or else, they can present as combined syndromes also.

Keywords: Orbital apex disorders, Orbital apex syndrome, Superior orbital fissure syndrome, Cavernous sinus thrombosis, Head and neck malignancy

INTRODUCTION

Orbital apex disorders (OADs) are uncommon and they include – orbital apex syndrome (OAS), superior orbital fissure syndrome (SOFS), and cavernous sinus syndrome (CSS). They all share similar etiologies, diagnostic evaluation, and management strategies. These syndromes can occur isolated or combined, where SOFS progresses and develops into OAS or CSS.[1] Combined presentation has been reported with infective etiologies, but no literature is available for neoplasms.[2] OAD secondary to head and neck cancer is exceptionally rare.[3]

CASE REPORT

A 44-year-old male patient diagnosed with retromolar trigone carcinoma in April 2022 following which he underwent left hemi-mandibulectomy with modified neck dissection, radiotherapy,

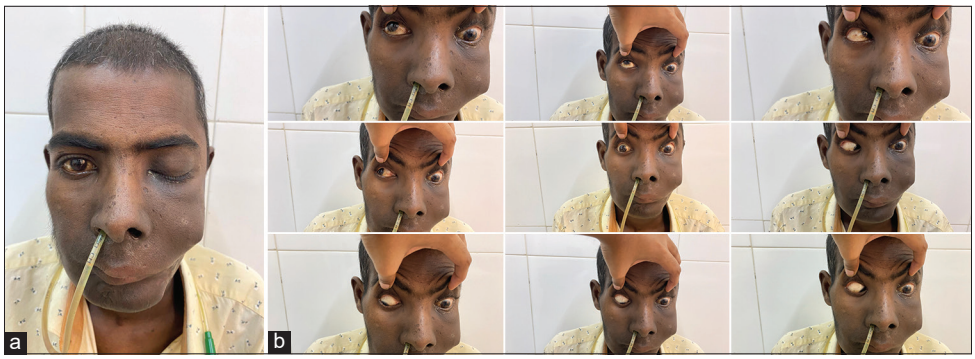

Figure 1: (a) Left eye complete ptosis. (b) Restriction of extraocular movements in all gaze in the left eye.

and two cycles of chemotherapy in May and from second cycle, the patient was started on concurrent chemotherapy and radiotherapy in June 2022. In October 2022, the patient presented to ophthalmology OPD with complaints of inability to open left eye (LE) and defective vision in the same eye for 5 days, which was not associated with pain. The patient gave no history of trauma, spectacle usage, or ocular surgery. He was not a known case of diabetes mellitus, systemic hypertension, tuberculosis, acquired immunodeficiency disorder, or thyroid disorder. He was not an alcoholic. History of tobacco chewing and smoking for past 15 years. On examination, the patient was poorly built and nourished, well oriented to time, place, and person. Visual acuity was 6/6 in the right eye and 6/24 NIP in LE. Examination of LE showed complete ptosis [Figure 1a], painless restriction of extraocular movements in all gaze [Figure 1b], diminished corneal sensation, and pupil was 5 mm and not reacting to light. LE fundus examination showed clear media and blurring of disk margins with dull foveal reflex in macula [Figure 2]. Sensations were also diminished in the left forehead, left cheek, and left upper and lower lid. Right eye examination was within normal limits. Both eye intraocular pressures were 16 mmHg, measured with non-contact tonometer. The primary diagnosis was made as OAS. Contrast enhanced-MRI brain showed features suggestive of recurrence with intracranial extend involving left cavernous sinus, orbital apex, and left orbit [Figure 3a-c]. Later considering the extent of spread in MRI and the clinical presentation, diagnosis was made to be OAD and was sort for oncology opinion. The patient had eastern cooperative oncology group score 2 and was started on third cycle of chemotherapy; Injection paclitaxel 200 mg (intravenous – IV) OD for 1 day, injection cisplatin 100 mg (IV) OD for 1 day, Injection dexamethasone 4 mg (IV) BD for 5 days, injection palonosetron 0.25 mg (IV) OD for 1 day, injection 5-fluorouracil 700 mg (IV) OD for 4 days, Injection ondansetron 4 mg (IV) TDS for 4 days, and injection pantoprazole 40 mg (IV) OD for 5 days. Following which the patient was given tablet dexamethasone 4 mg twice a day for 3 days and has been prescribed eye crutch glasses. The patient is asked to review after 3 weeks for repeat assessment.

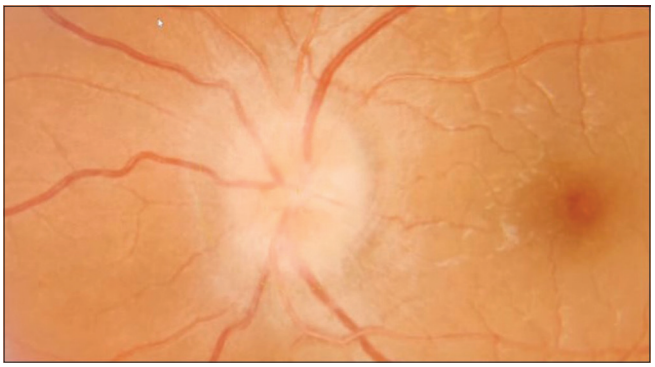

Figure 2: OS Fundus picture shows blurring of disc margins and dull foveal reflex in macula.

DISCUSSION

Orbital apex is the posterior part of orbit, where the four orbital walls converge. OAS is also known as Jacod syndrome characterized by ophthalmoplegia; proptosis; ptosis; palsy of III, IV, and VI cranial nerves; hypoesthesia of ipsilateral forehead, upper lid, and cornea (V1-division of trigeminal nerve); and visual deficit.[1]

Superior orbital fissure is a bony cleft between the roof and lateral wall at the orbital apex. SOFS is also known as Rochon-Duvigneaud syndrome includes all signs of OAS except optic nerve involvement.[1]

Cavernous sinus is located on either side of pituitary fossa and body of sphenoid bone. CSS includes all signs of OAS except optic nerve involvement and, in addition, has hypoesthesia of cheek and lower lid also (V2-division of trigeminal nerve). CSS may present with Horner's syndrome, if sympathetic chain adjacent to the cavernous segment of the internal carotid artery is involved.[1]

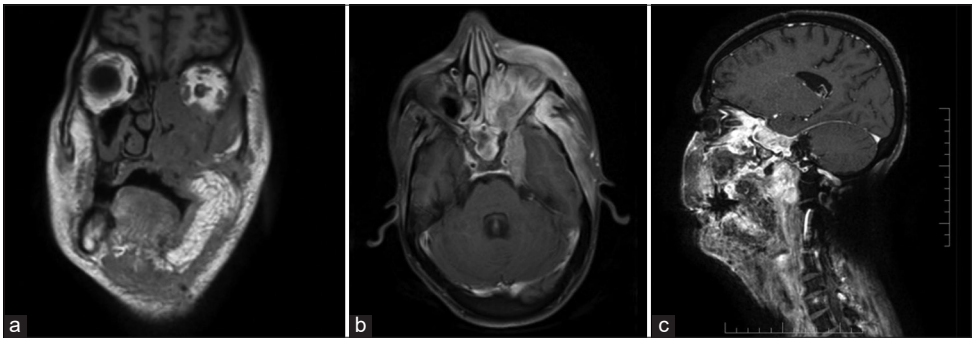

Figure 3: (a) CE-MRI NECK – An ill-defined T2/STIR high signal intensity lesion noted involving the flap region showing no significant restriction of diffusion and no blooming in gradient sequences. (b) CE-MRI NECK – An ill-defined high signal intensity lesion appears to extend into the left orbital apex and left cavernous sinus with left intraconal extension. (c) CE-MRI NECK – Ill-defined high signal intensity lesion appears to encase the optic nerve within the optic canal and intraconal region.

Various etiologies for OAD are infective, inflammatory process, neoplasms, trauma, iatrogenic, and vascular disorders. Management is toward the causative process. Therefore, in our case, the treatment is directed toward the underlying buccal mucosa cancer.[1]

Most common etiology among all is neoplasm followed by infection.[4] The incidence of intracranial metastasis from head and neck carcinoma is only 0.4%. Intracranial metastasis is rarely a clinical diagnosis in patients with head and neck squamous carcinoma.[5]

Failure to diagnose and treat these conditions will result in permanent disability to the patient. Although OAD are not commonly seen in the daily practice of maxillofacial surgeon, it is important to recognize it and sort a multidisciplinary approach with involvement of ophthalmologist, oncologist, otolaryngologist, and neurosurgeon. Thereby ensuring a timely recognition and treatment, to provide the best possible outcome.[6]

Along with the systemic treatment under oncology, the patient was given temporary crutch glasses to provide binocular vision. If ptosis does not improve post systemic treatment, surgical correction of ptosis must be done. Visual improvement will depend on the course of the disease process.

CONCLUSION

Knowledge regarding head and neck secondaries causing orbital apex disorders will result in early diagnosis and optimal patient care.

Acknowledgment

I would like to acknowledge the department of neurology, department of oncology, and department of radiology for their expertise and assistance throughout all aspects of our study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Jayamurthy P, Devi R, Nallamuthu P, Angalan V. Secondaries from retromolar trigone carcinoma leading to orbital apex disorders. Glob J Health Sci Res 2023;1:48-50.