IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 20, Issue 4 Ser.10 (April. 2021), PP 60-63
www.iosrjournals.org

A Case Report of Unilateral Eccentric Proptosis secondary to Maxillary Osteosarcoma

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Abstract: *Proptosis, the forward protrusion of the eyeball, is a common manifestation of a wide variety of diseases inside the orbit and its spaces. Its diagnosis is usually a combined effort of the ophthalmologist, otolaryngologist, neurosurgeon, and radiologist. This is a case of 24 years old male patient who presented to the OPD with gradually progressive proptosis and diminished of left eye in the last three months, associated with left sided nasal block and diffuse swelling over the left cheek. A detailed ophthalmic clinical examination was done including Exophthalmometry. ENT examination revealed the swelling to be bony growth of maxillary bone, biopsy of which was taken and tissue sent for histopathological examination. The diagnosis of Osteosarcoma of left maxillary bone was established from the histopathology and immunohistochemistry reports, supported by radiological reports. Case was referred to higher centre for Left maxillectomy, orbital exenteration and post-operative radiotherapy.*

Key words: *Proptosis, Osteosarcoma, Exophthalmometry, Orbit, Maxilla*

Date of Submission: 14-04-2021 Date of Acceptance: 28-04-2021

I. Introduction

Proptosis is defined as the forward protrusion of eyeball/displacement of globe relative to the orbital rims. The causes of Unilateral proptosis are innumerable and is usually a multidisciplinary problem requiring collaboration of ophthalmology with various branches. The eye is a major crossroad for all the structures around it which help in its support and functioning, which when affected extends into the orbit causing proptosis. It can be the most dramatic of the orbital symptoms, especially if it has an acute onset. A clear knowledge of the aetiologies will help the ophthalmologist to suspect, diagnose early, and provide treatment.

II. Case Report

History: A 24 years old male patient presented to the OPD with chief complaints of protrusion of left eye and diminution of vision in left eye in the last three months. There is history of gradually progressive protrusion of left eyeball and gradual painless progressive diminution of vision in left eye for the last three months, associated with left sided nasal block and diffuse swelling over the left cheek. Patient is also k/c/o seizure disorder, on anti-epileptic drugs.

Ocular examination:

BCVA in RE is 6/6, LE is CF at 1mt

Left eye examination: Eccentric proptosis seen, deviated in upward and outward direction. Ocular movements restricted in all gazes. Lid edema +, Conjunctival congestion+, Chemosis+, Exposure keratitis seen in inferior part of Cornea, AC- normal, Pupil – round, reacting to light (figure 1)

RE examination: within normal limits

Exophthalmometry was done, values are - RE-21mm, and LE-25mm (figure 2)

Dilated fundus examination of both eyes done, and showed normal fundus study.

ENT examination revealed the swelling to be bony growth of maxillary bone, biopsy of which was taken and tissue sent for histopathological examination.

HPE Report: Sheets of spindle cells with focal atypia, stroma shows lace like osteoid tissue. Suggestive of Osteosarcoma of left maxillary bone.

Immunohistochemistry report: KI67-50%, SATB2-Positive, P53-Positive, S100-Negetive. Suggestive of Osteosarcoma.

Radiological investigations:

CT brain with orbits report – expansile lytic lesion with ground glass appearance noted in floor of left orbit in maxillary base, with fibrous dysplasia.

MRI orbits report – mass involving left maxilla, showing intermediate signal on T1 and low signal on T2 and STIR, with extraosseous soft tissue component extending superiorly into left orbit not reaching the orbital apex, anteriorly into subcutaneous soft tissues, medial extension to medial wall of maxillary sinus and inferiorly into left alveolar margins at the level of molars. Left eyeball, extraocular muscles and optic nerve are normal (figure 3)

Figure 1:

Figure 2:

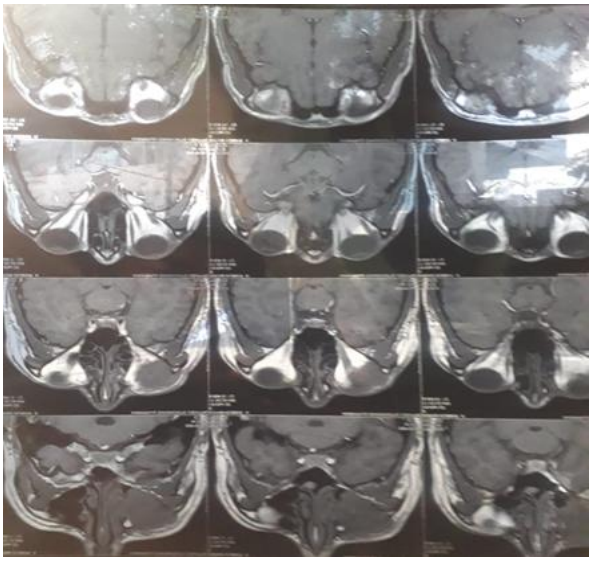

Figure 3:

Diagnosis: With the above clinical findings, supported by histopathology, immunohistochemistry and radiological reports, this is diagnosed to be a case of Left eye Eccentric proptosis with exposure keratitis, secondary to osteosarcoma of left maxillary bone.

Treatment: General measures – topical lubricants, antibiotics and vitamin supplementation given. Lid taping advised. Case was referred to higher center for Left maxillectomy, orbital exenteration and radiotherapy.

III. Discussion

Orbital pathology usually presents as proptosis. Symptoms reflect the orbital volume increase. Direction indicates the site of lesion. The causes of adult unilateral proptosis may be a retrobulbar haematoma following trauma, inflammatory conditions like orbital cellulitis, an orbital abscess, usually following frontal or ethmoid sinusitis, a pseudotumor of the orbit due to a granuloma of unknown cause, an epidermoid or dermoid cyst, a mixed lacrimal tumour (lacrimal adenoma), or a haemangioma. Malignant tumours include malignant melanoma, carcinoma of the maxillary or ethmoidal sinuses invading the orbit, and meningioma of the sphenoid. Thyroid eye disease which is usually bilateral also can present as a unilateral proptosis in its initial stages. Primary tumours of the orbit are usually mixed tumours of the lacrimal gland and dermoid cysts. Anterior temporal lobe lesions into the orbit can lead to proptosis and blindness. The causes of unilateral proptosis in a child include retinoblastoma in the first 5 years of life and infective orbital cellulitis.

The direction of exophthalmos may indicate the likely aetiology and site of lesion. Axial proptosis is seen in tumours arising within the muscle cone like optic nerve glioma. The eyeball is displaced down and/or lateral in diseases of frontal or ethmoid sinuses. Lacrimal gland or temporal fossa tumours have a medial displacement.

To evaluate and treat the patient with unilateral proptosis, an ophthalmologist must work closely with the ENT surgeon, neurosurgeon, and radiologist to ensure a successful outcome in each case. During the past few decades, advances in diagnostic instrumentation and surgical technique have helped to elevate the orbit to an anatomical area of great clinical interest. CT, MRI, and orbital echography have dramatically improved diagnostic accuracy and allowed a more careful therapeutic planning.

As regards the management, inflammatory cases and benign orbital neoplasms were most amenable to satisfactory treatment, medical or surgical, while malignant primary tumours if detected early could be eradicated with fair chances of success.

IV. Conclusion

Unilateral proptosis is a multidisciplinary problem and requires collaboration of different specialties of an ophthalmologist along with an otorhinolaryngologist, neurosurgeon, oncologist, and radiotherapist. A thorough ENT examination is mandatory in proptosis. A small number of cases can never go noticed, but in proptosis, however small the bulge, malignancy has to be ruled out.

CT and MRI scans was valuable in evaluating a case of proptosis, but histopathological examination provides a definitive diagnosis of the exact aetiology.

Early diagnosis of the underlying cause and prompt intervention will help in protecting the eyeball and preserving the visual function.

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Dr.M.Nirmala M.S, et. al. "A Case Report of Unilateral Eccentric Proptosis secondary to Maxillary Osteosarcoma."*IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 20(04), 2021, pp. 60-63.