Case Report

Olfactory neuroblastoma presenting as Non axial proptosis - A rare case report

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Abstract

Olfactory Neuroblastoma (ONB) is a rare neuroectodermal nasal tumour arising from the olfactory epithelium that is normally found in the upper part of the nasal cavity which frequently invades the orbit, cranial base and cranial vault. ONB accounts for approximately 2% of all sinonasal tract tumours with an incidence of 0.4 per million population. ONB has a bimodal age distribution between 11 and 20 years and between 51 and 60 years. The diagnosis may be delayed for several months due to its slow-growing nature. But in some cases may progress rapidly and aggressively. Here we describe a case of olfactory neuroblastoma in a 16-year-old male presented to our ophthalmology department with left-sided non-axial proptosis of 30-degree exotropia with nasal blockade on the same side.

Key Words: Olfactory neuroblastoma, Homer Wright Pseudorosettes, Flexner Wintersteiner rosettes, Proptosis, External ophthalmoplegia.

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INTRODUCTION

Olfactory Neuroblastoma (ONB) is a rare neuroepithelial tumour that arises from the olfactory epithelium that is normally present in the upper nasal cavity including the roof of the nose, cribriform plate of the ethmoid, superior nasal concha, the upper part of the septum. First described in 1924 by Berger it has a histological pattern similar to that of sympathetic ganglia, retina, and adrenal medulla and only recently became recognized as a distinct pathologic entity probably as a result of immunohistochemistry and employing electron microscopy techniques. ONB accounts for 1 to 5% of malignant neoplasms of the nasal cavity. The symptoms are related to site and invasion of the tumour. It's a locally aggressive malignancy that may invade orbit via local destruction of lamina papyracea. Orbital invasion can lead to vision loss, ophthalmoplegia and proptosis. The staging system based on tumour extension presented by Kadish *et al*. in 1976 has been widely accepted. The treatment of choice is a multidisciplinary craniofacial surgical resection followed by radiotherapy/ chemotherapy that has improved the prognosis considerably. In a large series, including 38 cases of ONB, Rakes *et al*. reported that 53% of patients had orbital or ocular symptoms, and the most common symptoms were periorbital pain and lacrimation. In this study, we present the clinical features of 16 years old boy with ONB showing orbital invasion.

CASE REPORT

A 16-year-old male presented to the Ophthalmology department with a complaint of forward and outward protrusion of left eyeball associated with pain for ten days. All ocular movements were restricted except elevation and depression in the left eye.

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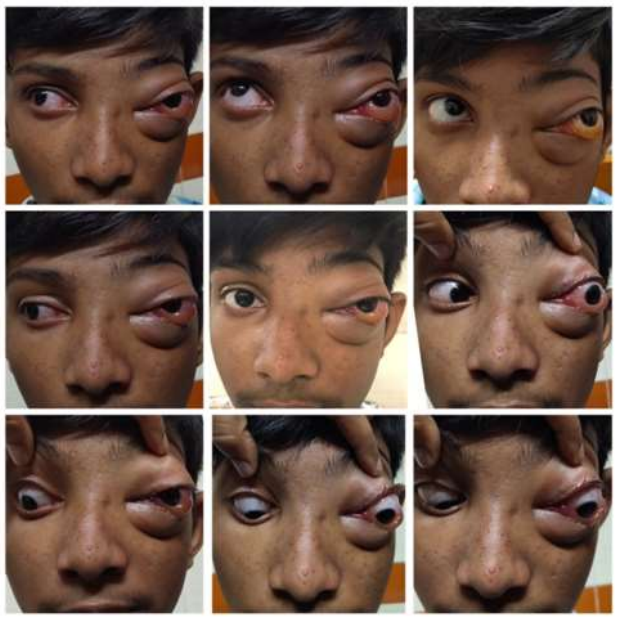
	OD	OS
UCVA	6/24	6/60
BCVA	6/6	6/18
CV	21/21	21/21
Lids and adnexa	Normal	Peri orbital oedema + Chemosis of lids +
Conjunctiva	Quiet	Extreme chemosis +
Cornea	Clear	Exposure keratopathy +
Anterior chamber	Well-formed, quiet	Well-formed, quiet
Pupil reflex direct and indirect	Normal, no RAPD	Normal, no RAPD
Iris	NCNP	NCNP
Lens	Clear	Clear
IOP		
Schiotz tonometry	17.3mmHg	20.6mmHg
Confrontation test	Normal	Normal
Fundus	Normal	Normal

On proptosis evaluation non-axial proptosis of LE with orbital mass occupying the medial orbital space mostly, pushing the globe outwards. Exophthalmometry reads RE 21mm, LE 27 mm with a baseline of 113mm. Compressive optic neuropathy was not present in our case in spite of huge tumour mass, probably due to sparing of intraconal space partially. So also diplopia was not a presenting symptom. He also complains of left-sided nasal blockage and over the past ten days associated with headache. A nasal exam was performed using rigid endoscopy, which revealed a large mass, medial to the middle turbinate in the left nasal cavity. The mass extended through the choana, extending into the nasopharynx clinically. It was attached superiorly with no medial or lateral attachment identified. The lesion was pink and lobulated with a rubbery, non-friable texture. Patient also had left-sided enlarged non-tender cervical lymph nodes of size 3x3cm.



STAGING

ENB spread to cervical lymph nodes is common, typically spreading to level II nodes, with frequent involvement of level I, level III, and retropharyngeal nodal groups at later stages. Locoregional and distant metastasis occurs in upto 38% of patients. Late recurrences or metastatic disease can occur up to two decades after the initial presentation. staging system that has been proposed in 1976 by Kadish *et al*. is still in use for staging, even though Dulgeuerov and Calcaterra have proposed TNM classification or ONB Kadish classification 4,5,7,8 is used for staging and recognizes three stages:

Stage A - tumours that are localized to the nasal cavity

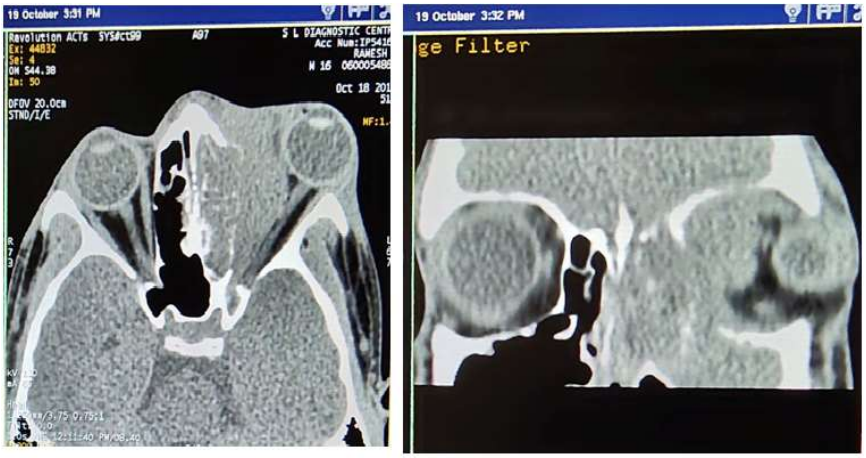
stage B - nasal cavity and paranasal sinuses

stage C - extension beyond the sinonasal cavities, including intracranial and orbital involvement.

MANAGEMENT

1. Haematological examination - was within normal limits.
2. Radio imaging

CT evaluation of nasal cavity, PNS and orbits revealed a mass lesion almost completely filling the left nasal cavity. The lesion is a hyperdense mass filling the left nasal cavity causing focal erosion of left lamina papyracea with extension into the extraconal compartment of the left orbit. The lesion caused focal erosion of part of the nasal septum. Superiorly, lesion caused erosion of the left cribriform plate. Laterally, the lesion extended into left maxillary antrum with blockage and expansion of osteomeatal complex.

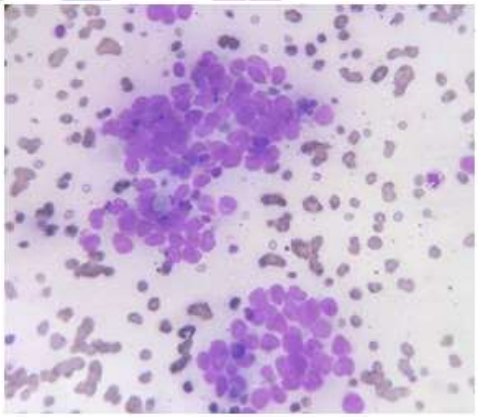


Lesion eroding the medial wall of left orbit with extension into the extraconal compartment of left orbit.
The lesion was causing focal erosion of part of the nasal septum. Superiorly eroding the left cribriform plate. Laterally extending into left maxillary antrum with blockage and expansion of osteomeatal complex.

3. Immunohistochemistry

Positive for Chromogranin ,CD 56 , Neuron specific enolasse, S-100 protein

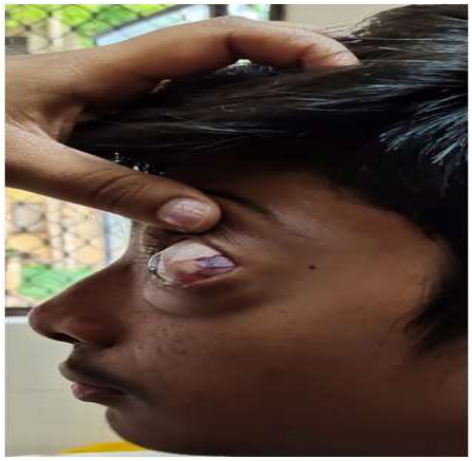
4. . Histopathological examination of lymph node (FNAC)



smears highly cellular and consists of pseudorosettes, clusters and discretely placed small round cells exhibiting moderate anisonucleosis

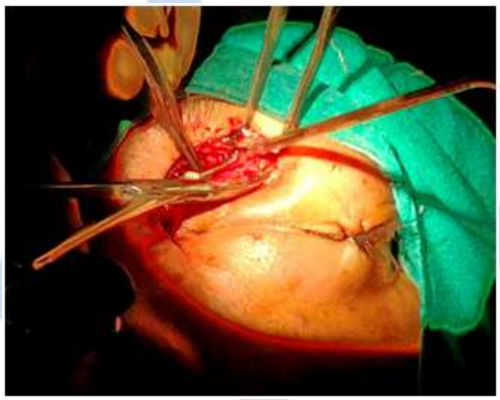
TREATMENT

Medical treatment: was given for ten days.
Inj Ceftriaxone IV BD
Inj DECADRON 2cc IV BD
Cyclopentolate e/d 2t/d
Lubricating e/d 6t/d
Day 3 of treatment
Lid oedema and chemosis of conjunctiva decreased, dilated conjunctival vessels noted



Surgical management

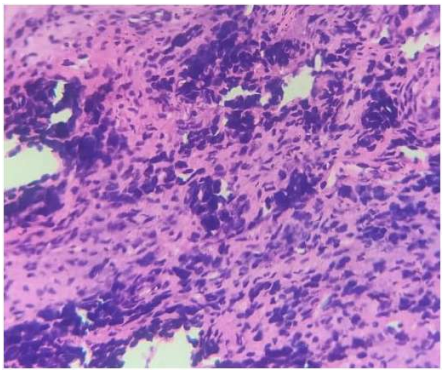
Medial orbitotomy combined with endoscopic resection of mass was performed with the help of an ENT surgeon as an initial procedure and samples were sent for histopathological examination. Adjuvant radiotherapy was given to patient because combined surgery and irradiation results in disease-free status6,8 in 92% of cases as per literature.



Intraoperative picture of mass removal by medial orbitotomy approach(Lynch incision)

Presently patient is under radiotherapy, and he will be reviewed regularly for recurrence and followup of visual function.

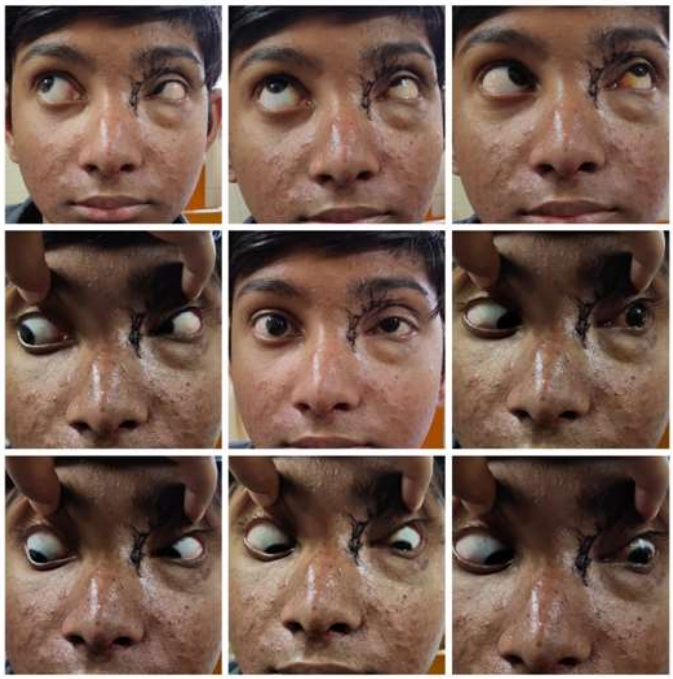
Histopathological examination report



Section showing tumour tissue composed of round to oval cells with mild to moderate pleomorphism exhibiting scant cytoplasm and darkly stained nuclei, at places arranged as nests and lobules in fibrillary matrix.at places, Homer wright rosettes were seen.

Features in favour of Olfactory Neuroblastoma(ONB)

Post Op Visual prognosis		
	OD	OS
BCVA	6/6	6/6
CV	21	21
Fundus	Normal	Normal



CONCLUSION

ONB is a rare endonasal tumour with orbital invasion. It may manifest with initial manifestations of external ophthalmoplegia, proptosis, either with or without compressive optic neuropathy. Therefore the rare aetiology of ONB must be considered in the differential diagnosis of non-axial proptosis associated with cranial nerve palsy and nasal signs. Since ONB is a locally aggressive tumour, the evaluation and management is a multidisciplinary approach as orbital invasion significantly associated with adverse outcomes.

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