A case of primary orbital neuroblastoma

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ABSTRACT

Introduction: Neuroblastoma is a pediatric neoplasm that is most common cancer diagnosed during infancy. Male to female ratio is 1:1:1 showing slight preponderance towards males. Neuroblastoma occurs primarily in the abdomen in 60% cases but in 8% cases the tumour arises in the orbit where it arises from ciliary ganglion. Seventy-five percent cases occur before the age of four years. Case Report: We describe a case of five year old boy who presented with a rapidly progressing large tumour in his left orbit. CECT revealed a mass in the left orbital region pushing the eyeball anteriorly. A histopathologic diagnosis of neuroblastoma was made. Subsequent medical evaluation including chest X-ray, USG of the abdomen, whole body computerized tomography and bone scintigraphy showed no evidence of systemic involvement or metastasis. Conclusion: Neuroblastoma although an uncommon primary orbital tumour in children but it should be considered in the differential diagnosis of orbital tumours in children.

Keywords: Primary, Orbital, Neuroblastoma, Child

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INTRODUCTION

Neuroblastoma is an undifferentiated malignant tumour of the primitive neuroblasts. Neuroblastoma is a pediatric neoplasm that is most common cancer diagnosed during infancy [1, 2]. Male to female ratio is 1:1:1 showing slight preponderance towards males [3]. Neuroblastoma occurs primarily in abdomen in 60% cases but in 8% cases the tumour arises in the orbit where it arises from ciliary ganglion. Seventy-five percent of cases occur before the age of four years. Primary orbital neuroblastoma has been reported previous in adults [2] and neonates [4].

CASE REPORT

A 5-year-old boy was brought to the Outpatient Department of our hospital with complaints of painless, rapidly progressing protrusion of the left eye since five months which was associated with loss of vision. The mass between the lids was red and firm in appearance and impeded any view of his left globe (figure 1).

Patient was physically normal and had no signs of congenital malformation. There were gross restriction in movement of proptosed mass in all positions of gaze.
Symblepharon was present. No, other details could be
seen. The lids were oedematous and ecchymosed. The
mass was non-tender and globular. It was immobile and
non pulsatile. It was not expansible on coughing or
crying. Right eye appeared normal.

**Investigations:** Complete hemogram, including
general blood picture, ESR, urine and stool examination
were found to be within normal limits.

Computed tomography (CT) scan showed soft tissue
density enhancing mass in the left retro-orbital region
measuring 3x2.7 cm pushing the left eye ball anteriorly.
The optic nerve could not be separated out and lesion
seemed to be adherent to the eye ball. There was no
evidence of mass in the sinuses, nasal cavities or intra-
cranial cavities (figure 2).

![Figure 1: View of the left eye man at presentation.](image1)

![Figure 2: Coronal CT images showing the left orbital mass.](image2)

![Figure 3: Histopathological analysis showing the Homer-
wright pseudorosettes and high mitotic activity (H&E, x200).](image3)

Ultrasoundography (USG) scan of the left eye ball was
performed using high frequency and showed a large
hypoechoic mass.

Fine needle aspiration cytology (FNAC) of the tissue
showed features suggestive of Neuroblastoma.

Excision biopsy on histopathological examination
showed tumour consisting of small round cells with high
mitotic activity in a neurofibrillary and haemorrhagic
background. The tumour cells were forming Homer –
Wright pseudorosettes. Some sites showed haemorrhagic
necrosis (figure 3). The tumour cells were positive for
synaptophysin, neuron specific enolase, chromogranin
and focially for glial fibrillary acidic protein. A
histopathological diagnosis of neuroblastoma was made.

Subsequent medical evaluation including chest X-
ray, USG of the abdomen, whole body CT scan and bone
scintigraphy showed no evidence of systemic involvement
or metastasis. Tumour was diagnosed as primary orbital
neuroblastoma. The whole orbital exenteration
operation was done under general anesthesia and the
patient further received 11 cycles of chemotherapy and
36 Gy local EBRT. Adequate treatment was given and
patient was further referred to pediatric oncology
department for further management.

**DISCUSSION**

Neuroblastoma is an undifferentiated malignant
tumour of primitive neuroblasts which may be
metastatic to the orbit. It represents second most
common orbital tumour in children after
rhabdomyosarcoma. It arises from the sympathetic
system and ganglia and represents the peripheral
nervous system counter part of retinoblastoma. Rarely
neuroblastosmas may represent primary lesions in the
orbit where they may arise from ciliary ganglion [5].

Only 8% cases first present with an orbital lesion; in
92% of cases the presence of extra orbital primary
tumour is already known [5]. Forty percent of orbital
lesions are bilateral. Mean age at presentation is two-
years-old [5]. 75% of cases occur before the age of four
years [5].
Commonly there is bone destruction particularly of lateral orbital wall. Ten to forty percent of systemic neuroblastomas result in orbital metastasis within three months after diagnosis. Forty percent of orbital metastasis are bilateral. Other possible ophthalmic manifestations of metastatic neuroblastomas are horner’s syndrome, papilledema, retinal striae, anisocoria, nystagmus and cranial nerve paralysis. About 90% of orbital lesions originate from abdomen [6, 7, 8].

Histopathologically other small round cell tumors especially non-hodgkin’s lymphomas and rhabdomyosarcomas must be taken into consideration for differential diagnosis of neuroblastoma [4].

The pathological features and immunohistochemistry certainly pointed out to the neuroblastoma in our case. Further-more the systemic examination ruled out any other primary lesion or metastatic foci pointing this to be the case of primary orbital neuroblastoma. The primary orbital neuroblastoma is definitely a rare case presentation. Till now very few cases of primary orbital neuroblastoma have been reported [1].

CONCLUSION

Neuroblastoma although an uncommon primary orbital tumour in children should be considered in the differential diagnosis of orbital tumours in children.

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Author Contributions

Rayees Ahmad Sofi – Substantial contribution to the conception and design, acquisition of data, analysis and interpretation of data, drafting the article, revising it critically for important intellectual content, final approval of data

Sajad Bashir Khanday – Substantial contribution to the conception and design, analysis and interpretation of data, drafting the article, revising it critically for important intellectual content, final approval of data

Manzoor Qadir Keng – Substantial contribution to the conception and design, revising it critically for important intellectual content and final approval of version to be published

Junaid Salam Wani – Analysis and interpretation of data, revising it critically for important intellectual content, final approval of version to be published

Ankur Goel – Acquisition of data, drafting the article, revising it critically for important intellectual content, final approval of data

Tufaila Shafi – Acquisition of data, revising it critically for important intellectual content, final approval of the version to be published

Guarantor

The corresponding author is the guarantor of Submission.

Conflict of Interest

The authors declare no conflict of interest.

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