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Rhabdomyosarcoma Left Trigeminal Nerve in a 12-year-old Male Child - A Case Report from Tertiary Cancer Centre from North India

Jyoti Sharma[1], Mrinalini Upadhyay[2]

[1,2]MBBS, MD Radiotherapy, MO Specialist, IGMC Shimla

Corresponding Author: Jyoti Sharma

ABSTRACT

Rhabdomyosarcoma (RMS) is a rare but aggressive malignancy of infants and children. Alveolar, embryonal, pleomorphic, and spindle/sclerosing rhabdomyosarcoma are the common types. Multimodality treatment[5] is almost always required. It Includes surgery, radiation therapy and chemotherapy. RMS were initially described by Weber in 1854.[6]

Keywords: Rhabdomyosarcoma, VAC, RT

INTRODUCTION

Rhabdomyosarcoma is a rare highly malignant soft tissue sarcoma that arises from mesoderm or myotome-derived skeletal muscle[1] and highly malignant soft tissue sarcoma with most frequently involved sites[2] are the orbit (9%) head and neck (excluding Para meningeal tumors) (7%) Para meningeal (25%) and rest genitourinary 31% and others

CASE REPORT

We report a case of 12yr old young adult who presented to tertiary cancer centre IGMC Shimla with complaint of swelling left side face, deviation of left eye and headache since past 3 months

Physical examination revealed normal HMF with signs of developmental delay. C.N.S examination revealed involvement of 2,3,4,5,6 cranial nerves on left side with presence of left sided ptosis.
MRI report revealed 5th nerve Schwannoma CE-MRI revealed well defined lesion in the course of left TG nerve 5.3x3.8x5.4 with temporal lobe extension.

Patient reported to us post op with these symptoms after undergoing Left FTOZ craniotomy with GTE at a higher centre. Detailed HPE revealed highly infiltrative cellular tumor arranged in islands large groups and central necrosis nests trabeculae surrounded by desmotrophic stroma. There was infiltration of adjacent soft tissue and skeletal muscles with no bone invasion revealed positivity for desmin, Myogenin, CD 56, Vimentin, and focal positivity for S-100.

F/S/O Rhabdomyosarcoma NOS. Child was evaluated by CECT brain after 1.5months post-op for residual disease which revealed heterogeneously enhancing mass left infratemporal fossa with pterygopalatine fossa involving mastication space with compression and displacement of IJV and L CCA
Metastatic workup revealed no mets elsewhere.

He was then started on Radiation therapy 36Gy/20# followed by boost 5.4 Gy/3# and was started on CCT based on VAC. After 3 cycles patient had symptomatic improvement.
Patient had episode of seizure at home and then died 8months after treatment.

DISCUSSION

RMS represents an aggressive tumor and age has been identified as an

independent predictor of prognosis, with children <1 year and >10 years having inferior survival.

Para meningeal RMS represents the majority of nonorbital head and neck RMS, and radiotherapy is essential for maximizing the chance of cure.

Data from the IRS studies show improved local control in patients with intracranial tumor extension when radiotherapy is started within 2 weeks of diagnosis,

In our set up, patient reported late after surgery leading to delayed RT. Moreover there was gross residual disease after surgery before commencement of RT and CCT which led to dismal prognosis.

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