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Radiation Oncology

PAINLESS UNILATERAL PROPTOSIS PRESENTING AS EWING'S SARCOMA- A RARE CASE REPORT

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ABSTRACT Ewing's sarcoma is a part of a spectrum of the Ewing family of soft tissue tumors that have a primitive neuro-ectodermal origin most commonly originating from the appendicular and axial skeleton. Occurrence in the orbit is a rare presentation. case report a 15-year-old girl presented with painless unilateral swelling of the right eye since 4 months on histopathological examination came out to be a small round cell tumor with primitive neuroendocrine origin. she was started on chemotherapy with VAC+IE regimen as induction chemotherapy but as the disease was progressive and not responding after 17 cycles of alternating VAC+IE second-line chemotherapy with irinotecan and temozolomide was started and after three cycles surgery was deferred i/v/o intracranial extension so she was treated with external beam radiotherapy 59.4gy in 33#'s at 1.8gy/# with concurrent irinotecan and temozolomide with complete response after radiotherapy. she was on follow-up for 4 months and was later diagnosed with secondary deposits in the lung which she could not cave in and expired.

KEYWORDS : Ewing's sarcoma, Orbit, Proptosis, Radiation.

INTRODUCTION

Ewing's sarcoma is a part of a spectrum of the Ewing family of soft tissue tumors that have a primitive neuro-ectodermal origin most commonly originating from the appendicular and axial skeleton. occurrence in the orbit is a rare presentation. The median age of presentation is 14 years.

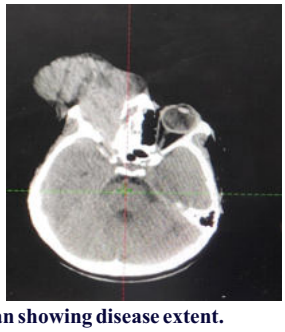

Figure 1 CT scan showing disease extent.

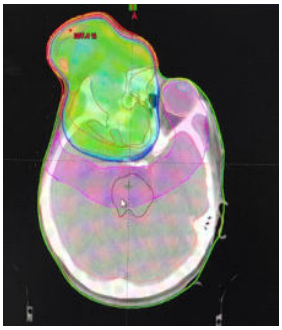

Figure 2. Figure showing the isodose distribution during rt planning.

CASE REPORT

A 15-year-old girl presented with painless unilateral swelling of the right eye since 4 months which on histopathological examination came out to be a small round cell tumor with primitive neuroendocrine origin. On immunohistochemistry with cd99 positive confirmed to be Ewing's sarcoma.

- CT scan shows lesion in the medial aspect and superior aspect of orbit involving eyelid 29*22mm with moderate proptosis.
- bone marrow aspiration was within normal limits.
- ophthalmic examination was normal

IHC PANEL

CD 99: POSITIVE
LCA: NEGATIVE
Tdt: NEGATIVE

she was started on chemotherapy with a VAC IE regimen as induction chemotherapy but as the disease was progressive and not responding after 17 cycles of alternating VAC IE second-line chemotherapy with irinotecan and temozolomide was started and after three cycles surgery was deferred i/v/o intracranial extension so she was treated with external beam radiotherapy 59.4gy in 33#'s at 1.8gy/# with concurrent irinotecan and temozolomide with complete response after radiotherapy.

she was on follow-up for 4 months and was later diagnosed with secondary deposits in the lung which she could not cave in and expired.

DISCUSSION:

Ewing's sarcoma family of tumors (Esfts) are malignant tumors of neuroectodermal origin. these include Ewing's sarcoma (bone 87%), extraosseous Ewing's sarcoma (8%), peripheral pnet (5%), and Askins tumor (PNET of the chest wall). they are small blue round cell tumors. markers that differentiate Ewing's sarcoma are vimentin, beta 2 Microglobulin, and hba-71. Esfts exhibit rapid progression and have a poor prognosis. it is assumed that most patients have a subclinical metastatic disease at presentation. the treatment plan includes initial cytoreduction with chemotherapy to eradicate micrometastatic disease and facilitate effective local tumor control measures, definitive radiation to eradicate all known disease, and consolidation therapy for eradication of the occult residual disease to reduce the likelihood of tumor recurrence. intracranial tumor extension, older age at presentation, and systemic metastasis are poor prognostic features in primary orbital Esfts.

CONCLUSION:

Ewing's sarcoma of the eye being a rare entity emphasizes the importance of immunohistochemistry staining in primitive cells to conclude the diagnosis.

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