ORIGINAL RESEARCH PAPER

Oncology/Radiotherapy

KEY WORDS: Orbital rhabdomyosarcoma, Proton-therapy, Radiotherapy, Brachytherapy.

ORBITAL RHABDOMYOSARCOMA- A CASE REPORT

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ABSTRACT

Introduction: Rhabdomyosarcoma is mesenchymal originated soft tissue tumours. It is commonest head and neck soft time Ca in childhood malignancy (1,2). It Constituted about 5% of all paediatric malignancy about 40% rhabdomyosarcoma occurs in head and neck area (3,4). Tumour arises from indifferent pluripotent Mesenchymal cells that can differentiated during embryonic period between skeletal muscles. Rarely it is caused by trauma. **Case Presentation:** 6-year female presented with proctoring Most in left eye for last 9 months, pain and watery discharge from left eye for last 3-4 months. Biopsy has been done which shown spindle cell carcinoma **Discussion:** The current European paediatric soft tissue sarcoma society recommended following treatment protocol. The treatment protocol consists of Proton-therapy, Radiotherapy and Brachytherapy. **Conclusion:** Orbital rhabdomyosarcoma in life threeling disease with high morbidity early diagnosis and treatment can save the life. Addition of radiotherapy improves survival and prognosis.

INTRODUCTION

Rhabdomyosarcoma is mesenchymal originated soft tissue tumours. It is commonest head and neck soft time Ca in childhood malignancy (1, 2). It Constituted about 5% of all paediatric malignancy about 40% rhabdomyosarcoma occurs in head and neck area (3, 4). Tumour arises from indifferent pluripotent Mesenchymal cells that can differentiated during embryonic period between skeletal muscles. Rarely it is caused by trauma (5). Histologically four types of rhabdomyosarcomata reported embryonal alveolar, pleomorphic and spindle cell or scalarising rhabdomyosarcoma. Embryonal rhabdomyosarcoma is commonest variant reported with good prognosis about 50-70% of orbital rhabdomyosarcoma are reported embryonal type. 2nd commonest is alveolar, pleomorphic and spindle cell are rare to be found in orbit (6, 7) 5-year survival rate reported about 84.3% (8,9) other factors responsible for survival were younger age <10 years, female sex (8,9,10,11,12)

Case Presentation

6-year female presented with proctoring Most in left eye for last 9 months, pain and watery discharge from left eye for last 3-4 months. MRI Report shown there is evidence of large lobulated exophytic altered signal intensity lesion is seen involving left orbit. Lesion encasing left eye globe and shows extension into overlying skin and subcutaneous tissue region. Lesion shows extension upto left orbital apex. Lesion shows multiple septations. It is measuring approx. 120 x 108mm - likely mitotic.

Biopsy has been done which shown spindle cell carcinoma biopsy report complete excisional and reconstruction with cervical flap done by ENT and ophthalmic surgeons histopathological report shown embryonal rhabdomyosarcoma base was involved by tumours, tumor extended around eyeball all eye ball structured spared.

Superior inferior nasal, inferior skin margin nasal skin, temporal skin resection margin were involved. On Histo-Chemistry examination confirmed as Embryonal rhabdomyosarcoma. Botryoid type. Desmin, Vimentin, Myogenin, Myo-01 were positive Ki-67 was 30-40%. Bone marrow examination was done which shown 2% plasma cells.X-Ray chest PA View and routine blood investigation were normal.

Fig-1 Pre-Operative Picture

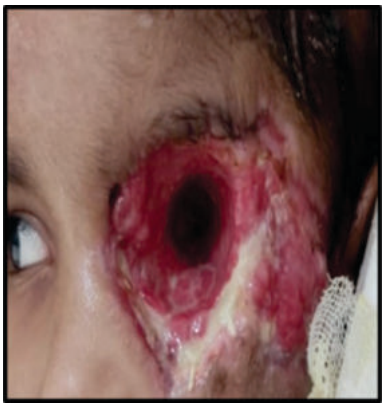


Fig-2 Post Operative Picture

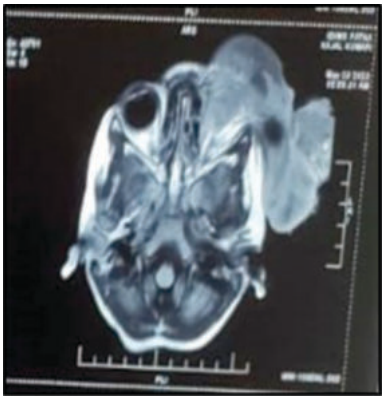


Fig-(i)

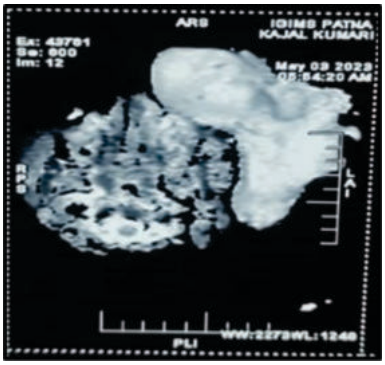


Fig-(ii)

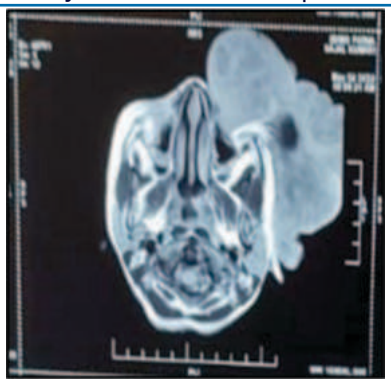


Fig-(iii)

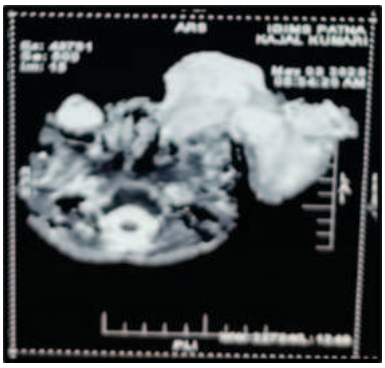
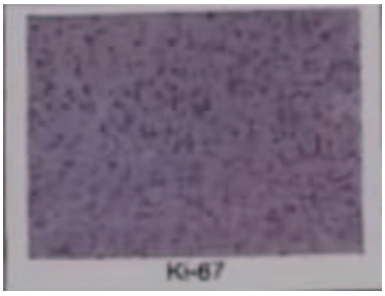
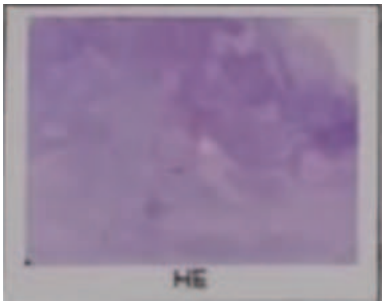


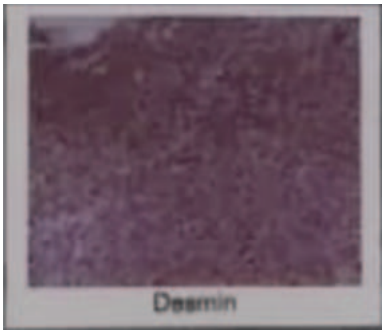
Fig-(iv)

Figures: CT Scan



Ki67- Immunoreactive in 30-40% of neoplastic cells.





Desmin- Immunoreactive, score 4+ in neoplastic cells.

Vimetin- Immunoreactive, score 4+ in neoplastic cells.

Table: Immuno Histo Chemistry Report shown orbital rhabdomyosarcoma

IHC MARKER(S)	RESULT
KI-67	Immunoreactive in 30-40% of neoplastic cells.
Desmin	Immunoreactive, score 4+ in neoplastic cells.
Myo-01	Weak immunoreactive, score 1+ in neoplastic cells.
Vimentin	Immunoreactive, score 4+ in neoplastic cells.
Myogenin	Weak immunoreactive, score 1+ in neoplastic cells.

DISCUSSION

The current European paediatric soft tissue sarcoma society recommended following treatment protocol (42,52)
Group-I Chemotherapy consisting of VA
Group-II & III Chemotherapy with VA

Complete Ifosfamide after complication of 3 cycle with remission
Add RT or Remove RT
Add more Ifosfamide or Remove Ifosfamide

Group-IV Chemotherapy IVA+ ..... followed by one year maintenance therapy and Radiotherapy.

Optimum available for consideration wolder et al. reported that (15, 22, 43, 57, 58) IMRT with fusion image achieved good local control and minimum dose to organ at risk (10, 36, 59).

Hein et al. also concluded that MRT resulted less dose to organ at risk as compare to 3DCRT (60)

Proton Therapy

Jack et al reported superior result by using fractionated proton radiotherapy for orbital RMS, it reduced doses to surrounding Structure (10, 36, 54, 61) disadvantage associated with proton therapy in paediatric age group is to develop second malignancies and growth retardation due to excellent survival.

10 years event free survival was better in a pooled analysis 306 patients of orbital rhabdomyosarcoma in comparison to those who did Receive radiotherapy (82 53%) without statical significant. (40)

Brenemor et al reported no effect on prognosis with reduced dose to radiotherapy and addiction of chemotherapy with moderate dose of cyclophosphamide (31).

Radiotherapy

Dose advise in these cases 36 Gy to 54 Gy over 4 to 5 weeks 3D conformal therapy. Intensity modulated radiotherapy. Prolong therapy and implant brachytherapy provide excellent result with minimal side effects. (15, 22, 43, 57, 58)

Brachytherapy

Brachytherapy delivers high dose to tumour area with law

dose to surrounding structure. Following SOP guideline. Blank et al reported use of brachytherapy in treatment of orbital rhabdomyosarcoma (54). We have planned her Chemotherapy 6 cycles Followed by radiotherapy 45 Gy in 36 fractions

Management

Main goal of treatment in these types of patients to prevent local area recurrence and systemic spread of disease (26,27). Now a days multi-disciplinary approach which include surgery, chemotherapy, Radio therapy is commonly preferred treatment (3). Zhang et al. shown that surgical excision is associated with 5.7 times more survival (30)

After Biopsy part internationally staging dose (44-46)
Group-I
Group-II
Group-III

Current management included surgery irradiation and chemotherapy depending on disease stage (47, 49)

Use abdomen shown internal echoes present in urinary bladder.

CONCLUSION

Orbital rhabdomyosarcoma in life threatening disease with high morbidity early diagnosis and treatment can save the life. Addition of radiotherapy improves survival and prognosis. A multi-disciplinary approach in the form of surgery chemotherapy and radiotherapy should be considered for improvement of survival. Embryonal rhabdomyosarcoma has good 5 years survival rate.

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