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Original Research Article

Pediatric Orbital Rhabdomyosarcoma: A Single Institute Retrospective Study

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ABSTRACT

Background: Orbital rhabdomyosarcoma is the most common orbital malignancy of childhood. Orbit is the primary site in approximately 10% cases. Orbit is the most favorable site with a 5 year survival of 85%.Outcome in Indian patients is largely unknown, as only few series are reported. **Methods**: A total of 11 children with orbital RMS were evaluated retrospectively. Treatment of it involved a multimodal approach, using radiotherapy and chemotherapy (based on the IRS-III) protocol. Progression during therapy and relapse were considered as events. **Results**: Of the 11 patients diagnosed during the study period there were 6 male and 5 female patients. The median age was 7 years (age range 1 yrs to 14 yrs). Commonest presentation was proptosis with diffuse involvement of the orbit. The commonest histopathological subtype was embryonal rhabdomyosarcoma. All our patients had stage I/group III. Only one of our patients died during treatment due to febrile neutropenia and multiorgan failure. 73% of the patients achieved complete remission following chemotherapy (CT) and radiation (RT). In two patients not responding to CT+RT, exenteration was required. Only one patient in our series had a local recurrence after being disease free for 1 year. At a median follow up of 30 months (range 6 -96 months), the event free survival was 80.18±10.12%. **Conclusion**: Orbital RMS commonly presents as early stage disease in young children, with unilateral proptosis being the most common presenting symptom. Orbital RMS has good prognosis.

Key Words: orbital rhabdomyosarcoma, chemotherapy, radiotherapy, embryonal rhabdomyosarcoma

INTRODUCTION

Orbital rhabdomyosarcoma (O-RMS) is the most common orbital malignancy of childhood. [1,2] The orbit is the primary site in approximately 10% of RMS. [3,4] Although O-RMS was once believed to arise from extraocular muscles, it is now accepted that O-RMS develops from undifferentiated mesenchymal cells that have the capacity to differentiate into striated muscle. [1] In majority of published series on RMS, orbit is the most favorable site with a 5year survival of 85%. [4,5] Outcome in Indian patients is largely unknown, as only a few series have been reported.

Since there is dearth of data from India, we conducted the present retrospective study over 9 years.

MATERIALS AND METHODS

Eighty nine children were diagnosed and treated for RMS during January 2003 to December 2010 at our institution. Of these, 38 patients had head and neck as primary site with 11 patients has O-RMS. The case files of these were analyzed for demographic profile, histological subtype, clinical staging and outcome. Staging workup included CT-scan/MRI of site, chest X-ray, abdominal USG, bone scan, bone marrow examination and CSF examination. Diagnosis was confirmed by histopathological examination and immunohistochemistry of primary lesion. Patients were staged according to IRSG presurgical Staging Classification and grouped as per IRSG Postsurgical Grouping.

Treatment of O-RMS involved a multimodal approach, using radiotherapy and chemotherapy (Vincristine and Dactinomycin for 1 year with radiation therapy beginning at week 2, based on the IRS-III protocol).[4] Dactinomycin was omitted during radiation. The prescribed dose of radiation was 45 to 55 Gy given by conventional fractionation over 4-6 weeks.
Post therapy patients were followed up every 3 months for the first 2 years, and 6 monthly thereafter. Progression during therapy and relapse were considered as events. The event-free survival (EFS) was calculated for all patients using the Kaplan Meier curve (SPSS 19 - SPSS Inc, USA).

Table 1: Demographic profile.

Age/ Gender	Clinical Presentation	HPE	Stage	Group	Date of starting treatment	Treatment Given	Follow-up
12/M	Proptosis	Embryonal	I	III	Aug 2003	CT+RT	EFS for 8 years
11/M	Proptosis, Eye movement Restriction	Embryonal	I	III	October 2003	CT+RT	EFS for 8 years
1/F	Eye pain & Swelling	Embryonal	I	III	December 2004	RT+CT	EFS for 7 years
4/M	Proptosis with chemosis	Undifferentiated	I	III	July 2006	CT+RT	EFS for 5 years
10/F	Proptosis	Embryonal	I	III	December 2006	CT+RT	EFS for 5 years
6/F	Eye pain & Swelling	Embryonal	I	III	May 2007	CT+RT+ EX	EFS for 3 years
13/M	Proptosis	Alveolar	I	III	Oct 2007	CT+RT	Died during treatment
3/M	Eyelid swelling & Chemosis	Embryonal	I	III	May 2008	CT+RT	EFS for 2 years
9/M	Proptosis	Embryonal	I	III	November 2008	CT+RT	EFS for 2 years
7/F	Proptosis	Alveolar	I	III	Jan 2010	CT+RT	Recurrence after 1 year
5/F	Eyelid swelling & Ptosis	Embryonal	I	III	April 2010	CT+RT+EX	EFS for 1 year

RESULTS

Demographic profile of patients is shown in table 1. Of the 11 patients were diagnosed during the study period there were 6 male and 5 female patients. The median age was 7 years (age range 1 yrs to 14 yrs). Commonest presentation was proptosis with diffuse involvement of the orbit, other symptoms like eyelid swelling, chemosis and ptosis were present in only 2 patients. The commonest histopathological subtype was embryonal. All our patients were stage I/group III.

Only one of our patients died during treatment due to febrile neutropenia and multiorgan failure. Complete remission rate

with CT+RT in present study was 73%. In two patients not responding to CT+RT, exenteration was required. Only one patient in our series had a local recurrence after being disease free for 1 year. At median follow up of 30 months (range 6 -96 months), the event free survival was 80.18±10.12% (figure1). Alveolar histology correlated with poor outcome (p=0.0182).

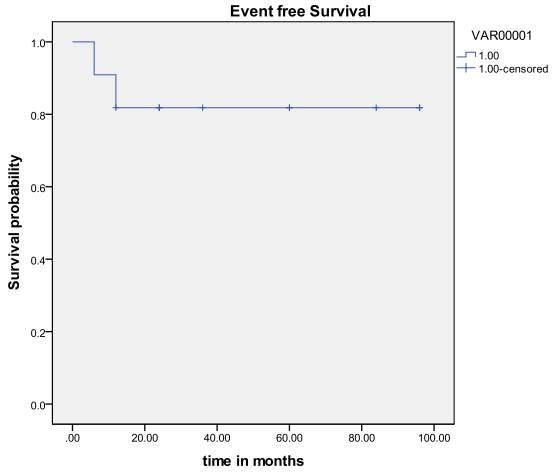

Figure 1: Event free survival.

DISCUSSION

Primary Orbital RMS is mainly a disease of young children, with 90% occurring before the age of 16 years with a mean age of onset of 5-7 year. [6]There is a slight male predilection with a male: female ratio of 5:3. [7] The present study had similar demographics.

The most characteristic presentation for primary orbital RMS is the rapid onset of unilateral proptosis and inferior or inferotemporal displacement of the globe. Proptosis can develop rapidly within a few days, or less commonly, present insidiously as a gradual painless process.[1] Most of the patients in present analysis presented with rapid onset of proptosis with diffuse involvement of the orbit.

Since most orbital tumors are biopsied without an attempt at resection, there is gross residual disease (group III). Thus, most orbital RMSs are stage I/group III, minorities are stage I/group I or II, and rarely, a primary orbital RMS is stage IV. [8] In present study all the patients had stage I/group III tumor.

In a comprehensive report of 264 patients from the Intergroup Rhabdomyosarcoma Study (IRSG), the tumor type was classified as Embryonal in 84%, Alveolar in 9% and Botyroid in 4%. Pleomorphic RMS is very rare in the orbit and generally occurs in adults. [9] In present study out of 11 patients, 9 patients had embryonal type of RMS. Histology was also correlated with survival, with an unfavorable outcome for patients with alveolar histology compared with patients with embryonal RMS. [9] This was seen in the present study as well, were patients with alveolar histology had significantly poor survival. The event free survival in present study (80%) was similar to majority of published series. [4,5]

CONCLUSION

Orbital RMS commonly presents as early stage disease in young children, with unilateral proptosis being the most common presenting symptom. Orbital RMS has good prognosis.

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