ORIGINAL ARTICLE

# Rhabdomyosarcoma of Head and Neck– A Ten Year Review

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**Abstract**

Rhabdomyosarcoma is an aggressive malignant skeletal neoplasm arising from embryonal mesenchyme. It accounts for over 50% of all paediatric soft tissue tumours. It may occur in any site of the body but the most common sites of tumor occurrence are orbit (12%) and head and neck (16%). Use of multimodality approach to treatment, including multidrug chemotherapy, radiotherapy has resulted in a dramatic improvement in the outlook of affected children. Orbit has long been recognized as a favourable site as compared to other head and neck sites due to paucity of lymphatics and high response rates seen with radiation therapy and chemotherapy. A ten year retrospective analysis of 31 patients of Rhabdomyosarcomas of head and neck was done. The main outcome measures were age, gender , histopathologic type, treatment received ,follow up period. and eventual outcome. Most of our patients presented with an advanced stage. A complete response of 91.6% and 28.2% was seen in orbit and other non orbital head and neck sites respectively. The 5- year disease free survival in patients of orbital rhabdomyosarcoma was 83.3%

**Key Words**

Rhabdomyosarcoma, Head and Neck, Orbit, Radiotherapy, Chemotherapy

## Introduction

Rhabdomyosarcoma is a common soft tissue sarcoma in infants ,children and adolescents, accounting for 4-8% of malignant tumours in less than 15 years of age (1). Rhabdomyosarcoma is a locally invasive tumour and has the potential for local spread along fascial or muscle planes, lymphatic or hematogenous dissemination. Head and neck is one of the commonest sites for this tumour in children. The frequently involved sites are non parameningeal-16%, orbit-12%,parameningeal-11%. Orbit has been considered a favorable site as compared to other head and neck sites due to the commonly encountered favorable histology of the embryonal subtype and the paucity of lymphatics in this area which accounts for a low incidence of lymph node metastasis as compared to other sites. Non orbital rhabdomyosarcomas are further grouped into parameningeal and non-parameningeal sites. The mainstay of treatment of rhabdomyosarcomas of head and neck is combined modality with radiotherapy and chemotherapy and surgery reserved for salvage.

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### Aims & Objectives

The objective of this study was to observe the clinico-pathological features, evaluate the prognostic factors and correlate the outcome in patients of rhabdomyo-sarcomas of head and neck treated in the department of Radiotherapy, Post Graduate Institute of Medical Education and Research, Chandigarh.

### Material & Methods

A reterospective analysis of patients of rhabdomyosarcoma of orbit and head and neck was done who were treated between 1991 - 2001. Total number of patients studied were thirty-three. The main outcome measures were age ,gender, histopathologic subtype , treatment received and eventual outcome. The age of the patients ranged from 3 - 21 years with mean age of 9 years. Total number of males was 21 and females, 12. The male to female ratio was 1.75:1. Orbit was the commonest site with 24 patients . Nine patients had primary tumour in parameningeal sites. Histological subtypes were embryonal -28 and pleomorphic-5.The patients were staged according to the IRS pretreatment staging system (Table 1). As orbit is a favorable site and none of the patients had metastasis, all the 24 patients were grouped under stage I. However, when patients were grouped according to clinical grouping classification based upon extent of surgery 30 patients were grouped under group III (Table 2). Majority of patients presented with locally advanced disease and in a poor general condition. Patients were treated with combined modality treatment with radiotherapy, chemotherapy and surgery. All patients received chemotherapy withVAC regimen (vincristine, adriamycin and cyclophosphamide). Sixteen patients received radical doses radiotherapy 45-50 Gy in 20-25 fractions. Nine patients who had locally bulky disease and those with a poor general condition received palliative doses of 30Gy in 10 fractions over 2 weeks.Thirteen patients received combined treatment with chemotherapy and radiotherapy; 12 underwent treatment with all three modalities. Follow up period ranged from 0 - 66 months with a median follow up of 14 months.

**TABLE 1**

**IRS CLINICAL GROUPING CLASSIFICATION**
**(International Rhabdomyosarcoma study)**

| | |
|---|---|
| GROUP - I | NONE |
| GROUP - II | 3 |
| GROUP - III | 30 |
| GROUP - IV | NONE |

**TABLE 2**

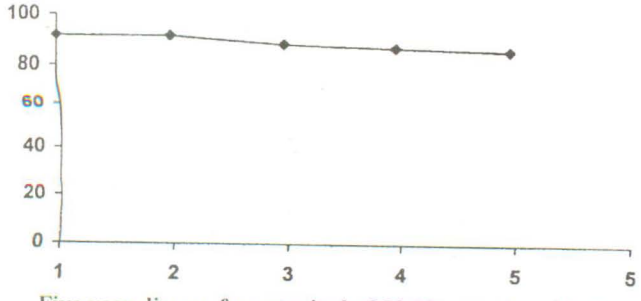
**IRS PRE-TREATMENT STAGING SYSTEM**
**(International Rhabdomyosarcoma study)**

| | | |
|---|---|---|
| GROUP - I | 24/33 | 72.7% |
| GROUP - II | 5/33 | 15.1% |
| GROUP - III | 4/33 | 12.1% |
| GROUP - IV | NONE | NONE |

### Results

Response was assessed after four weeks of treatment. A complete response was seen in 18 out of 33 patients. Patients who received radiotherapy and chemotherapy had a 61.5%, those who received radiotherapy, chemotherapy and surgery had a 58.3% complete response as compared to 37.5% for those who received only surgery and chemotherapy. Hence, a maximum response was seen in patients receiving radiotherapy .An initial complete response of 91.6% (22/24patients) was seen in those with orbit as the primary site whereas a complete response of only 28.2% (2/7 patients) was seen in those with parameningeal disease. A five year disease free survival for patients was 83.3% in those with orbital disease ie 20/23 patients were disease free at 5 years. All relapses occurred by four years. One out of the four patients who recurred had orbital disease. This patient underwent salvage by exenteration of the eye and continues to be locally disease free till date. Three patients had parameningeal as the as the primary site

and received salvage chemotherapy with vicristine, adriamycin, actinomycin D, ifosphamide and etoposide. One of the three patients continues to be disease free. Two patients had both local recurrence and distant metastasis. One had local recurrence in orbit with metastasis in the breast. The patient received radiation to the breast with salvage chemotherapy.




**Five year disease free survival of 83.3% seen in orbital rhabdomyosarcoma**

## Discussion

According to early reports of treatment of rhabdomyosarcomas, orbital disease was curable with orbital exenteration but was associated with a high incidence of local and distant failures and hence poor overall survival. Abramson in 1979, reported a 91% local control rate in 58 patients of orbital rhabdomyosarcoma treated with combined chemotherapy and megavoltage radiation therapy (1). High dose radiation therapy has been shown to provide local control in 90% cases when used alone and when used with chemotherapy has resulted in greater than 90% cure rates (2). Heyn *et. al.* demonstrated a 2 year survival of 82% in patients receiving adjuvant chemotherapy as compared to 53% for patients who received only surgery and radiation (3). Though initial intensive chemotherapy is being used as a means of providing pharmacological debulking, however when used alone in tumours of head and neck and pelvis, most children require radiation therapy because of incomplete response or local recurrence (4).Chemotherapy alone without radiation has resulted in local relapse, poor event free survival, and loss of functional vision (5). Non orbital parameningeal rhabdomyosarcomas have a propensity for invasion into base of skull. However, the prognosis of these patients has markedly improved with the use of adequate irradiation of primary tumour and adjacent meninges (6) Overall survival of 62.2% has been reported in the IRS III with the use of adjuvant chemotherapy after radiation (7). But as compared to orbital site, the survival for parameningeal tumours continues to be poor (8). For patients with orbital tumours,there is no longer an indication for exenteration at diagnosis. Orbital exenteration should be reserved for salvage treatment and enucleation for management of post treatment ocular complications (9). The current protocol for children with orbital relapse consists of chemotherapy with radiotherapy with surgery reserved for those with relapse.

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