Rhabdomyosarcoma-Report of Two Cases and Review

Rajasekhar G.,[1] Nandagopal Vura,[2] Sudheer,[3] Srikanth Dhanala,[4] Damera Srikanth[5]

ABOUT THE AUTHORS

1. Dr. Rajasekhar. G M.D.S
Professor &Head of the Department,
Department of Oral and Maxillofacial Surgery,
Mamata Dental College,
Giriprasadnagar,
Khammam.
Mobile: +91 9000018128
Email: drrajasekharg@yahoo.com

2. Dr. Nandagopal Vura M.D.S
Associate Professor
Department of Oral and Maxillofacial Surgery,
Mamata Dental College,
Giriprasadnagar,
Khammam.

3. Dr. Sudheer M.D.S
Reader
Department of Oral and Maxillofacial Surgery,
Mamata Dental College,
Giriprasadnagar,
Khammam.

4. Dr. Srikanth Dhanala
M.D.S
Senior Lecturer
Department of Oral and Maxillofacial Surgery,
Mamata Dental College,
Giriprasadnagar,
Khammam

5. Dr. Damera Srikanth
Postgraduate
Department of Oral and Maxillofacial Surgery,
Mamata Dental College,
Giriprasadnagar,
Khammam.
Mobile: +91 9948325066
Email: damerasrikanth@gmail.com

Corresponding Author:

Dr.Rajasekhar.G M.D.S
Professor &Head of the Department,
Department of Oral and Maxillofacial Surgery,
Mamata Dental College,
Giriprasadnagar,
Khammam.
Mobile: +91 9000018128
Email: drrajasekharg@yahoo.com

Abstract

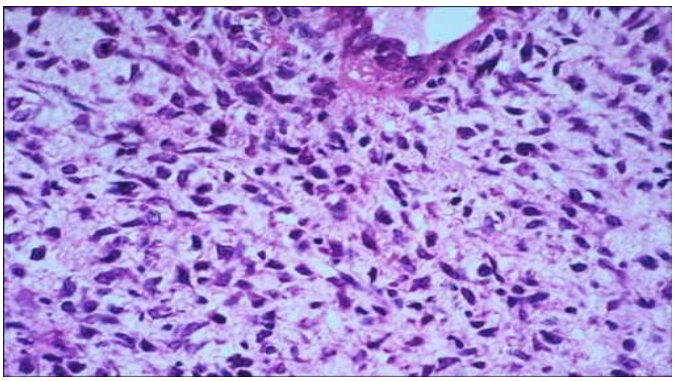
Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children. It is seen rarely in adults. There are three variants of RMS which include embryonal, alveolar which are commonly seen in children and pleomorphic type seen in adult. In recent years, spindle cell and sclerosing variants of RMS have also been recognized in adults. Because of the scarcity of data further extensive research has to be carried regarding the tumor. The prognosis of the tumor is worse in adults when compared to children, with an overall 5-year survival of less than 30%. This review will discuss the clinicopathological characteristics of RMS, the different histological variants, and two case reports of our Institution.

KEYWORDS: Embryonal, Alveolar, Pleomorphic, RMS-Rhabdomyosarcoma

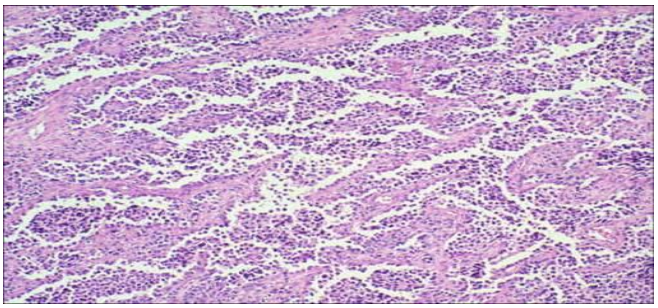
Introduction

Rhabdomyosarcoma is a malignant tumor of the skeletal muscle .It is one of the most common sarcomas of head and neck soft tissue in the pediatric population. It is rarely found in adults.
Description about the Rhabdomyosarcoma was given well before in 1854 by WEBER but the published documentation was done by STOUT in 1946.It is one of the very rare kind of tumor ,there are only 350 reported cases every year in united states in children under the age of 21 and there are only 400 cases of adults which were reported in united states and Europe over the past 20 to 30 years.[1]
The most common site of the occurrence of tumor is in the head and neck region accounting for almost 40% ,with 25% occurrence in genitourinary tract and 20% in the extremities of the body and 15% in parameningeal site.[1]
Rhabdomyosarcoma occurs in three variants based upon its histology Embryonal , Alveolar ,Pleomorphic.[3] Of the three subtypes the most common is embryonal type which accounts for 60 to 70 percent observed in children. Alveolar subtype making up about 20%which generally occurs between 10 to 20 years of age while the majority of these occur in extremities. Pleomorphic is the least common of all, occurs in extremities and generally seen in elderly.[4]

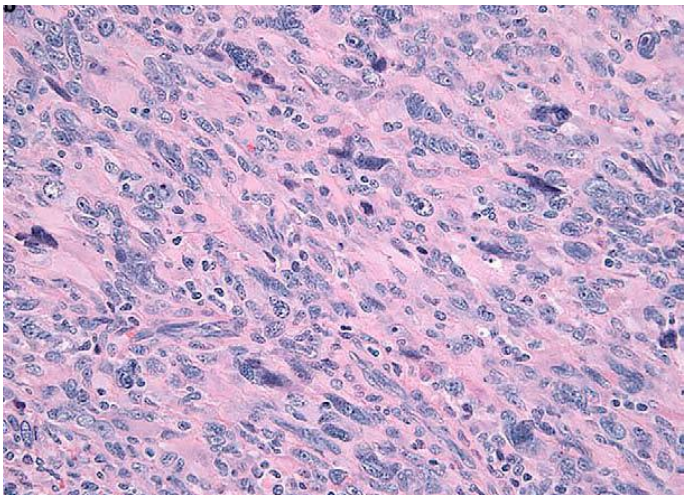
HISTOLOGY OF RMS TYPES
EMBRYONAL RHABDOMYOSARCOMA exhibits mixture of four cell types Eosinophilic spindle cell ,usually arranged in fascicles. Round eosinophilic cells , large and intermediate in size, with small nucleus and granular eosinophilic cytoplasm, interspersed among other cell type ,Broad elongated eosinophilic cells , occasionally with cross striations .Small round and spindle cells with dark staining nuclei and little cytoplasm[1]



ALVEOLAR RHABDOMYOSARCOMA is comprised of small, poorly differentiated round and oval cells aggregated into irregular clusters separated by fibrous septa. Degenerated cells in the center lack cohesiveness and peripheral cells are arranged in a single layer[1,3],

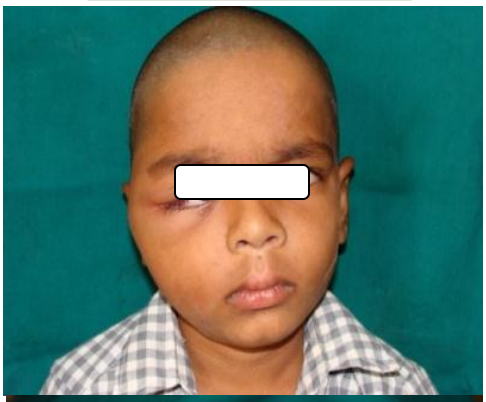


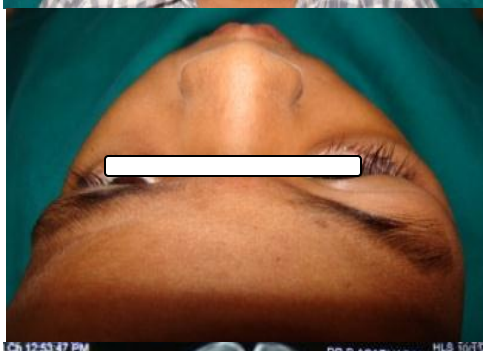
PLEOMORPHIC RHABDOMYOSARCOMA is composed chiefly of spindle cells in a haphazard arrangement. These cells are generally large and show considerable variation in appearance. The characteristic feature of this tumor is large bizarre shape cells with nuclei situated in the expanded end of the cell. [3] The tumor is so undifferentiated that the identification of the cell of origin is difficult.

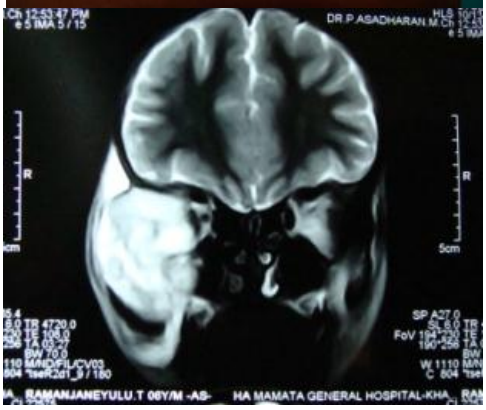


Case report 1:

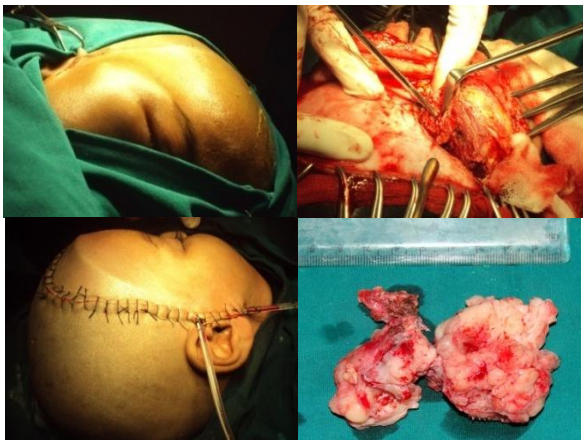
Preoperative



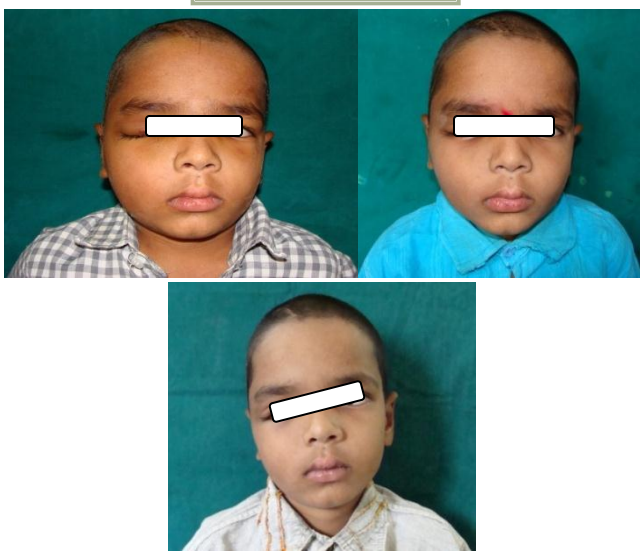




Intra operative



Post operative

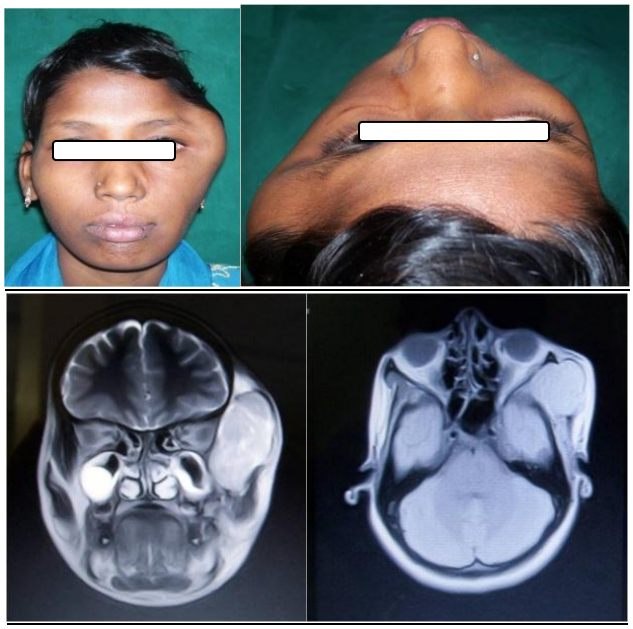


A six year old male with complaint of swelling and pain on right side of face since 2 months which was growing rapidly since 1 month. On examination diffuse swelling extending superiorly from supra orbital notch to zygomatic arch inferiorly, posteriorly up to preauricular region, anteriorly up to infraorbital region. On palpation the swelling was tender, firm in consistency. Mouth opening was adequate. Intra oral findings are noncontributory.

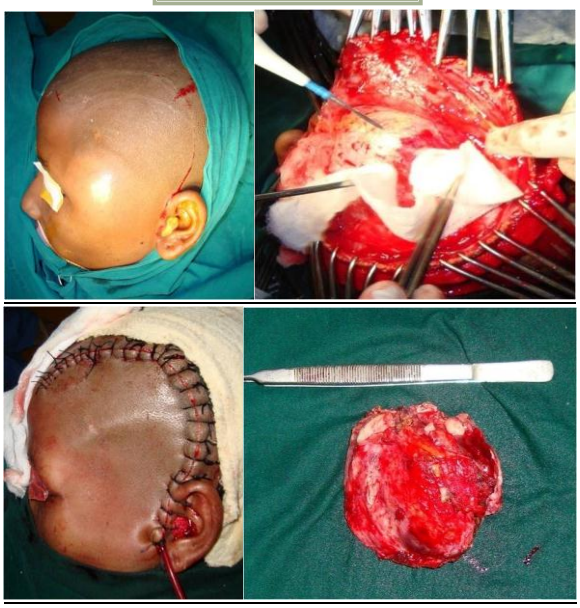
Aspiration cytology revealed rhabdomyosarcoma. MRI contrast showed the extension of lesion in the infra temporal area. Hemicoronal approach is planned with zygomatic arch access osteotomy for surgical excision of the Tumor.

Case report 2 :

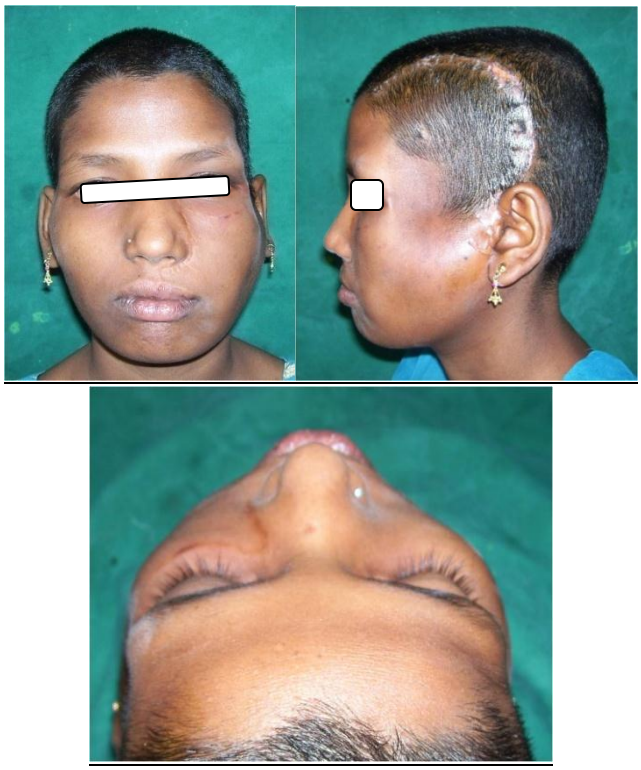
Preoperative



Intraoperative



Postoperative



A 14 year old female patient with complaint of swelling on the left side of face rapidly growing since three months. On examination the swelling was non tender and firm in consistency. Facial asymmetry was evident on left side of face with diffuse swelling on the left zygomatic and infra temporal region. Aspiration cytology study shows Rhabdomyosarcoma.

Extension of tumor mass in infratemporal fossa was removed after fracturing the zygomatic arch and later

coronoidectomy is carried out. Hemostasis achieved and irrigation done and incision closed in layers with vicryl and silk by interrupted suturing with drain in place. Post operative healing was satisfactory and patient was advised for radiotherapy.

DISCUSSION

Rhabdomyosarcoma is a malignant tumor arising from the skeletal muscle cell and the tumor is most commonly seen in children with in the age of ten years, usually seen in 1-4 years of age and is uncommon in infants and below the age of one year[1]

Rhabdomyosarcoma is rarely seen in adults but the embryonal variant occurs more common and later preceded by Pleomorphic variant.[11] The patients reported to our department are of young age group with six year old male and fourteen year old female of Indian origin. Males and especially Caucasians are slightly more affected than females accounting for almost eighty percent of cases seen in adults.[3]

Alveolar Rhabdomyosarcoma accounts for approximately 30% of all which affects mainly adolescents and young adults of both sexes. These tend to occur in deep soft tissues of the extremities .[5,10] The histopathological report of our cases confirms Alveolar Rhabdomyosarcoma, which is the most common variant of all. The occurrence of this condition in our cases was in the deep tissues (Infratemporal fossa).

BARNES noted that the tongue and palate are the most commonly affected intra-oral sites of RMS. The clinico-epidemiological analysis on Nigerian population by OA FATUSI , identified the palate and cheek as the most common intra-oral sites. No tongue lesion was recorded.[5]

L. WALLACE FRANK reported a case of Rhabdo-myosarcoma occurring unusually at neck region. Spindle cell variant of Rhabdomyosarcoma is rarely seen in adults but a case of radiation induced spindle cell RMS has been reported by V.GOOSENNS ET AL.[2] We present here an unusual cases of Alveolar Rhabdomyosarcoma occurring in the Infratemporal region.

To evaluate imaging findings of Rhabdomyosarcoma of the head and neck in adults, CT and MRI plays a very important role in establishing the extent of disease. Tumors arising from the maxillary sinus shows poorly defined homogenous mass extending into surrounding spaces. CT findings of head and neck Rhabdomyosarcoma have been described as showing poorly defined, inhomogeneous soft-tissue masses destroying adjacent bone.

The most commonly reported MR imaging appearance of Rhabdomyosarcoma in the head and neck is that of a homogeneous mass, iso intense or minimally hyper intense relative to muscle on T1-weighted images and hyper intense relative to both muscle and fat on T2-weighted images, with post contrast images showing enhancement of the tumor.[7] Precisely, the MRI images in our cases produced exact extensions of the tumor mass with the boundaries. As the tumor extended into the Infratemporal space, the emphasis was made on proper preoperative investigations for ideal management of the tumor which was extending into surgically inaccessible site.

A new classification based on the prognostic value of different forms of RMS has been proposed by WILLIAM A. NEWTON et al.

The histologically recognizable types are assigned to one of three prognostic groups. Based on the results of this study, Botryoid and spindle cell RMS are associated with a superior prognosis; conventional Embryonal RMS is associated with an intermediate prognosis. Alveolar RMS (all types) is associated with a poor prognosis.[4] We have successfully followed our cases up to two years.

C. ASKEW proposed a multi modal approach in the successful management of Rhabdomyosarcoma. Initially surgery was the sole therapeutic modality available for RMS .Surgery tended to be radical and associated with significant morbidity if complete excision was to be achieved. The potential role of radiotherapy in management of recurrent RMS emerged.[11]

The multimodal approach of Rhabdomyosarcoma is accordingly applied in our cases by surgical debulking of tumor mass to control further extension of tumor into base of skull. Surgical excision was followed by radiotherapy which resulted in complete regression of the apparent tumor including the lesion noted in radiographic examination with a successful outcome.[11]

F. GHAVIMI et al has proposed the multi disciplinary protocol for the treatment of Embryonal Rhabdomyosarcoma for children under **15** years with surgical removal of the tumor followed by chemotherapy, and also with radiation therapy in patients with gross or microscopic residual disease. Radiation therapy was given in the **4500-7000** rads range. The chemotherapy consisted of cycles of sequential administration of dactinomycin, Adriamycin, vincristine, and cyclophosphamide, with obligatory periods of rest. The drug therapy was continued for 2 years with high successful prognosis [14]

Chemotherapy was not administered in our cases as the surgery and radiotherapy resulted in complete regression of the tumor mass. Considering the young age of the patients and weighing the potential adverse effects of chemotherapy the earlier modality was preferred and we could achieve a satisfactory result.

RICHARD E. ALBIN, has described Transzygomatic approach for the surgical management of Rhabdomyosarcoma involving the pterygoid fovea. The approach with caution produce an excellent preservation

of branches of facial nerve both temporal and zygomatic. Following surgery radiotherapy provides complete eradication of tumor at in accessible sites.[8] Surgical excision was performed under General Anesthesia by Transzygomatic approach, which provided good visibility and accessibility with minimal facial asymmetry and negligible loss of function post operatively.

In spite of advances in the field of surgery, chemotherapy and radiotherapy the recurrence of 30 percent Rhabdomyosarcoma has been observed. ALBERTO S. PAPPO analysis proposes a probability of five year survival of such patients. The prognosis depends on several factors including initial diagnosis, histologic subtype, disease group, and stage.[6] The follow up was done for two years in our cases with no signs of recurrence.

CONCLUSION

Gross removal of these sarcomas appears to provide better disease control. Resection of the tumor followed by radiotherapy remains an important factor in prognosis. Whenever there is no evidence of distant metastasis or CNS invasion, an aggressive surgical approach forms primary treatment modality. The Trans zygomatic approach has a wider application in the surgical treatment of the tumors of infra temporal fossa. Co operation among both surgical and non surgical specialties should result in continued advancement in the treatment of these difficult oncologic problems.

REFERENCES

1. Leonard H Wexler. Rhabdomyosarcoma. Soft Tissue Sarcoma Section Memorial Sloan-Kettring Cancer .esun article 2004.

2. V.Goosens, Vanden Berghe, et al: Radiation induced Mandibular adult spindle cell Rhabdomyosarcoma. Int. J. Oral Maxillofac. Surg. 37: 395-397, 2008

3. Sabrina Rossia , Antonio G nascimentob, et al: Small round – Cell neoplasms of soft tissues: An integrated diagnostic approach. Current Diagnostic Pathology . 13: 150-163, 2007

4. Roberet. Horn , Horatito, et al: Rhabdomyosarcoma : A clinico pathological study and classification of 39 cases. Cancer vol 11.Jan -1958.

5. O. A. Fatusi, S. O.Ajike, et al: Clinico-Epidemiological analysis of Orofacial Rhabdomyosarcoma in a Nigerian population. Int. J. Oral Maxillofac. Surg. 38: 256–260. 2009.

6. Alberto S. Pappo, James R. Anderson, et al : Survival after Relapse in Children and Adolescents with Rhabdomyosarcoma: A Report From the Intergroup Rhabdomyosarcoma Study Group. J Clin Oncol 17:3487-3493.1999.

7. Jeong Hoon Lee, Mi Sook Lee, et al : Rhabdomyosarcoma of the Head and Neck in Adults: MR and CT Findings. Am J Neuroradiol 17:1923–1928, 1996.

8. Markku Miettinen: Rhabdomyosarcoma in Patients Older Than 40 Years of Age. Cancer 62:2060-2065, 1988.

9. Gary W**.** Mierau, Blaise E. Favara : Rhabdomyosarcoma In Children: Ultrastructural Study Of 31 Cases.Cancer 46:2035-2040, 1980.

10. Ramzi Dagher, Lee Helman: Rhabdomyosarcoma: An Overview. The Oncologist 1999;4:34-44

11. Askew, P.S. Fleming, et al: Successful Multimodal Management of Rhabdomyosarcoma and dento- facial sequelae of treatment. Oral Oncology 42: 52-55, 2006.

12. Harold M. Maurer, Mohan Beltangady, et al: The Intergroup Rhabdomyosarcoma Study-I .A Final Report. Cancer 61:209-220,1988.

13. Ruth Heyn, Abdel Ragab, et al: Late Effects of Therapy in Orbital Rhabdomyosarcoma in Children. A Report From the intergroup Rhabdornyosarcoma Study. Cancer 57,1738-1743, 1986

14. F. Ghavimmi, D, P. R. Exelbym et al : Multidisciplinary Treatment of Embryonal Rhabdomyosarcoma in Children. Cancer 35:677-686, 1975.

15. Charles B. Pratt, H. Omar Hustu et al : Coordinated Treatment of Childhood Rhabdomyosarcoma with Surgery, Radiotherapy, and Combination Chemotherapy. Cancer research 32, 606-610, 1972.