
Rhabdomyosarcoma of Lip: A Case Report**Dr Bhavya Thakur^{#1}, Dr Shirin Garg^{\$2}, Dr Harpreet Puri^{*3}, Dr Ruchika Tyagi^{*4}, Dr Ramneesh Garg^{#5}, Dr Sheerin Shah^{#6}**¹MCh Resident [#], ²Consultant Gynaec- Oncosurgeon^{\$}, ³Professor, ⁴Associate Professor ^{*}, ⁵Associate Professor[#], ⁶Assistant professor^{#@}^{\$}Department of gynaec- oncology, Oswal Hospital and cancer institute, Ludhiana.^{*}Department of Pathology, Dayanand medical college and hospital, Ludhiana[#]Department of Plastic Surgery, Dayanand Medical College & Hospital, Ludhiana – 141001, Punjab, India***Corresponding author**

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Abstract: Rhabdomyosarcomas (RMS) are the malignant tumors of the striated skeletal muscles. RMS was first described by Weber in 1854 and accounts for 6% of all malignancies and represents 50% of all soft tissue sarcomas in children under 15 years of age. 1The most common sites of occurrence are the head and neck (40%), genitourinary tract (25%), and extremities (20%). 2Oral Rhabdomyosarcoma is rare neoplasm, and accounts for only 0.04% of all head and neck malignancies. 3 Even in the oral cavity when it occurs the most common site is soft palate.1 Here we present a case of 4 year old female child who was diagnosed as having rhabdomyosarcoma of the upper lip diagnosed by histopathology and confirmed by immunohistochemistry.**Keywords:** Rhabdomyosarcoma lip, paediatric rhabdomyosarcoma, paediatric lip swelling, Cancer paediatric lip

INTRODUCTION

Rhabdomyosarcomas (RMS) are the malignant tumors of the striated skeletal muscles. RMS was first described by Weber in 1854, and accounts for 6% of all malignancies and represents 50% of all soft tissue sarcomas in children under 15 years of age[1]. The most common sites of occurrence are the head and neck (40%), genitourinary tract (25%), and extremities (20%)[2]. Oral Rhabdomyosarcoma is rare neoplasm, and accounts for only 0.04% of all head and neck malignancies[3]. Even in the oral cavity when it occurs the most common site is soft palate[1].

Here, we present a case of 4 year old female child who was diagnosed as having rhabdomyosarcoma of the upper lip diagnosed by histopathology and confirmed by immunohistochemistry.

CASE REPORT

A 4 year old female child presented with history of swelling right upper lip since 6 months of age. The swelling kept on increasing size and started causing disfigurement of face which is when the parents took the child to a private hospital when the child was one and a half years of age where it was operated upon, however there were no records of surgery or any histopathology report. The child then remained asymptomatic for 2 years, but the parents noticed recurrence which increased progressively in size reaching its present size in 4 months. The patient apart from disfigurement of face was asymptomatic.

Examination revealed a non-tender, non-pulsatile, non-compressible mass of 3x3 cm with irregular surface and well defined margins involving the right upper lip and obliterating the right nasolabial sulcus. [Figure 1] Both the underlying mucosa and overlying skin could not be pinched separately. There was no cervical lymphadenopathy.

The patient was subjected to wide local excision [Figure 2] and primary closure [Figure 3], tissue was sent for histopathological assessment after the sample was oriented. [Figure 4]

The histopathology report impression was consistent with malignant neoplasm and possibility of rhabdomyosarcoma was kept and immunohistochemistry was advised for confirmation. [Figure 5, 6]

The immunohistochemistry showed vimentin, myogenin and S-100 focally positive in tumour cells, desmin positive and CD34, SMA and cytokeratin negative in the tumour thus being consistent with the diagnosis of rhabdomyosarcoma.

The patient remained in our follow up for 1 month and no recurrence was noted [Figure 7]. The patient was advised chemotherapy and therefore sent to oncology for the same however the patient went absconding after that.



Fig-1: Shows swelling right upper lip obliterating the right nasolabial sulcus



Fig-2: Showing the defect after local wide excision.

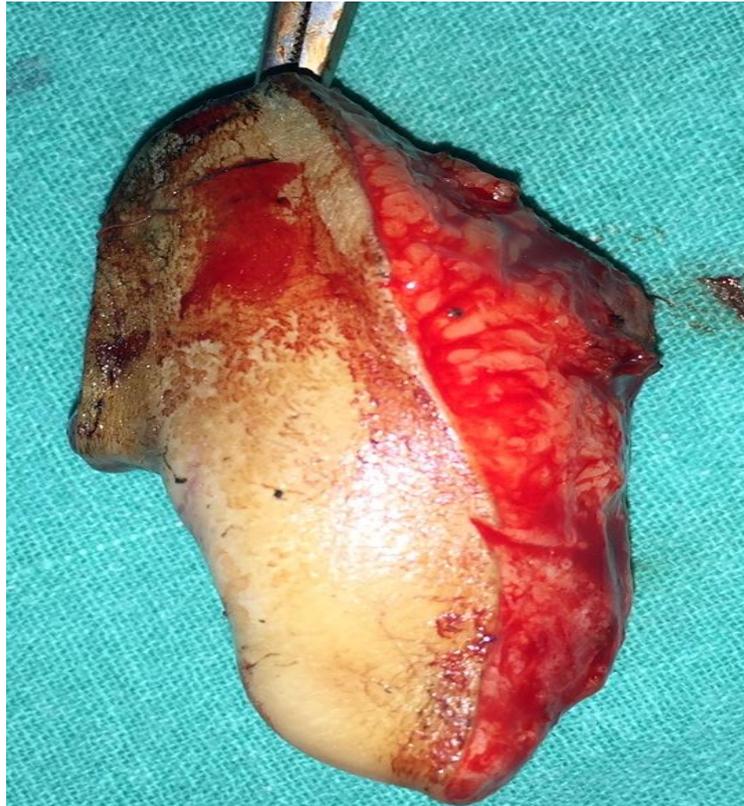


Fig-3: Excised tumour along with rim of healthy tissue



Fig-4: Immediate post-operative picture

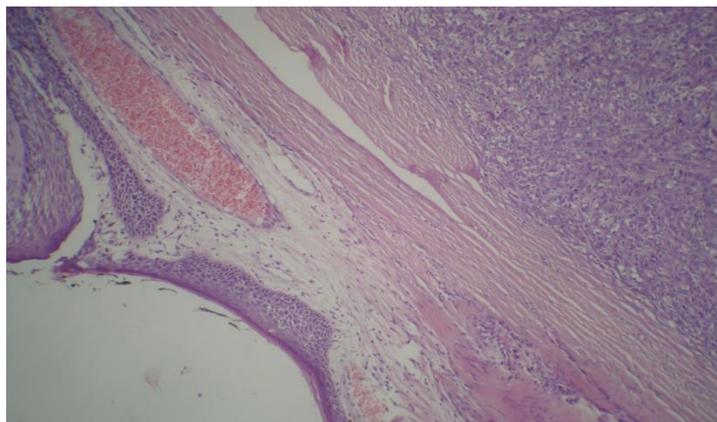


Fig-5: Shows a well circumscribed tumour with tumour cells arranged in diffuse sheets and fascicles.

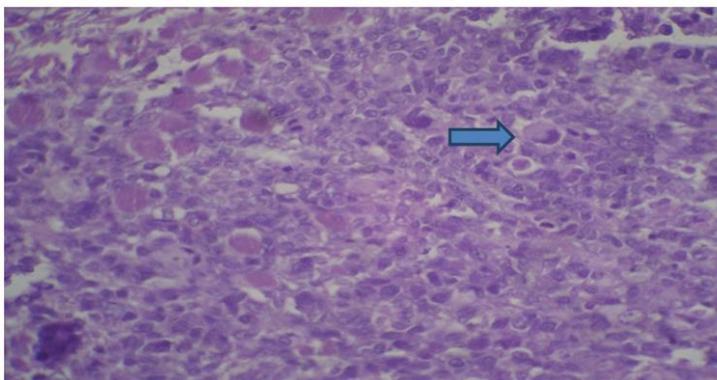


Fig-6: Arrow shows a rhabdomyoblast eccentric nucleus and abundant bright eosinophilic cytoplasm

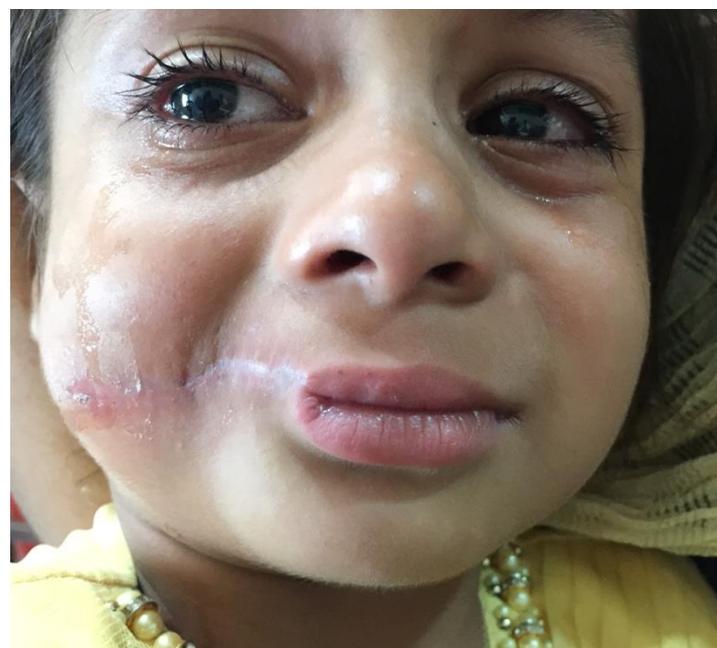


Fig-7: 1 month postoperative picture.

DISCUSSION

RMS is a rare disease, more prevalent in young boys [1]. The highest incidence of RMS is in the children aged 1-4 years, lower rate in children aged 10-14 aged and lowest rates between 15-19 years of age[4]. In our case we had a 4 year old female which falls in high risk category.

Clinically, the manifestations of RMS may vary from a small cutaneous nodule on the face to an extensive fast-growing facial swelling, which may be painless or occasionally associated with pain, trismus, paresthesia, facial palsy, and nasal discharge[5]. Our

patient too had history of rapid increase in size however there were no other complaints.

Four basic histological patterns of RMS are recognized: embryonic, botryoid, alveolar and Pleomorphic. The embryonic type represents more than 70% of all cases[6]. RMS involving the head or neck is most commonly of the embryonic subtype and rarely involves regional lymph nodes, lymphatic spread is unusual with head and neck primaries particularly in children under the age of teen[7]. Oral RMS is seen more commonly in males and usually occurs during the first two decades of life[8].

The head and neck region is the most common site for RMS, with the orbit being the most frequent primary site. In the oral cavity the most common sites are tongue, palate and buccalmucosa[8,9]. In our patient the upper lip was involved which is very rare.

The diagnosis of RMS is confirmed through biopsy of the primary tumor and an adequate amount of tissue should be obtained[1]. In the described case, we chose to do an excision biopsy. Microscopic examination in our patient revealed a well circumscribed tumour, showing brisk mitosis and rhabdomyoblast .

Treatment of the lesions is by a multidisciplinary approach. It consists of surgical removal of the tumor followed by multiagent chemotherapy. Prognosis of the lesion depends on age of the patient, anatomic site, clinical staging (tumor size, node involvement and metastasis) and H/P subtype. Twenty percent of patients develop metastasis at the time of diagnosis; and lung, lymph node and bone marrow are the common sites with or without radiotherapy[10,11]. Classic embryonal rhabdomyosarcoma has an intermediate prognosis; 5-year survival rate is around 75%[12].

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