

INTRAORAL PLEOMORPHIC RHABDOMYOSARCOMA : A CASE REPORT

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Abstract

Rhabdomyosarcoma (RMS) is a malignant neoplasm of primitive mesenchyme exhibiting skeletal muscle differentiation. Intraoral Rhabdomyosarcomas are rare. We report here a case of pleomorphic Rhabdomyosarcoma of oral cavity in a 10-year-old child. The patient was diagnosed clinically and radiographically and planned for surgery. The clinical specimen was sent for histopathological studies. Definitive diagnosis is made by microscopic analysis and other auxiliary techniques such as immunohistochemistry, electron microscopy, cytogenetic analysis and molecular biology. Clinical, histological and immunohistochemical aspects suggests the diagnosis of rhabdomyosarcoma. Although rare, Rhabdomyosarcoma should be included in the differential diagnosis of intra-oral lesions, especially in children. This case illustrates clinical, histopathological aspects and diagnostic difficulties of Rhabdomyosarcoma. Taken together, we conclude that in children, any fast growing swelling should be carefully examined with a high degree of suspicion of this entity. Timely diagnosis and multidisciplinary treatment approach may improve the patient's survival.

Key Words: Immunohistochemistry, Rhabdomyosarcoma

Introduction

Described by Weber in 1854, RMS accounts for 6% of all malignancies in children under 15 years of age^{1,9}. The most commonly affected areas are the head and neck region, genitourinary tract, retroperitoneum, and to a lesser extent the extremities^{2,10}. In the head and neck, the most frequently affected sites are orbit, paranasal sinuses, soft tissues of the cheek and the neck³. Oral RMS is rare, and when occurring, it is more frequent in the soft palate⁹. Intraoral RMS corresponds to 10 to 12% of all head and neck rhabdomyosarcoma. Clinically the manifestations of rhabdomyosarcoma may vary from a small cutaneous nodule to an extensive fast-growing facial swelling, which may be painless or occasionally associated with pain, trismus, paresthesia, facial palsy, and epistaxis¹. The diagnosis is generally made

by microscopic analysis and auxiliary techniques such as immunohistochemistry, electron microscopy, cytogenetic analysis, and molecular biology.

Histologically four broad subtypes of Rhabdomyosarcoma has been identified: botryoid and spindle cell Rhabdomyosarcoma, embryonal Rhabdomyosarcoma, alveolar Rhabdomyosarcoma, and pleomorphic Rhabdomyosarcoma^{4,5}. Within the microscopic patterns, the embryonal type is the most frequent in oral cavity. Undifferentiated / pleomorphic Rhabdomyosarcoma is very rare in children. Rhabdomyosarcomas are associated with high rates of recurrence and generalized metastases through haematogenous and /or lymphatic routes. Prognosis is relatively poor compared to that of other oral soft tissue malignant lesions and depends on the clinical staging and the anatomic site of the tumor.

Case Report

A ten years female patient reported to the Department of Oral and Maxillofacial Surgery department, KGMC, Lucknow with complaint of progressively increasing swelling in left maxillary vestibule which was associated with pain and difficulty in mastication and swallowing. Extra oral examination showed extensive left sided swelling of face with ill-defined limits and smooth surface, reaching the mandibular ramus, zygomatic and infra-orbital regions (Fig 1). Left submandibular and upper jugular lymph nodes were palpable. Intraoral examination showed a large, firm and tender swelling on left upper deciduous first molar up to the maxillary tuberosity region extending from left side palate to buccal vestibule measuring 3x5 cm (Fig 2) . It was tender, firm, fixed and bleed on probing. CT scan showed ill defined destructive lesion involving maxilla and maxillary sinus and floor of left orbit with extension to adjacent structures (Fig 3) .On the basis of history and clinical examinations, we came to the provisional diagnosis of Rhabdomyosarcoma and Ewing's sarcoma and surgery was planned under general anaesthesia after routine haematological investigation informing the patient about the disease and taking the written consent for the same. The upper buccal vestibular incision was given and layer wise dissection was done, lesion was exposed and taken out. The flap was repositioned and sutured. Subsequent histopathological examination showed tumor composed of interlacing fascicles and solid sheets of predominantly spindled cells with pleomorphic hyperchromatic nuclei, prominent nucleoli and moderate amount of eosinophilic cytoplasm. Occasional Rhabdoid cells having moderate to abundant eosinophilic cytoplasm and eccentric nucleus along with moderate number of mitotic figures were also seen (Fig 4a). Immunohistochemistry showed Desmin and Ki 67 positivity while CK, S100 and smooth muscle actin were negative (Fig 4b,4c). Thus a definitive diagnosis of pleomorphic

Rhabdomyosarcoma was given. The patient was referred to Department of Oncology, KGMC, Lucknow where chemotherapy was started. The drug regime included Vincristine, Ifosfamide, Actinomycin and Mensa. Unfortunately the patient passed away after receiving 3 cycles of chemotherapy.



Fig1: Extraoral examination showing extensive left sided ill-defined swelling of face involving mandible ramus, zygomatic and infra-orbitary regions.



Fig 2: Intraoral examination of the same mass lesion.



Fig.3: CT Scan showing ill-defined destructive lesion involving left maxilla and maxillary sinus and floor of left orbit with extension into adjacent structures.

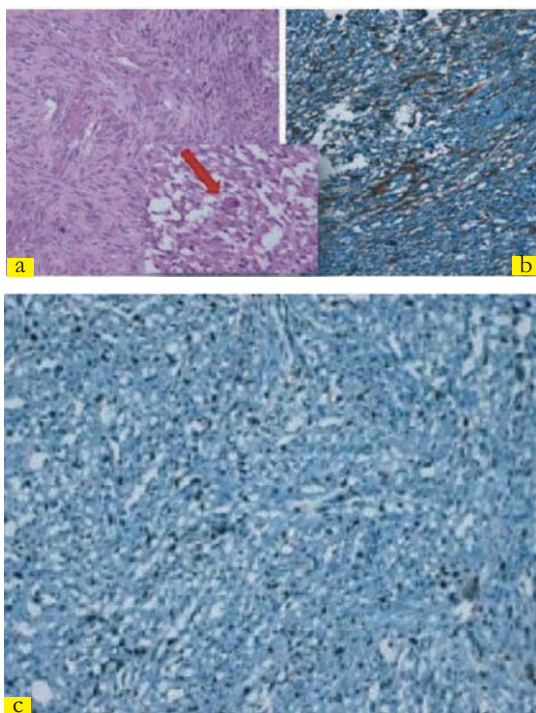


Fig.4:

- (a) Microphotograph showing tumor composed of interlacing fascicles and solid sheets of predominantly spindled cells with pleomorphic hyperchromatic nuclei. Occasional Rhabdoid cells were also seen (Inset marked by arrow)
- (b) Immunohistochemistry showing Desmin positivity
- (c) Immunohistochemistry showing SMA negative

Discussion

Rhabdomyosarcoma is the most common soft-tissue sarcoma of childhood and adolescence. Of all soft tissue sarcomas, Rhabdomyosarcoma accounts for only 2–5% tumors in adults but approximately 60% tumors in children⁶. The incidence of Rhabdomyosarcoma is highest in children between 1-4 years of age. Our patient was 10 years old and did not fall in the very high-risk category. Head and neck Rhabdomyosarcoma is anatomically divided in two categories: parameningeal (including nose, nasopharynx, paranasal sinuses, mastoid region, infra-temporal and pterygopalatine fossae and middle ear) and non-parameningeal (which include scalp, orbit, parotid gland, oral cavity, oropharynx and larynx). Most common site of involvement in oral cavity is the tongue followed by the soft palate, hard palate, and buccal mucosa^{7,12}. In the case we presented here, the lesion was involving upper jaw and was extensive in size measuring 3x5 cm in dimensions. This tumor is usually fast-growing and infiltrative and often appears as an enlarging, painless or sometimes slightly painful mass. Initial symptoms may be vague, and may mimic other benign neoplastic, inflammatory, or infectious processes. In cases of Rhabdomyosarcoma, radiographic examination reveals the size of the lesion, spatial relations, and the extent of bone destruction, while CT scans reveal the affected areas such as the mandible, condyle, maxillary sinus, and infra-orbital regions. Diagnosis is based on histological findings. A careful histological examination is required to differentiate Rhabdomyosarcoma from other more frequent and aggressive lesions affecting the concerned site like Ewing's sarcoma, Leiomyosarcoma or Neurofibrosarcoma. In our case, histological picture revealing predominantly spindle cell population, marked pleomorphism, fascicular pattern, etc was suggestive if not confirmative for differentiating Rhabdomyosarcoma from Ewing's sarcoma and Neuroblastoma. Features favouring Rhabdomyosarcoma over soft-tissue sarcomas such as fibrosarcoma, leiomyosarcoma and neurofibrosarcoma was

the presence of Rhabdoid cells but definitive confirmation requires immunohistochemistry. According to literature, clinical diagnosis of Rhabdomyosarcoma is difficult; a fact that can markedly affect the patient's prognosis. In the case reported the lesion was extensive and was involving maxillary bone. Embryonal Rhabdomyosarcoma are more common in younger age group while in this case presence of diverse morphological features suggested pleomorphic Rhabdomyosarcoma; this rather aggressive tumor type does not occur in children⁷. Immunohistochemistry is mandatory for definitive diagnosis. Stains such as myoglobin, muscle specific actin HNF35, vimentin, desmin and myosin are useful immunohistochemical markers for the disease.

Prognosis of Rhabdomyosarcoma is relatively poor compared to that of other oral soft tissue malignant lesions⁸ and depends on the clinical staging and the anatomical site of the tumor. Treatment of Rhabdomyosarcoma consists of surgical resection, when possible, associated with multiagent chemotherapy and/ or radiotherapy. Possible complications of this multidrug chemotherapy includes myelosuppression, oral mucositis, nausea and vomiting¹¹.

Conclusion

Rhabdomyosarcoma is derived from primitive mesenchymal tissues that exhibit tendency towards myogenic differentiation and probably originate from satellite cells associated with the embryogenesis of skeletal muscle. This case illustrates clinical, histopathological aspects and diagnostic difficulties of Rhabdomyosarcoma. Taken together, we conclude that in children, any fast growing swelling should be carefully examined with a high degree of suspicion of this entity. Microscopy and immunohistochemistry are helpful in

diagnosis. Prompt diagnosis and multidisciplinary treatment approach can improve the prognosis.

References

1. Chigurupati R, Alfatooni A, Myall RW, Hawkins D, Oda D. Orofacial rhabdomyosarcoma in neonates and young children: a review of literature and management of four cases. *Oral Oncol.* 2002;8:508–515.
2. Duraes GV, Jham BC, Mesquita ATM, dos Santos CRR, Miranda JL. Oral embryonal rhabdomyosarcoma in a Child: A case report with immunohistochemical analysis. *Oral Oncol Extra.* 2006;42:105–108.
3. T. Seth and P. Kempert, Embryonal rhabdomyosarcoma of lower lip, *Indian Pediatr* 41 (2004), pp. 858–859.
4. França CM, Caran EM, Alves MT, Barreto AD, Lopes NN. Rhabdomyosarcoma of the oral tissues -two new cases and literature review. *Med Oral Patol Oral Cir Bucal.* 2006;11:136–140.
5. Gordon-Nunez MA, Piva MR, Dos Anjos ED, Freitas RA. Orofacial rhabdomyosarcoma: report of a case and review of the literature. *Med Oral Patol Oral Cir Bucal.* 2008;13:765–769.
6. Sekhar MS, Desai S, Kumar GS. Alveolar rhabdomyosarcoma involving the jaws: a case report. *J Oral Maxillofac Surg.* 2000;58:1062–1065.
7. Hicks J, Flaitz C. Rhabdomyosarcoma of the head and neck in children. *Oral Oncol.* 2002;38:450–459.
8. Yamaguchi S, Nagasawa H, Suzuki T, Fujii E, Iwaki H, Takagi M, et al. Sarcomas of the oral and maxillofacial region: a review of 32 cases in 25 years. *Clin Oral Investig.* 2004;8:52–55.
9. Stuart A, Radhakrishnan J. Rhabdomyosarcoma. *Indian J Pediatr* 2004;71:331–337
10. Peters E, Cohen M, Altini M, Murray J. Rhabdomyosarcoma of the oral and paraoral

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region. Cancer 1989;63:963-6

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11. Kraus DH, Saenz NC, Gollamudi S, et al.
Pediatric RMS of the head and neck. Am J
Surg 1997;174:556-60

Rhabdomyosarcoma of the tongue. British
Journal Of Oral And Maxillofacial
Surgery(1994),32 183-186.

12. D. C. Doval, V. Kannan, R. S. Acharya,
G. Mukberjee, A. M. Shenoy, P. P. Bapsy.

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