

Alveolar Rhabdomyosarcoma of Oral Cavity – A Rare Case

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Abstract: Rhabdomyosarcomas are the malignant tumors of the striated skeletal muscles. Rhabdomyosarcomas are the most common soft tissue sarcoma of the children, adolescents and young adults. An eleven year old boy who presented with a painless progressive lump in the floor of the mouth since 4 months which was rapidly increasing in size without any systemic symptoms or signs of any metastatic spread. A wide excision of the lump was done and histopathology was reported to be an alveolar variant of rhabdomyosarcoma. Alveolar rhabdomyosarcomas account for almost 30% of all rhabdomyosarcomas. They have predilection for deep soft tissue of the extremities. The tumor may arise at other places also though they are rare. We report a rare case of alveolar rhabdomyosarcoma occurring in the oral cavity.

Key words: Rhabdomyosarcoma, Alveolar rhabdomyosarcoma, floor of mouth.

Introduction

Rhabdomyosarcomas are the malignant tumors of the striated skeletal muscles. Rhabdomyosarcomas are the most common soft tissue sarcoma of the children, adolescents and young adults. They are classified histologically into Embryonal, Botryoid, Alveolar, Pleomorphic varieties. They occur predominantly in three regions: the head and neck, genitourinary tract, and upper and lower extremities. Alveolar Rhabdomyosarcomas have predilection for the extremities [1]. We report a rare case of alveolar rhabdomyosarcoma occurring in the oral cavity in a eleven year old boy.

Case History

An eleven year old boy presented with a painless progressive swelling in the oral cavity in the floor of the mouth of four months duration. It was rapidly increasing in size and progressed from approximately 3 cm in the beginning to approximately 8 cm in size and was not associated with any other symptoms either locally or systemically. On Examination the swelling was 10 cm X 6 cm in size, occupying the floor of mouth, displacing the tongue to the left, non tender, regular margins and variegated in consistency and was not mobile (Figure 1). There was no cervical lymphadenopathy and the systemic examination was normal. FNAC of the swelling revealed a malignant spindle cell neoplasm (Figure 2), CT of the oral cavity was suggestive of malignant mass invading floor of the mouth. A metastatic work up was done which revealed no metastasis. Mandibulotomy was done and wide excision of the swelling was done. Histopathology was reported as alveolar rhabdomyosarcoma. Post operative course was uneventful. Chemotherapy and Radiotherapy was given to the patient post operatively. The patient is under follow

up since 4 years and there is no sign of residual tumor or any recurrence till date. (Figure3)

Legends



Figure1 Preoperative photograph of patient showing intraoral tumour.

Figure 2: Microphotograph of histopathological analysis of excised tumour.

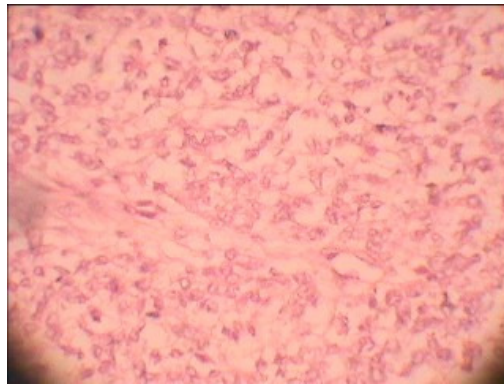


Figure3: Post operative photograph of patient.

Discussion

Rhabdomyosarcoma (RMS) is an aggressive malignant skeletal muscle neoplasm arising from embryonal mesenchyme. The head and neck is the most common site for this tumor in children.^[2] Alveolar rhabdomyosarcomas are relatively rare. Rhabdomyosarcomas occur predominantly in infants and children and somewhat less frequently in adolescents and young adults.^[1] Site predilections were found for the soft palate, maxillary sinus and alveolus, posterior mandibular region, cheek and lip and possibly tongue. The gingiva and floor of mouth were uncommon sites [3]. There are predominantly three types of Rhabdomyosarcomas:

1. Embryonal variant: Accounts for approximately 49% of all rhabdomyosarcomas and affects mostly children younger than 10 years of age, but it also occurs in adolescents and young adults. It is rare in patients older than 40 years of age [4].
2. Alveolar variant: It accounts for almost 30% of all rhabdomyosarcomas and tends to arise in patients of age group 10-25 years. It has predilection for deep soft tissue of the extremities. The tumor may arise at other places also though they are rare [5].
3. Pleomorphic variant: This is a rare variant which almost arises in adults older than 45 years. They arise mostly in the deep soft tissues of the extremities [6].

Treatment of the lesions is by a multidisciplinary approach. It consists of surgical removal of the tumor followed by multiagent chemotherapy with or without radiotherapy. Since RMS tend to metastasize to bone marrow, bone marrow aspiration should be a part of the staging procedure [7]. Metastasis may develop during the course of illness in approximately 20% of the cases with major sites being the lung, lymph node and bone marrow followed by heart, brain, meninges, pancreas, liver and kidney. Lungs are involved in nearly two thirds of patients of metastasis [8].

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