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An Exceptionally Rare Case of Palatal Botryoid Rhabdomyosarcoma in a Child

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Abstract
Background: Rhabdomyosarcoma (RMS) is the most common malignant mesenchymal tumor in children and adolescents, representing approximately 5% of all pediatric solid tumors. Although the head and neck region is a frequent site for botryoid RMS its occurrence in the oral cavity particularly on the palate is extremely rare.
Case presentation: We report the case of a 6-year-old patient who presented with a 3 cm nodular lesion on the palate. Histopathological analysis of the biopsy specimen revealed a malignant mesenchymal tumor showing a cambium layer of poorly differentiated, round or spindled cells arranged in a desmoplastic stroma. Immunohistochemical study demonstrated strong and diffuse positivity for Desmin with focal positivity for Myogenin. The Ki-67 proliferation index was estimated at 100%. These findings confirmed the diagnosis of botryoid variant of RMS.
Conclusion: This case underscores the diagnostic challenges associated with rare palatal botryoid RMS and highlights the necessity for early diagnosis using combined histopathological and immunohistochemical approaches. A personalized multimodal treatment strategy, including surgery, chemotherapy and radiotherapy when appropriate, is critical for achieving favorable outcomes. Reporting such rare cases is essential to enrich the literature and refine management protocols for these unusual presentations.

Keywords: Rhabdomyosarcoma, Palatal tumor, Pediatric malignancy, Rare localization, pathology, Immunohistochemistry, Botryoid

Introduction
Rhabdomyosarcoma (RMS) is the most common malignant mesenchymal tumor encountered in pediatric and adolescent populations, constituting approximately 5% of all solid tumors in these age groups. Despite its overall frequency, botryoid RMS arising in the oral cavity and particularly in the palatal region is exceedingly rare.

Case report
We report the case of a 6-year-old patient who presented with a nodular lesion of the palate measuring 3 cm in GA. A biopsy was performed. Histopathological study of the specimen showed a highly cellular malignant neoplasm arising beneath a focally ulcerated palatal mucosal surface (Fig. 1).

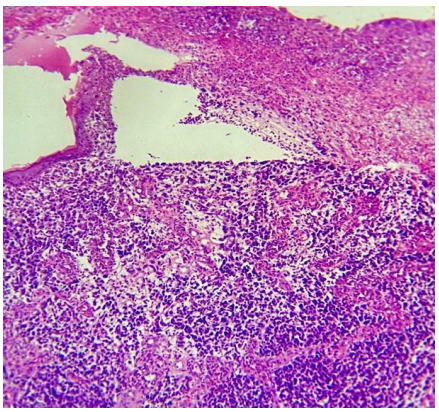

Fig 1: (H&E stain, x100): Ulcerated surface overlying loose sheets of undifferentiated cells.

The tumor was arranged in loose sheets of undifferentiated round to spindle-shaped cells**.** A dense, subepithelial zone of tightly packed tumor cells directly beneath the mucosa, giving the appearance of a "cambium" layer, was observed (Fig. 2).

Tumour cells displayed eosinophilic cytoplasm. The nuclei were hyperchromatic, atypical and often showing mitotic activity (Fig. 3). There were no evidence of keratinization or glandular differentiation. The background stroma was desmoplastic**,** with scattered inflammatory cells and areas of hemorrhage. An immunohistochemical study was performed. It showed diffuse and strong cytoplasmic staining for desmin (Fig. 4) and focal positivity for myogenin (Fig. 5), supporting skeletal muscle differentiation.

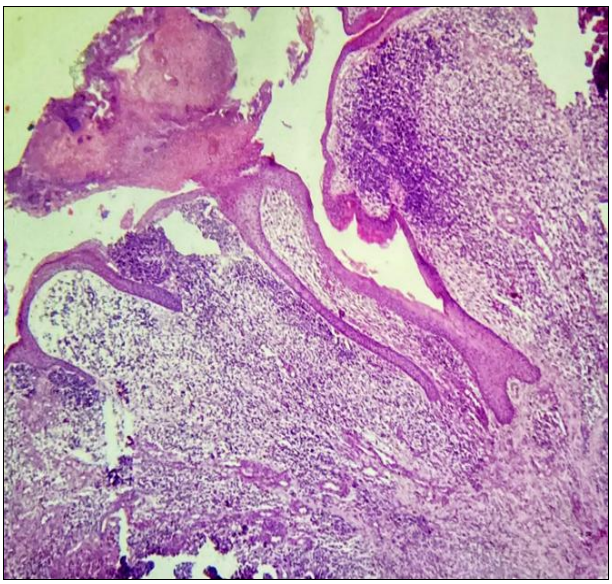

Fig. 2 (H&E stain, x40)**:** A hyper cellular cambial zone is observed beneath palatal mucosal surface.

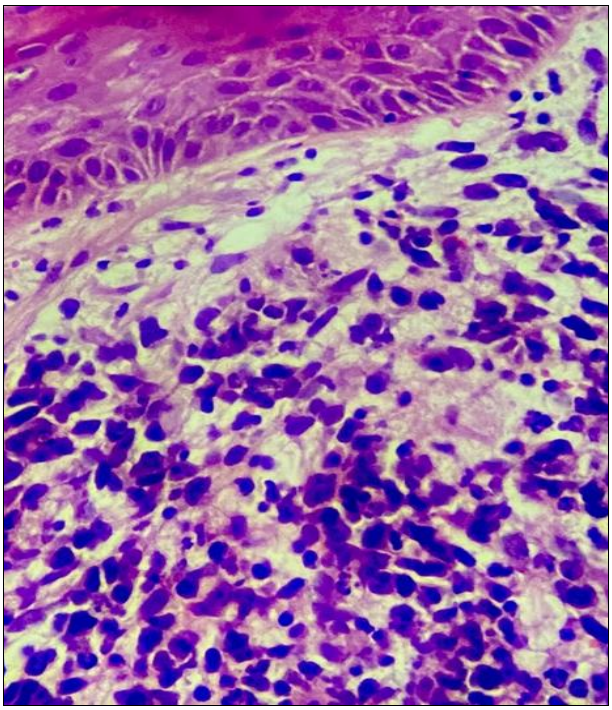

Fig 3: (H&E stain, x400): Undifferentiated round to spindle-shaped cells found subjacent to the epithelial covering.

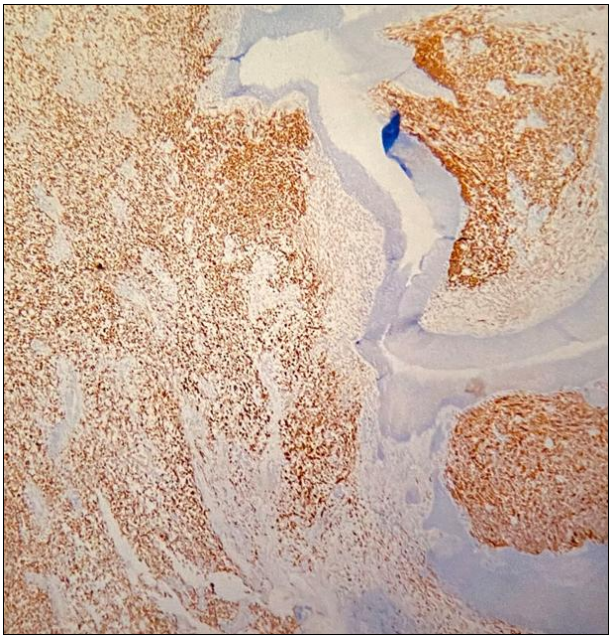

Fig 4: (H&E stain, x100): Markedly diffuse desmin positivity observed throughout the neoplastic cell population.

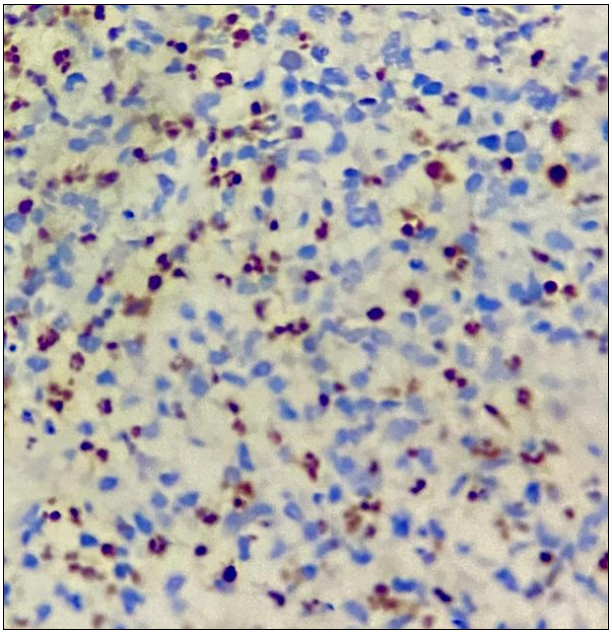

Fig 5: (H&E stain, x200): Myogenin expression detected as nuclear staining in scattered malignant cells

Ki-67 expression was estimated at 100%, indicating a high proliferative index. The tumor cells were negative for HHV8 and CD34 staining. These findings confirmed the diagnosis of botryoid variant of RMS.

Discussion

Rhabdomyosarcoma (RMS) is the most common malignant mesenchymal tumor of childhood and adolescence and accounts for about 5% of all solid tumors in these age groups [1].

The most common localizations of RMS in children are head and neck region (40%), genitourinary system (25%) and limbs (20%) [2].

RMS comprises several histological subtypes, including embryonal, alveolar, pleomorphic, and spindle cell/sclerosing variants with the embryonal type being the

most frequently encountered [3–5].

Although RMS classically occurs in sites such as the orbit, head and neck or genitourinary tract with the botryoid variant most frequently occurring on mucosa-lined surfaces like the vagina or bladder its manifestation in the oral cavity, particularly in the palatal region, is exceptionally uncommon and is generally reported only as isolated case reports [6].

Children's rhabdomyosarcoma of the palate is extremely rare. Individual data about its exact prevalence do not exist as it is unusual and the vast majority of the present case reports are not connected but standalone.

Literature review reports a paucity of documented instances of this specific localization. Most head and neck rhabdomyosarcomas occur in the orbit, parameningeal sites (such as nasopharynx, nasal cavities, and paranasal sinuses) or non-parameningeal sites with the palate being an infrequent site [6].

Botryoid RMS is macroscopically described as nodule-shaped polypoid mass, which is found in the mucosa-lined organs of the nasopharynx, paranasal sinus, genitourinary and gastrointestinal tracts [7, 8].

Microscopically, the tumor is made up of small, round, and ovoid cells with hyperchromatic nuclei and scant cytoplasm. The cells are loosely dispersed in a myxoid background and demonstrate variable stages of skeletal muscle differentiation. Moderate numbers of mitotic figures are seen, reflecting the high proliferative potential of the tumor. Strap cells with eosinophilic cytoplasm and cross-striations indicating rhabdomyoblastic differentiation are identified in some cases [9].

Histologically, botryoid rhabdomyosarcoma demonstrates differentiation of skeletal muscle, typically confirmed by immunohistochemical markers such as desmin, myogenin, and MyoD1 [10, 11].

Molecular studies may also be useful in the diagnosis. While embryonal rhabdomyosarcoma, including botryoid type, does not have a typical translocation like PAX3-FOXO1 or PAX7-FOXO1, present in alveolar rhabdomyosarcoma, loss of heterozygosity at 11p15.5 is usually detected [12].

Differential diagnoses include other small round blue cell tumors such as Ewing sarcoma, neuroblastoma and lymphoma. Therefore, immunohistochemistry is essential in distinguishing botryoid rhabdomyosarcoma from these conditions [13].

The standard treatment regimen consists of multimodal management with surgery, chemotherapy and, in a small selected number, radiotherapy. Due to the susceptible anatomic site, neoadjuvant chemotherapy is most commonly used to decrease the tumor size prior to surgical therapy for maintenance of function as well as local control. The approach has greatly improved the survival and 5-year overall survival rates are over 70% in certain series [13, 12].

Conclusion

Though botryoid rhabdomyosarcoma is rare, particularly in the palate, early diagnosis and personalized multimodal management strategy can lead to favorable outcomes. Such rare cases should be reported to add to the literature and help define clearer management in the future for these unusual presentations.

Abbreviations

- **Ac** – Antibody
- **CD34** – Cluster of Differentiation 34
- **GA** – Greatest Axis
- **HHV8** – Human Herpesvirus 8
- **Ki 67** – Cellular Proliferation Marker
- **PAX3-FOXO1** – Paired Box 3-Forkhead Box O1 Fusion Gene
- **PAX7-FOXO1** – Paired Box 7-Forkhead Box O1 Fusion Gene
- **RMS** – Rhabdomyosarcoma

Declaration

Ethics Approval and Consent to Participate

I declare no conflicts of interest. This work was conducted in full compliance with the code of ethics under the supervision of the Medical and Ethics Committee of La Rabta Hospital, Tunisia.

Data and Material Availability

All data used in this study were obtained from patients' medical records, available in the archives of our institution.

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Conflicts of Interest

None.

Authors' Contribution

- Data collection
- Literature review
- Drafting of the manuscript

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Not applicable.

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