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Pediatric Nasopharyngeal Rhabdomyosarcoma: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Rhabdomyosarcomas is a soft tissue tumor with a highly invasive malignant cells that originate of the skeletal muscle cells, specially occurs in the head and neck regions, the presentation in the parameningeal region, including the paranasal sinuses and nasal cavity, the presentation in the nasopharynx is infrequent.

The diagnosis is confirmed by appropriate immunohistochemical stains, while conventional radiology is used for evaluating the primary tumor, determining the extension to nearby organs, and detecting any potential metastases.

The treatment of nasal rhabdomyosarcoma is a real challenge given the unknown biological behavior of the pathology, the treatment is essentially based on surgery, chemotherapy and radiotherapy.

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Keywords: Nasopharyngeal rhabdomyosarcoma; immunohistochemical stains; CT scan.

1. INTRODUCTION

Rhabdomyosarcomas (RMS) are a tumors with a highly invasive malignant cells that originate of the skeletal muscle cells, about 35% of rhabdomyosarcoma arise in the head and neck regions, with approximately 50% occur in the parameningeal region, counting the sinuses and nasal cavities. Most tumors appear before 5 years but can affect patients of any age.

RMS is subdivided into 4 subtypes: embryonal, alveolar, spindle cell/sclerosing and pleomorphic, according to the current WHO classification.

We describe a case of embryonal nasopharyngeal RMS in a 4-year-old patient treated with chemotherapy and radiotherapy.

2. CASE PRESENTATION

We present the case of a 4 years old patient, with no significant medical history, was referred to our department for a cerebral and facial CT scan in view of a symptomatology that began 3 months ago, consisting of nasal congestion, mucopurulent rhinorrhea and voice changes, without any history of odynophagia or dysphagia.

The clinical examination revealed no neurological or ophthalmological disorders, there was no obvious lymphadenopathy.

The Computed tomography (CT) revealed a 53 × 33 × 49 mm tissue-dense tumor occupying the right nasal fossa and right maxillary sinus, dissolving their medial wall, and extending into the nasopharynx. The tumor spreads into the parapharyngeal space but does not invade the retrostylial or prevertebral space.

The histological sections of the nasopharyngeal process showed an embryonal rhabdomyosarcoma composed of primitive skeletal mesenchymal cells within a myxoid stroma, tumor cells show a significant atypia.

Immunohistochemistry the tumor cells express strongly the Desmin and Myogenin antibody.

The patient received four cures of combination chemotherapy (actinomycin D, cyclophosphamide, vincristine) followed by radiation therapy (4000 cGy) to the sinonasal region.

The follow-up clinical exam and CT scan reveals an increase in the size of the lesion, extending into the oropharynx, confirming a lesion advancement.

The patient received two additional cures of chemotherapy, with the introduction of another molecule (Adriamycin) to the initial chemotherapy treatment.

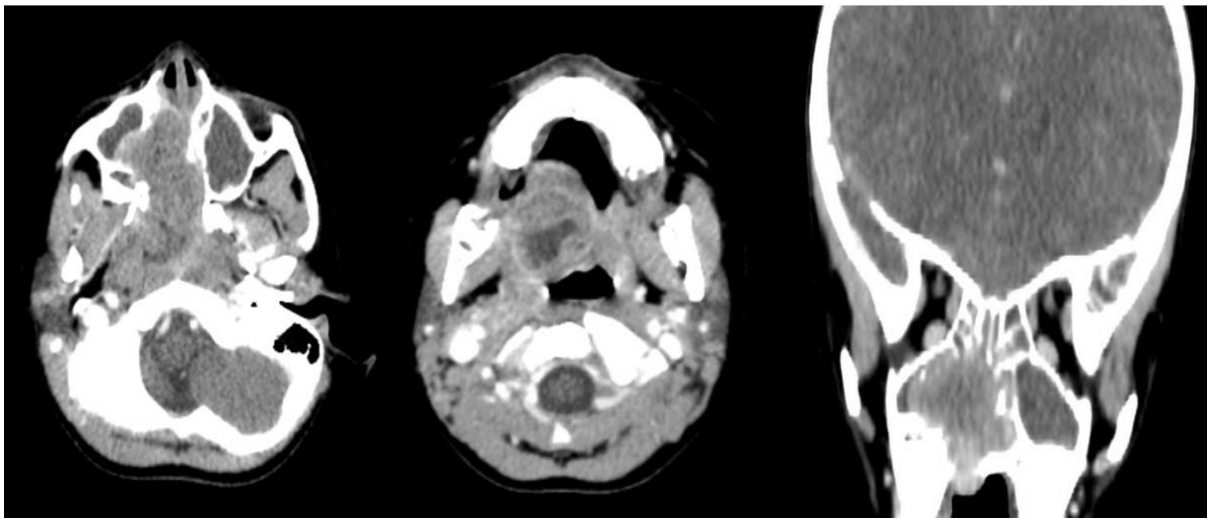

Fig. 1. Axial CT scan showing a tissular process centered on the right maxillary sinus extending to the homolateral parapharyngeal space

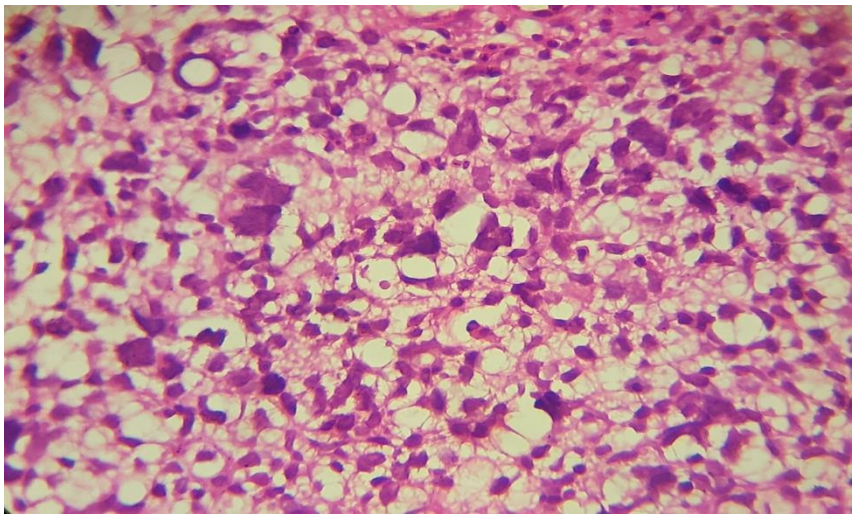

Fig. 2. Histological slide showing an Embryonal Rhabdomyosarcoma composed of primitive skeletal mesenchymal cells within a myxoid stroma. Tumor cells show a significant atypia. HEX10 (magnificationX 10 under the microscope)

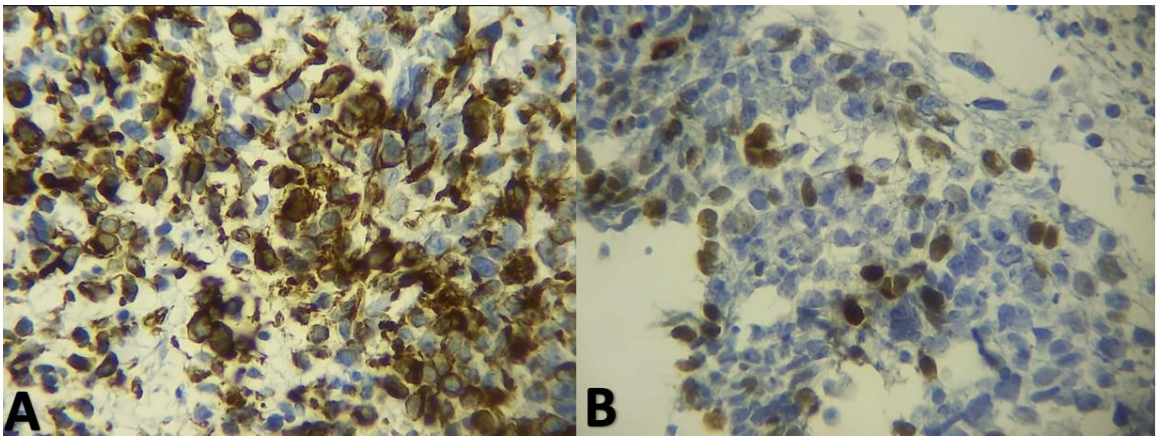

Fig. 3. At immunohistochemistry, tumor cells express strongly the Desmin antibody (A), and Myogenic antibody (B)

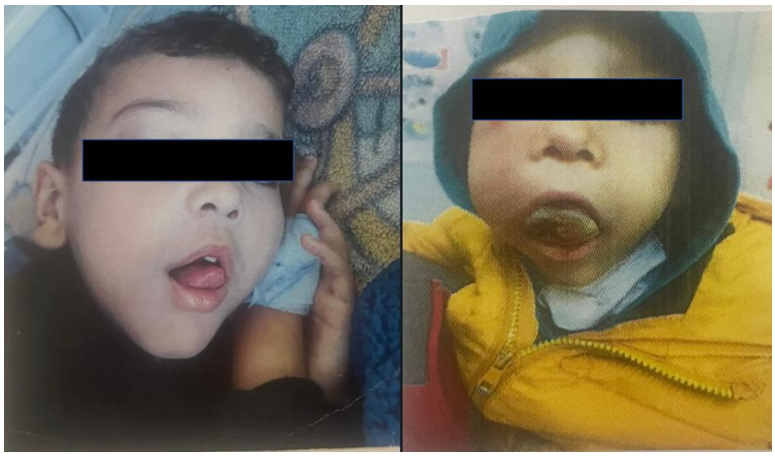

Fig. 4. Clinical examination demonstrates an increase in the size of the process, with invasion of the soft palate

3. DISCUSSION

RMS is a malignant soft tissue neoplasm arising from the skeletal muscle cells, described initially by Weber in Virchow's Archives in 1854 [1], about 35% of rhabdomyosarcoma arise in the head and neck regions, with approximately 50% occur in the parameningeal region, counting the sinuses and nasal cavities [2]. Typically emerging in children under the age of 10 years [3] but can develop at any age, although its presence in adults is exceptionally rare [4]. Rhabdomyosarcoma (RMS) has the potential to develop in various anatomical locations throughout the body. However, the most frequent primary tumor sites are found in the head and neck region with a prevalence of 35%, followed by genitourinary approximately 25% usually in the bladder, prostate, or vagina [5,6].

RMS, similar to all soft-tissue carcinomas, mostly without obvious clinical signs in the early stages, the majority of patients consult at an advanced stage, with signs depending mainly on the location of the tumor, ranging from nasal congestion, rhinorrhea, voice changes, nausea, recurrent otitis and headache, to a mass generally invading the various deep spaces of the face causing more alarming clinical signs such as dysphagia, dyspnea or even other symptoms secondary to metastasis [7,8]. Taking into account that 15% already have distant metastases at the time of diagnosis (9), the lung stands as the most prevalent site for metastasis, followed by occurrences in the bone marrow, other common sites include metastases to bones or distant lymph nodes [10].

The diagnosis specially based on the immunohistochemical profile, the embryonic variant is the most common RMS subtype, predominating in the pediatric patients comprising 58% of all cases, while alveolar RMS is more common in adolescents and young adults [11, 12].

Microscopically, the embryonic RMS is characterized by hypocellular and hypercellular areas composed of striated muscle cells in various stages of differentiation, surrounded by a myxoid stroma. Immunohistochemical staining is necessary to confirm the diagnosis, revealing tumor cells' reactivity to desmin, myoglobin, vimentin, actinMyoD1, and myogenin,the absence of staining for cytokeratin, S-100, and epithelial membrane antigen helps eliminate other potential differential diagnoses of RMS [13, 14].

Conventional radiology plays a pivotal role in evaluating the primary tumor, determining its extension to nearby organs, and detecting metastases; these assessments are crucial for formulating an appropriate therapeutic approach. MRI offers superior resolution for soft tissues, Fat suppression sequence rise the sensitivity of this approach to tumor spread, MRI also allows better distinguishing of tumor, muscle, secretion, and mucous thickening and it is particularly effective in assessing perineural and perivascular structures, as well as detecting signs of intracranial extension.

CT scan is mainly used to detect extension to bones, the bone remodeling indicates benign or slow-growing tumors, whereas bone destruction and soft tissue loss indicate malignancy, and also to detect metastasis which will have an impact on therapeutic [15,16].

The prognosis for patients with rhabdomyosarcoma is influenced by several factors, including age (the prognosis is less favorable in children younger than 1 year old or older than 10 years old), tumor size, origin site, (survival rates in cases involving the nasopharynx have been reported to be less favorable due to the high risk of tumor spreading to the central nervous system), the histologic type (the embryonic type is associated with a better post-treatment result than the alveolar type), and the presence of metastasis [17, 18, 19].

Advancements in radiotherapy, chemotherapy, surgery have improved the management of ERMS. Presently a multidisciplinary strategy incorporating surgery, surgical resection is usually performed prior to chemotherapy if it does not result in disfigurement, functional impairment or organ dysfunction. If this is not possible, only an initial biopsy is performed [20, 21].

In our case the patient did not improve following combined chemotherapy and radiotherapy, surgical resection is not an option, given the size of the process and its extension.

4. CONCLUSION

Pediatric nasopharyngeal rhabdomyosarcoma is a quit frequent soft tissue tumor, the diagnosis is

confirmed by an appropriate immune-histochemical stains, while conventional radiology is used for evaluating the primary tumor, determining the extension to nearby organs, and detecting any potential metastases.

CONSENT

Written informed consent was obtained from the parents of patients for the publication of this case report.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Weber CO, Virchow R. Anatomische Untersuchung einer hypertrophischen Zunge nebst Bemerkungen über die Neubildung quergestreifter Muskelfasern. Virchows Arch 1854;7:115–25.
2. Hicks J and Flaitz C: Rhabdomyosarcoma of the head and neck in children. Oral Oncol. 38:450–459. 2002.
3. L Wexler, W Meyer, L Helman: Rhabdomyosarcoma and the Undifferentiated Sarcomas 2006 ed 5 Philadelphia, PA Principles and Practice of Pediatric Oncology, Lippincott Williams & Wilkins
4. A Ferrari, P Dileo, M Casanova, etal: Rhabdomyosarcoma in adults. A retrospective analysis of 171 patients treated at a single institution Cancer 98: 571– 580,2003
5. Egas-Bejar D, Huh WW. Rhabdomyosarcoma in adolescent and young adult patients: current perspectives. Adolesc Health Med Ther 2014;5:115–25.
6. Linardic CM, Wexler LH: Rhabdomyosarcoma: Epidemiology and Genetic Susceptibilty. In Pizzo and Poplack's Pediatric Oncology, 8th ed., edited by SM Blaney, LJ Helman, PC Adamson. Philadelphia, Wolters Kluwer, 2021, p. 693.
7. Sultan I, Qaddoumi I, Yaser S, Rodriguez-Galindo C, Ferrari A. Comparing adult and pediatric rhabdomyosarcoma in the surveillance, epidemiology and end results program, 1973 to 2005: an analysis of 2,600 patients. J Clin Oncol 2009;27:3391–7. Available:https://doi.org/10.1200/JCO.2008.19.7483.
8. Ferman SE. Survival analysis of pediatric patients with rhabdomyosarcoma: 18 years of experience of the National Cancer Institute—RJ. This Presented to the Faculty of Medicine of the University of Sao Paulo, Sao Paulo; 2005.
9. Crist W, Gehan EA, Ragab AH, Dickman PS, Donaldson SS, Fryer C, Hammond D, Hays DM, Herrmann J, Heyn R et al. The Third Intergroup Rhabdomyosarcoma Study. Journal of Clinical Oncology. 1995;13:610-630. Available:https://doi.org/10.1200/JCO.1995.13.3.610
10. Oberlin O, Rey A, Lyden E, Bisogno G, Stevens MC, Meyer WH, et al. Prognostic factors in metastatic rhabdomyosarcomas: Results of a pooled analysis from United States and European cooperative groups. J Clin Oncol. 2008;26:2384–9.
11. Mondal PK, Pal I, Misra S, Biswas S, Bera SP. Rhabdomyosarcoma of Nose, Nasopharynx and Paranasal Sinuses. Indian Journal of Otolaryngology and Head & Neck Surgery. 2009;61:317-319. Available:https://doi.org/10.1007/s12070-009-0090-9
12. WHO Classification of Tumors Editorial Board: WHO Classification of Tumors. Volume 3: Soft Tissue and Bone Tumors. 5th ed, 978-92-832-4502-5, IARC Press; 2020.
13. Kumar S, Perlman E, Harris CA, Raffeld M, Tsokos M. Myogenin Is a Specific Marker for Rhabdomyosarcoma: An Immunohistochemical Study in Paraffin-Embedded Tissues. Modern Pathology. 2000;13:988-993. Available:https://doi.org/10.1038/modpathol.3880179
14. Gibson MC, Nasiri N. Rhabdomyoma. Vienna: WikiDoc; 2019. [cited 2023 Jan 4]. Available:https://www.wikidoc.org/index.php/Rhabdomyoma. [Google Scholar]20 Gibson MC, Nasiri N. Rhabdomyoma. Vienna: WikiDoc; 2019 [cited 2023 Jan 4].
15. Kobi M, Khatri G, Edelman M, Hines J. Sarcoma Botryoides: MRI Findings in Two Patients. J Magn Reson Imaging. 2009;29:708–12.
16. Burkat CN, Lucarelli MJ. Rhabdomyosarcoma masquerading as

acute dacryocystitis. Ophthalmic Plastic & Reconstructive Surgery. 2005;21:456-458. Available:https://doi.org/10.1097/01.iop.0000181349.79287.12

17. Weiss SW, Goldblum JR. (Eds.). Rhabdomyosarcoma. Enzinger and Weiss's Soft Tissue Tumors. 4th Edition. C. V. Mosby, St. Louis. 2001;785-835.
18. Oberlin O, Rey A, Lyden E, et al: Prognostic factors in metastatic rhabdomyosarcomas: Results of a pooled analysis from United States and European cooperative groups. J Clin Oncol 26(14):2384–2389, 2008. DOI: 10.1200/JCO.2007.14.7207
19. Oberlin O, Rey A, Lyden E, et al.: Prognostic factors in metastatic rhabdomyosarcomas: Results of a pooled analysis from United States and European cooperative groups. J Clin Oncol. 2008;26(14):2384-9.
20. Donaldson SS, Meza J, Breneman JC, et al. Results from the IRS-IV randomized trial of hyperfractionated radiotherapy in children with rhabdomyosarcoma--a report from the IRSG. Int J Radiat Oncol Biol Phys. 2001;51(3):718-28.
21. Guo LM, Zhao H, Chi FL, Yan YH. Clinical analysis of 17 cases of rhabdomyosarcoma in ear nose and throat. Journal of Clinical Otorhinolaryngology. 2001;15358-359. Available:http://dx.chinadoi.cn/10.3969/j.issn.1001-1781.2001.08.010

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