Case report

Pediatric nasopharyngeal rhabdomyosarcoma: a case report

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

introduction:

Rhabdomyosarcomas (RMS) are a tumors with a highly invasive malignant cells that originate of the skeletal muscle cells, specially occurs in the head and neck regions, it represents 4-6% of all malignant tumors in the pediatric population. The paranasal sinuses, nasal cavity, nasopharynx and are the most common sites for all RMS subtypes. most Tumors appear before 5 years but can affect all 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 child treated with induction chemotherapy, radiotherapy, and adjuvant chemotherapy.

Case report:

We present the case of a 4 years old boy, 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.

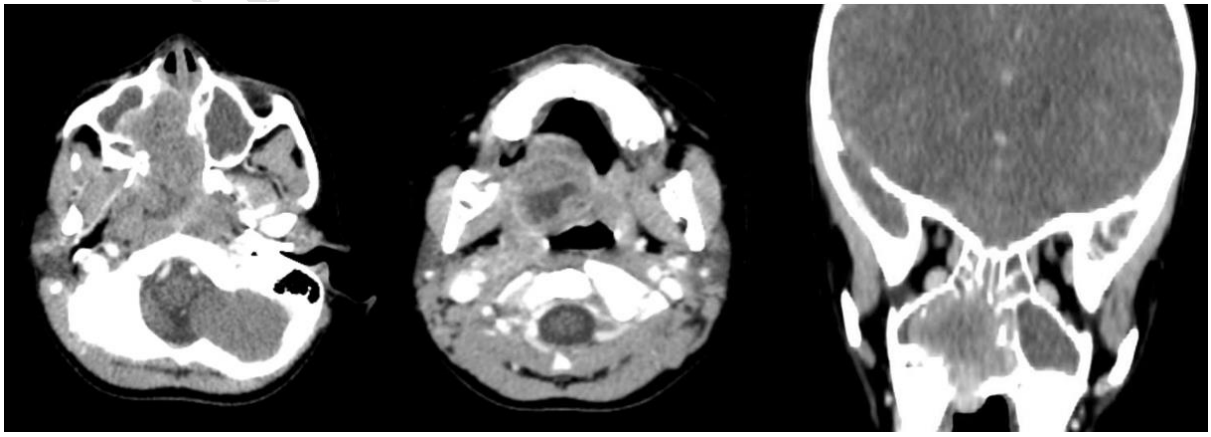

Figure 1: axial CT scan showing a tissular process centered on the right maxillary sinus extending to the homolateral parapharyngeal space.

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.

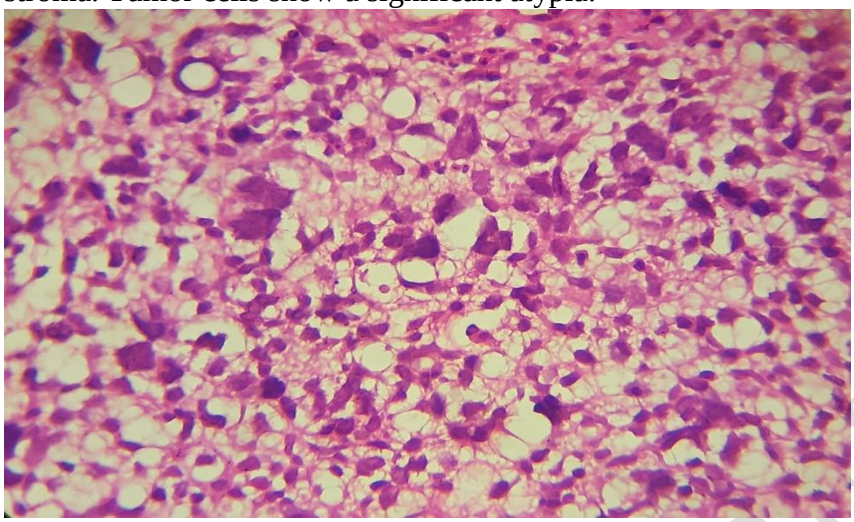

Figure 2: Histological slide showing an Embryonal Rhabdomyosatcoma compoed of primitive skeletal mesenchymal cells within a myxoid stroma. Tumor cells show a significant atypia. HEX10

Immunohistochemistry the tumors cells express strongly the Desmin and Myogenic antibody.

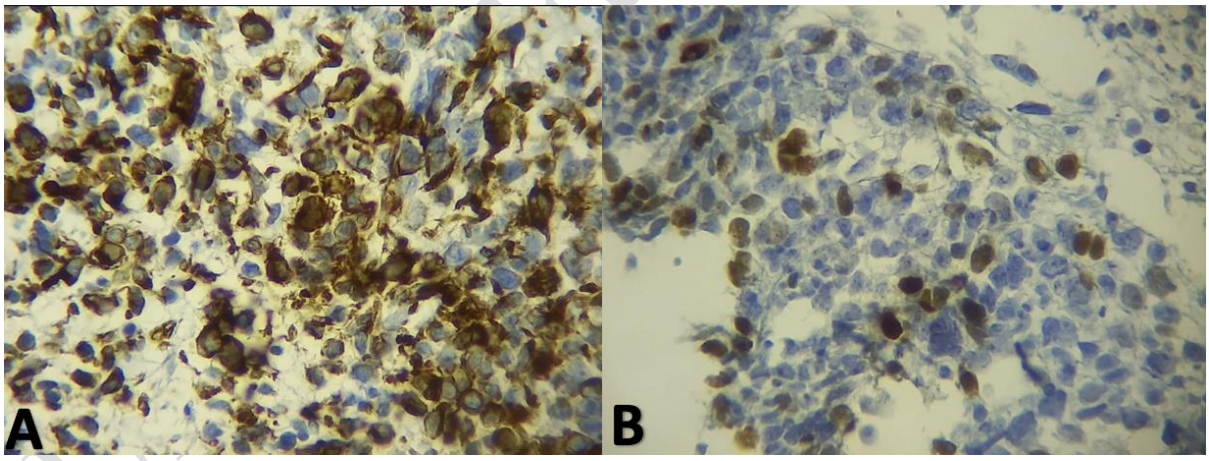

Figure 3: At immunohistochemistry, tumor cells express strongly the Desmin antibody (A), and Myogenic antibody (B).

The patient received a chemotherapy neoadjuvant and adjuvant radiotherapy after the elimination of cells or metastatic clusters, through a thoraco-abdominal CT scan, myelogram, osteo-medullary biopsy, and Positron Emission Tomography-Computed Tomography (PET-CT),

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

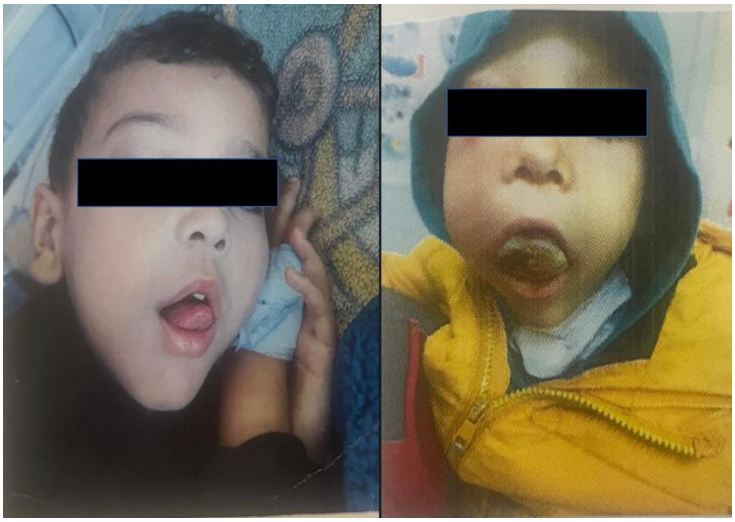

Figure 5: clinical examination demonstrates an increase in the size of the process, with invasion of the soft palate.

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),** It represents approximately 4-6% of all pediatric malignancies **(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%, typically in the orbit or nasopharynx: most common in school-aged children, followed by genitourinary approximately 25% usually in the bladder, prostate, or vagina, typically occurring in infants and newborns,
The extremity site about 20% predominantly in adolescents, and about 20 % in trunk/various sites. **(5,6)**

RMS, similar to all soft-tissue carcinomas, can present a range of clinical symptoms, mostly without obvious clinical signs in the early stages, most patients consult at an advanced stage, with signs depending mainly on the location of the tumor ranging from nasal congestion, rhinorrhea,nausea, headache, recurring otitis, voice changes, 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)**

A tissue biopsy is crucial for diagnosis specially the immunohistochemical profile, the 2020 WHO classifies rhabdomyosarcomas into four subtypes: embryonal, alveolar, pleomorphic and spindle cell/sclerosing, 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 it is characterized by hypocellular and hypercellular areas composed of striated muscle cells in various stages of differentiation, surrounded by a myxoid stroma, imunohistochemical staining is necessary to confirm the diagnosis. Immunohistochemical reveals 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 of 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 scans 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.

Conclusion:

Embryonal rhabdomyosarcoma is a soft tissue tumor, the presentation in the nasopharynx is quite rare, the diagnosis is confirmed by an appropriate immunohistochemical stains, while conventional radiology is used for evaluating the primary tumor, determining the extension to nearby organs, and detecting any potential metastases.

Informed Consent:

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

Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Reference:

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