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Case Report

Adolescent' Rhabdomyosarcoma of the Infra-Temporal Fossa: Case Report and Review of the Literature

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ABSTRACT

Rhabdomyosarcoma (RMS) is a malignant tumor of striated muscle. It is the most common malignant mesenchymal tumor. The prognosis of these tumors depends on the absence of metastasis at diagnosis, the age of the child, and the location, volume and operability of the tumor. The etiology of RMS is unknown, but certain syndromes such as neurofibromatosis type 1, Li-Fraumeni syndrome, Beckwith-Wiedman syndrome and Costello syndrome have been incriminated. Therapeutic management is based on surgical treatment whenever possible, as well as postoperative chemotherapy/radiotherapy. In this context, we present the case of a very advanced RMS of the infra-temporal fossa in a 16-year-old child.

Keywords: Rhabdomyosarcoma; Infra-temporal fossa; Case report; Surgery

Introduction

Developed from primitive mesenchymal cells involved in striated muscle differentiation, rhabdomyosarcoma (RMS) can occur anywhere in the body. In terms of extra-cranial solid tumors in children, RMS is the third most common after neuroblastoma and nephroblastoma[1].

Rhabdomyosarcomas that occur in the head and neck region, the most frequent site is the orbit, followed by the nasopharynx and paranasal sinuses. The infratemporal fossa, on the other hand, is an extremely rare site of such tumors. It is a highly malignant tumor, distinguished from other sarcomas by its locoregional aggressiveness. Early diagnosis and multidisciplinary management are therefore crucial[2].

In this context, we report the case of an adolescent who presented to our department with a symptomatology suggestive of rhabdomyosarcoma of the infra-temporal fossa. Given its location, a diagnostic biopsy was performed under general anesthesia before referring the patient for chemotherapy/radiotherapy.

Case Report

We present the case of a 16-year-old adolescent, with no particular medical history. Admitted to our ENT department for a right parotid swelling, with no other associated signs, notably no dyspnea, dysphagia or signs of facial paralysis. The patient's general condition remained stable. On physical examination, the parotid mass is firm, fixed and sensitive to palpation, with no inflammatory signs **(Figure 1)**. Rhinoscopy shows a bulging of the nasopharynx. Endo-buccal examination shows a bulging of the soft palate **(Figure 2)**.

Cervical examination showed no abnormalities, notably no palpable lymph nodes.

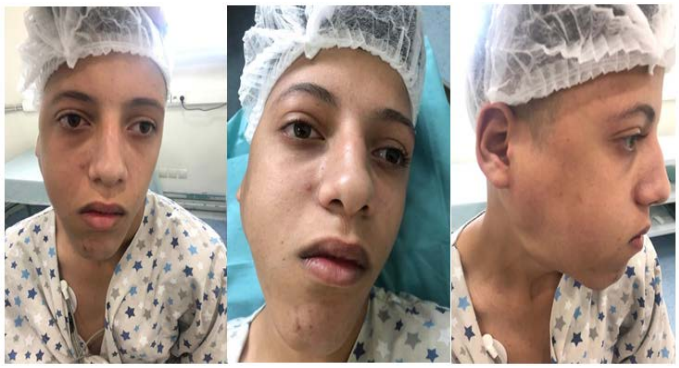


Figure 1: Image of the patient showing the tumefaction opposite the parotid region.

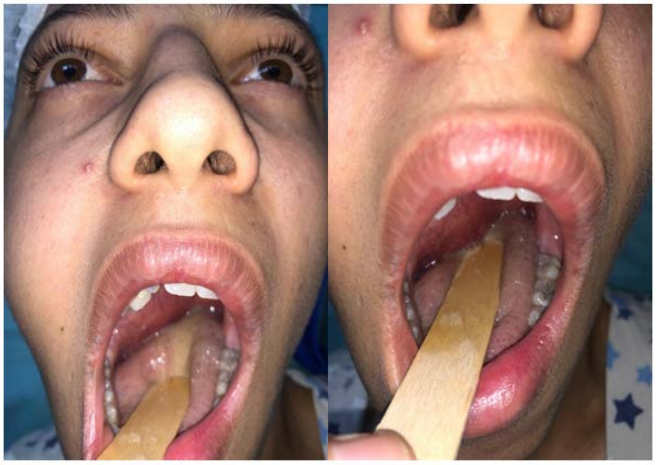


Figure 2: Image showing a bulging of the soft palate.

The patient then underwent MRI of the face, which revealed a voluminous, largely necrotic tissue lesion in the right infra-temporal fossa, measuring 94 mm in long axis, with irregular contours in T1 hyposignal, discrete T2 hyper signal, diffusion hypersignal with low ADC, intensely and heterogeneously enhancing after injection of gadolinium, delineating areas of necrosis. Superiorly and medially, it infiltrates the right parapharyngeal space and the nasopharynx, partially filling its lumen. Superiorly and laterally, it compresses the homolateral parotid gland and reaching the subcutaneous soft tissue **(Figure 3)**.

Inwardly and inferiorly, it infuses the hypopharynx and the proximal part of the larynx, which remain permeable **(Figure 5)**. Downwards and outwards, it infiltrates the ascending branch of the mandible, with bone lysis **(Figure 4)**. Anteriorly, it comes into contact with the right maxillary sinus **(Figure 6)**. Posteriorly, it fills the pre-stylial and retro-stylial space and infiltrates the pre-vertebral muscles on the homolateral side. At the top, it comes into contact with the floor of the orbit, with bone lysis of the anterior and base of the skull, and infiltrates the homolateral temporal lobe **(Figures 7 and 8)**. Below, it infiltrates the right submandibular gland and the base of the tongue **(Figure 5)**.

On this basis, the team decided to perform a biopsy under general anesthesia for histopathological confirmation, followed by chemotherapy/radiotherapy. The evolution was marked by a clear reduction in tumor volume. The patient is still under close follow-up to detect any recurrence or complication related to the tumor or radio-chemotherapy.

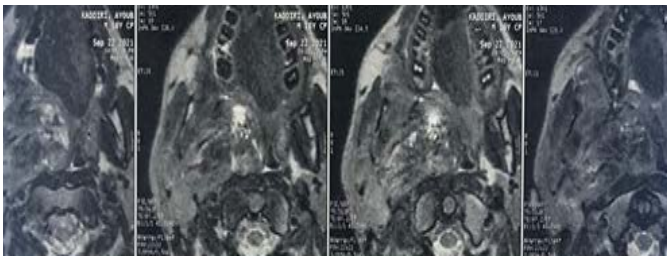


Figure 3: MRI on axial section showing the tumor that infiltrates the right parotid, the right parapharyngeal space and the nasopharynx, partially filling its lumen.

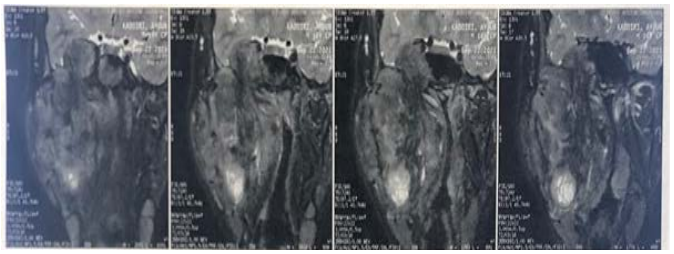


Figure 4: MRI on coronal section showing the tumor infiltrating the ascending branch of the mandible, with bone lysis.

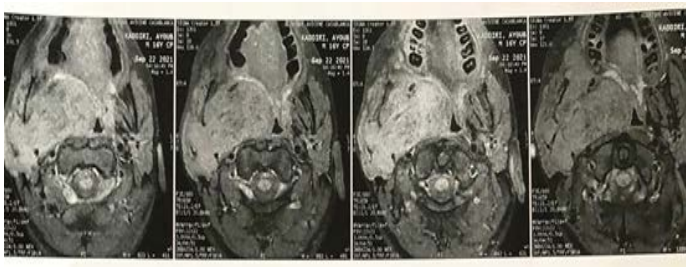


Figure 5: MRI on axial section showing the infiltration of the base the tongue and the hypopharynx.

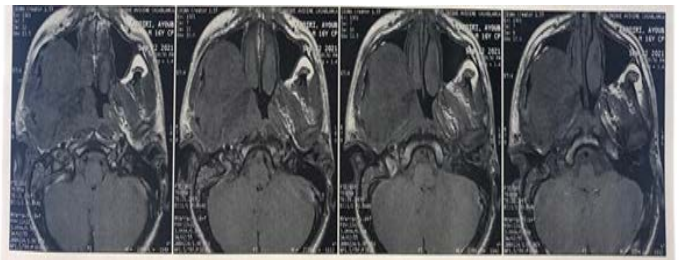


Figure 6: MRI on axial section showing the infiltration of the right maxillary sinus.

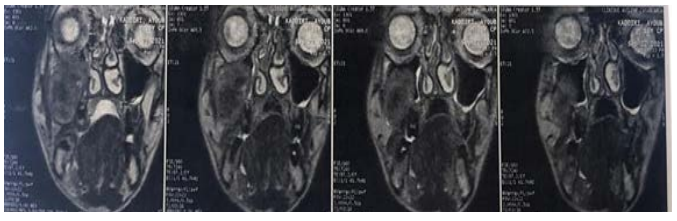
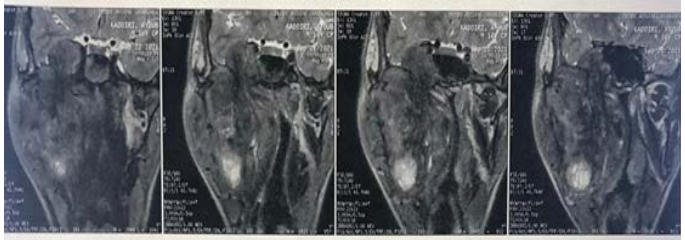


Figure 7: MRI on coronal section showing the tumor that comes into contact the floor of the orbit.



Figu.re 8: MRI on coronal section showing the tumor infiltrating the base of the skull, and the homolateral temporal lobe

Discussion

Rhabdomyosarcoma (RMS) represents the most prevalent mesenchymal tumor in children and adolescents. This is a malignant tumor of unknown etiology with more or less marked striated muscle differentiation, of mesenchymal origin. It can develop anywhere in the body, including sites where striated muscle tissue does not normally exist. The most common sites are the head and neck (40%). There are 350 new cases of rhabdomyosarcoma diagnosed every year in the United States, with an annual incidence estimated at 8/1000000[1,2].

About two-thirds of RMS cases are diagnosed in children under the age of six years, with two peaks in incidence under five and in adolescents[3].

Epidemiological data suggest that genetic factors play an important role in the etiology of at least some of these sarcomas. A number of specific genetic syndromes may be associated neurofibromatosis type 1, Rubinstein-Tayebi syndrome, Wiedemann-Beckwith syndrome, Costello syndrome, Noonan syndrome and Gorlin basic nevus syndrome. Among a series of 33 cases of sporadic RMS, evidence of a germline p53 mutation was found in 3 of 13 children under 3 years of age[3-5].

This tumor is distinguished by rhabdomyoblasts, slightly elongated cells with intracellular cross streaks and eosinophilic cytoplasm[6]. Diagnosis relies to a crucial extent on immunohistochemical analysis. RMS express Vimentin, testifying to the conjunctival origin of cell proliferation, smooth muscle actin (SMA) of striated muscle, and desmin, testifying to an intermediate filament between smooth and skeletal muscles. Myo-D1 or Myf-3 are specifically expressed in the nucleus of rhabdomyosarcoma cells in 80% of cases. Embryonal rhabdomyosarcoma is confirmed by immunohistochemistry with positive markers desmin, HHF35, possibly myoglobin and MyoD1; some cells may be cytokeratin positive and PS100 positive[7,8].

The most common molecular signature of ASMR involves the chromosomal translocation t(2 ;3)(q35 ;q14)[9]. In the case of embryonal rhabdomyosarcomas, differential diagnosis is often made with other round cell tumors, such as neuroblastoma, lymphoma, PNET (Peripheral Neuro Ectodermal Tumors), synovial-sarcoma or rhabdoid tumors[10].

The primary site of rhabdomyosarcoma has long been recognized as a key prognostic factor. It is also an important element to consider in the therapeutic strategy for RMS, as it determines the quality of the local procedure, with the existence of microscopic or macroscopic residue to a greater or lesser extent. Orbit, Non-para meningeal head and neck, Para meningeal head and neck, genitourinary organs and others (intra-thoracic, intra-abdominal-pelvic, walls and perineum) are all potential sites of localization[11,12].

Para-meningeal sites, involve the base of the skull (nasopharynx, sinuses, middle ear, infra-temporal fossa and pterygopalatine), with a propensity to erode adjacent bony structures and infiltrate intracranial structures by contiguity[13].

The American “IRS” system (Intergroup RMS Study, United States) has developed a classification that takes into account tumor operability. Group 1 or Localized disease, microscopic resection, confined to the muscle or organ of origin without lymph node invasion. Group 2 where macroscopic resection is total but microscopic residue, regional disease (extending beyond the muscle or organ of origin), completely resected or with lymph node involvement. Group 3 Incomplete resection with macroscopic residue or simple biopsy. Group 4 distant metastases at diagnosis[10].

Concerning which imaging modality should be used when a mass lesion of the infratemporal fossa is suspected, Levine et al reported a representative case series in which malignancies that involved the infratemporal fossa were better defined by axial MRI than by CT imaging[14]. (18).

It is important to consider at the time of diagnosis both the possibility of a cure after well-managed treatment, and the acute and late toxicities of the therapies used. Therapeutic management of these tumors therefore requires a multidisciplinary approach involving the oncologist, surgeon, radiotherapist, radiologist and pathologist[3].

The surgical approach not only allows the biopsy required for diagnosis, but also for complete primary removal of the tumour in sites where extended removal would not compromise functional and/or aesthetic prognosis, such as the limbs and trunk, or for a secondary removal leading to better local control after reduction of tumour volume by chemotherapy and/or radiotherapy. In head-neck cases, surgery is often limited to diagnostic surgical biopsy[15,16].

Chemotherapy has transformed the prognosis of these tumors. These drugs are always used in combination, depending on the protocol. Drugs with proven efficacy include actinomycin D, cyclophosphamide, vincristine, cisplatin, carboplatin, dacarbazine (DTIC) and doxorubicin. More recently, ifosfamide and etoposide have been added to the therapeutic arsenal. Duration and intensity of treatment vary according to initial prognosis and response to therapy[17].

Radiotherapy’s aim is to achieve local control, or to consolidate that achieved by chemotherapy. Doses used in recent studies range from 40 to 45 Gy for microscopic disease control, and 50 to 55 Gy in the case of macroscopic residue[10].

In terms of prognosis, we classify initial sites into two categories, Favorable (involvement of the orbit, non-paramedic head and neck, genitourinary region and bladder-prostate, with overall survival rates of around 80% respectively). And unfavorable (involvement of the para-meningeal region, bladder and prostate, limbs and other sites, with an overall survival of 60%)[18].

Conclusion

A definitive diagnosis relies on pathological examination plus immunohistochemical analysis. Despite improvements in therapeutic management, the prognosis for children head and neck rhabdomyosarcomas remains very poor, given the often initially advanced stage of the disease, frequent inoperability in the case of basicranial extensions, and high metastatic potential. The management of rhabdomyosarcomas is multidisciplinary, involving multi-drug therapy, surgery and external radiotherapy.

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