

Child Presenting with a Huge Cervicofacial Mass : About a Case Report

Case Report

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ABSTRACT

Introduction: Rhabdomyosarcoma (RMS) is a malignant mesenchymal neoplasm. It is one of the most common malignant head and neck tumors in children and the most frequent soft-tissue sarcoma. The main locations are the base of the skull and the nasopharynx. We present a rare case of a child presenting with parotid RMS.

Presentation of Case: A 4-year-old male was admitted to our hospital with the chief complaints of a progressively enlarging, cervicofacial swelling. There was no history of fever or any other constitutional symptoms. Physical examination revealed a 60 mm left cervicofacial swelling, trismus and grade IV left peripheral facial paralysis. There were no fever or palpable nodes. Biological findings showed an increased LDH. Computed tomography scan of the neck revealed a bulky tumor in the left cervicofacial region, involving both superficial and deep lobes of parotid gland, causing compression of oropharynx, extending to the left cervical lymph nodes measuring 77*55 mm with difficulties determining the starting point. Two days after his admission, he presented dyspnea and fever. A tracheotomy was performed associated to cervical biopsy under general anesthesia. The immunohistochemical analysis revealed intense positivity for desmin and myogenin favoring the diagnosis of embryonal rhabdomyosarcoma. The patient received chemotherapy and radiotherapy. A residual mass remained in the parotid region, in contact with the carotid artery. Tumor resection was discussed but ruled out due to high surgical risk.

Conclusion: Parotid RMS is a rare entity. Treatment follows a rigorous international protocol associating surgery, chemotherapy and radiation therapy.

Key Words: Children, embryonal rhabdomyosarcoma, malignant tumor, parotid gland.

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INTRODUCTION

Rhabdomyosarcoma (RMS) is one of the most frequent malignant head and neck tumors and the most common variety of soft tissue sarcoma in childhood. It is a rare but aggressive disease^[1,2].

RMS is a malignant mesenchymal neoplasm divided into four main groups: embryonal, alveolar, pleomorphic and botryoid^[3,4]. Embryonal and alveolar types occur mostly in children and adolescents. In contrast, botryoid RMS occurs over a broad age range affecting children and older adults^[5].

In pediatric populations, 30% to 40% of RMSs develop in the head and neck region. The predilection sites are the base of the skull, nasopharynx, nasal cavity and orbit^[6,7,8].

Parotid rhabdomyosarcomas are more frequent in children with a median age of 7 years^[5,9].

Treatment options include ablative surgery, chemotherapy and radiation therapy^[3].

PRESENTATION OF CASE:

A 4-year-old male presented to the Otolaryngology Department for evaluation of a huge cervicofacial mass, increasing in size progressively over 20 days.

Physical examination found a voluminous left cervicofacial mass extending to the parotid region, measuring about 70 mm, fixed in relation to the deep plane, painless with trismus and grade IV left peripheral facial paralysis (House Brackmann score). There were no fever or cervical lymphadenopathy.

Computed tomography (CT) scan demonstrated the presence of a voluminous and necrotic tissue density process on the left parotid region bordering against the lateral lymph node sector (Levels : IB - II - III - VA - VB - IV), enhanced heterogeneously, measuring 74 x 44 x 77 mm. It exerts a mass effect on the trachea and esophagus (Figure 1: A-B-C).

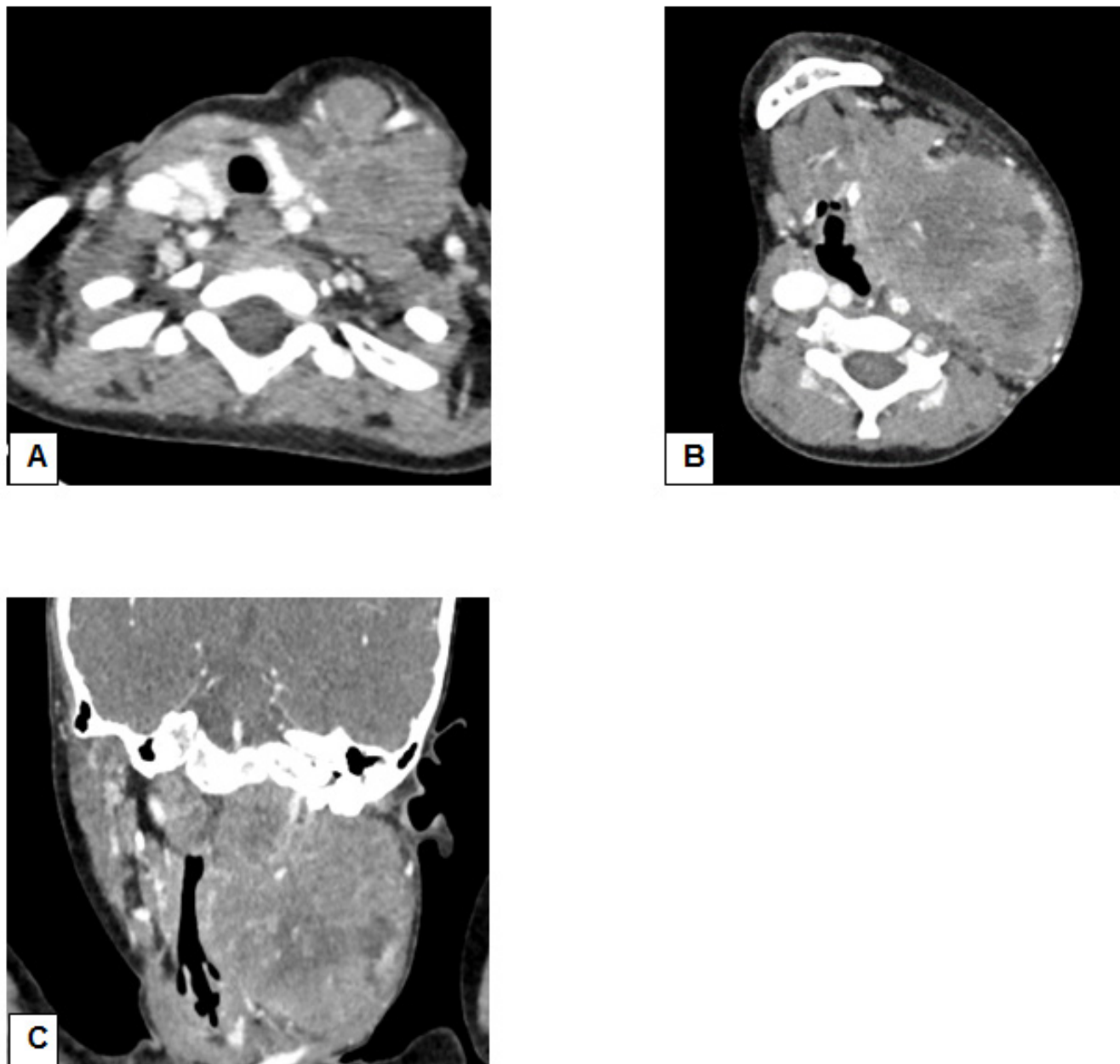


Fig. 1: Axial (A -B) and coronal (C) views of CT scan with contrast of the head and neck. The CT demonstrates an indistinctly bordered (arrows) mixed attenuation lesion with hypervascularity in the left face at approximately the parotid gland

He presented dyspnea and fever two days after his admission. A tracheotomy was performed to secure the airway associated to cervical biopsy under general anesthesia.

Histological examination revealed lymph node metastasis of an undifferentiated tumor.

Immunohistochemical studies were conducted to further classify this lesion.

Neoplastic cells were positive for myogenin and desmin.

Based on the immunohistochemical results, a diagnosis of embryonal RMS was made.

The patient received IVA-chemotherapy (ifosfamide, vincristine, and dactinomycin) and radiotherapy.

The evolution was good during the first-year follow-up without recurrences or distant metastases. The tumor showed considerable reduction in size but a residual mass remained in the parotid region, in contact with the carotid artery. Tumor resection was discussed but ruled out due to high surgical risk.

The patient presented no symptoms and his condition progressed well. The control CT scan showed no signs of recurrence.

DISCUSSION

RMS represents the most frequent soft-tissue sarcoma in children and accounts for 5% of all pediatric cancers^[10]. In the head and neck, it commonly affects orbit, paranasal sinuses, soft tissues of the cheek and the neck. The parotid location is rare^[11].

RMS is a highly aggressive mesenchymal neoplasm that exhibits skeletal muscle cells with varying differentiation degrees^[12]. It has been classified into four histological subtypes: embryonal, alveolar, botryoid, and pleomorphic.

The embryonal variant represents more than 80% of all cases and is considered at low or standard risk^[13]. This variant arises in children in the orbit, middle ear, nasal cavity, paranasal sinuses, or nasopharynx^[14]. Embryonal RMS of the parotid gland has never been described to our knowledge.

Symptoms of RMS are variable, depending on the site of initial presentation, the extent of the tumor, and the presence or absence of distant metastases and lymph node involvement^[3,15].

RMS can present as generalized painless swelling, oculobulbar cranial nerve deficits discharge, dysphagia, and significant ear and nose pain due to mass effect on the nasopharynx and paranasal sinuses^[14,16,17].

Common sites of metastasis are the lungs, skeletal system, lymph nodes, and brain, with hematogenesis being the common route of metastasis^[3,18].

The radiological characteristics of the tumor are not specific. Computed tomography (CT) of the head and neck is useful to descriptively identify the location and to assess bone erosion. Magnetic resonance imaging (MRI) gives a better definition of the mass and its involvement in adjacent sites^[3,14].

On CT-scan, RMS is a bulky tumor with multiple degrees of heterogeneous attenuation. On MRI, RMS is seen like a nonspecific low or isointense signal on T1-weighted sequences and high signal on T2-weighted images^[10].

Considering that RMS is a congenital tumor prenatal diagnosis can be suspected by MRI or ultrasound^[9]. A rare case of neonate with intra oral embryonal RMS; diagnosed on antenatal ultrasound scan has been reported by Skelton and Goodwin^[10]. Prenatal MRI can be used to perform, accurate diagnosis of fetal masse^[19].

The diagnosis can be suspected with CT-scan or MRI and confirmed by biopsy. Only histopathological examination permits definitive diagnosis.

Histologically, Embryonal RMS is characterized by small round blue cells, like neuroblastoma or Ewing's sarcoma, with primitive spindle cells and a myxoid background^[20].

Immunocytochemistry, including staining for desmin, smooth muscle actin and myogenin confirmed the diagnosis^[10,21].

The standard treatment in children includes a combination of chemotherapy, radiotherapy, and surgery^[14,22,23].

The aim of the treatment is to achieve loco-regional control and to prevent distant metastases^[13].

Prognosis factors of RMS are: age (less or greater than 10 years), tumor size (less or greater than 5 cm), histology subtype (alveolar or not), surgery status (complete, macro- or microscopically incomplete resection), primary site, lymph node invasion. They define risk groups and determinate the intensity and duration of treatment^[6].

The favorable prognosis group comprises favorable anatomopathology (non-alveolar type), complete primary resection, absence of lymph node invasion, favorable age (< 10 years) and tumor < 5 cm^[6].

Multidisciplinary approach in the management of RMS has improved the prognosis with a five-year survival rate of 74–77%^[19].

CONCLUSION

Parotid RMS is rare but should be considered as a differential diagnosis of any adenoiditis resistant to treatment in a child especially with facial palsy. Imaging has a key role in the initial staging and during the follow up. Current treatment includes a combination of ablative surgery, chemotherapy, and radiation therapy. Prognosis is good, as the site is accessible to a complete resection. All patients require long term follow-up to exclude recurrence.

CONFLICT OF INTEREST

There are no conflicts of interest.

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