MERIT RESEARCH JOURNALS
www.meritresearchjournals.org

Merit Research Journal of Medicine and Medical Sciences (ISSN: 2354-323X) Vol. 7(1) pp. 031-034, January, 2019
Available online http://www.meritresearchjournals.org/mms/index.htm

DOI: 10.5281/zenodo.2550814

Case Report

Lingual alveolar sarcoma in children: a rare soft tissue sarcoma

Adil Fouad[1*], Meriem El Baz[1], Nadia Mansouri[2], Abderrachid Hamdaoui[3], Dounia Basraoui[4], Hicham Jalal[4] and Jamila El Houdzy[1]

[1]Pediatric Hematology and Oncology Department, Mohammed VI University Hospital of Marrakesh, Morocco.

[2]Maxillofacial Department, Mohammed VI University Hospital of Marrakesh, Morocco.

[3]Center of Anathomocytopathology Zohor, Marrakesh, Morocco.

[4]Pediatric Radiology Department, Mohammed VI University Hospital of Marrakesh, Morocco.

***Corresponding Author's E-mail: adilfouad@hotmail.fr**
Tel /Fax: +212524300700

Abstract

Alveolar soft part sarcoma is a rare soft tissue sarcoma with a predilection for adolescents and young adults that tends to occur in the extremities. It is a well-differentiated and chemo-resistant solid tumor. Lingual alveolar sarcoma can occur at an early age and appears, in this case, to be chemo-sensitive. We report the case of a child with an ASPS of the tongue and the mouth floor with lung metastasis, never yet published in Morocco, and a review of the literature. Alveolar soft part sarcoma of the tongue and the mouth floor was diagnosed in a 13-year-old boy. He had a painful, occasionally bleeding, medio-lingual swelling, spanning the mouth floor and the pharynx, in progression for a year, complicated with dysphagia and dyspnea requiring emergency tracheotomy. He underwent a surgical excision of the mass, and then was treated by chemotherapy. The child died from hemorrhagic shock and worsening of the respiratory distress. ASPS should be included in the differential diagnosis of head and neck masses in children since early detection and treatment are essential. In particular, lingual alveolar sarcoma diagnosed at an early age seems to be chemo-sensitive.

Keywords: Alveolar soft part sarcoma, Chemotherapy, Child, Mouth floor, Tongue, Tracheotomy

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is a rare entity that most commonly affects adolescents and young adults in the extremities, but can occur in the orbit or on the tongue in younger patients (Katz AP et al., 2017). It is characterized by an almost specific translocation t(X,17) (p11;25) which creates a fusion protein, APSL-TFE3, acting as an aberrant transcription factor (Paillard C et al., 2015). ASPS is a classic chemo-resistant malignant solid tumor. The curative treatment is surgical. It associates the excision of the primary lesion with the resection, sometimes iterative, of possible metastases in the advanced stages. Alveolar sarcoma of the tongue can occur at an early age and appears to be chemo-sensitive (Marchac A et al., 2007). We report the case of a child with an ASPS of the tongue and the mouth floor with lung metastasis, never yet published in Morocco, and a review of the literature.

Case Report

A 13-year-old boy, one year before admission, presented a painless medio-lingual swelling, occasionally bleeding, with no inflammatory signs. Initially, the surgical excision, 7 months after the beginning of the symptoms, noted an epulis. The patient presented a recurrence of swelling 5

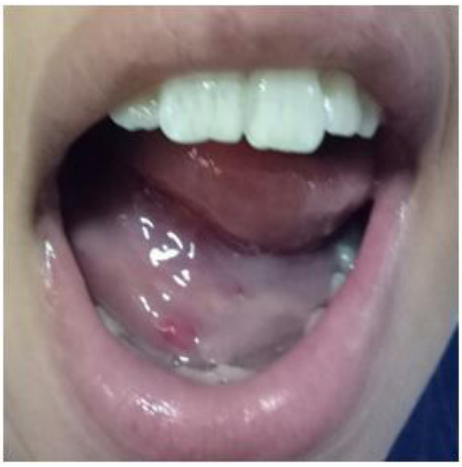

Figure 1. Swelling of the mouth floor with a small erythematous papule.

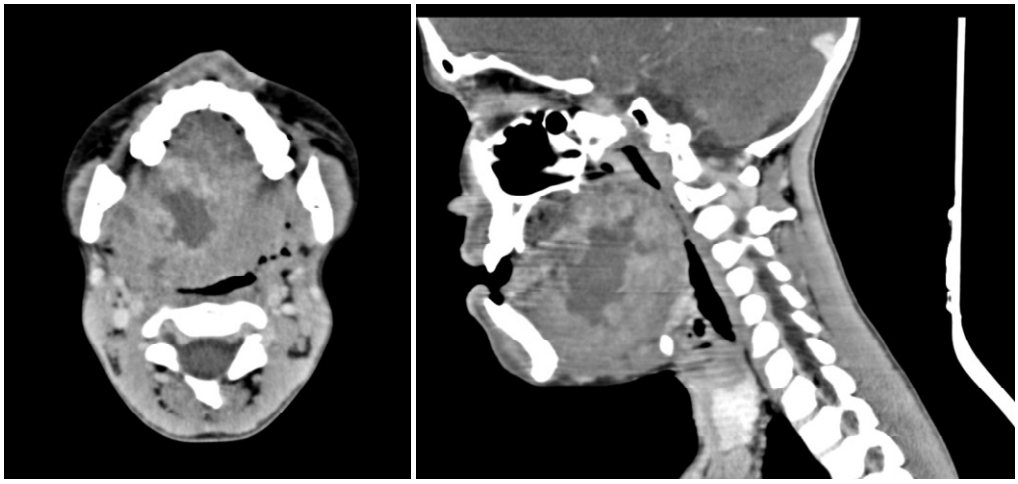

Figure 2. CT scan showing an endo-buccal tumor process locally infiltrating and completely obstructing the oropharyngeal lumen.

months after the surgical excision, stretching from the mouth floor to the pharynx, with a small erythematous papule on the mass, which bled upon contact (Figure 1); right jugulo-carotid lymphadenopathy measuring 1.5x1cm, with dysphagia, snoring, and stage 3 dyspnea on the NYHA scale, requiring emergency tracheotomy. Computed Tomography (CT) showed an endo-buccal tumor process measuring 7.4x7.3 cm, locally infiltrating and completely obstructing the oropharyngeal lumen (Figure 2), associated with thrombosis of the right external jugular vein, as well as pulmonary nodules and micronodules. The 2nd surgical excision of the mass confirmed lingual alveolar sarcoma of the tongue and the mouth floor without bone marrow infiltration. It was decided that the patient be put on the MMT 2005 protocol (malignant mesenchymal tumors of the child) IVADO cures (Vincristine, Actinomycin, Doxorubicin, and Ifosfamide). The child died on Day 7 of the protocol from hemorrhagic shock and aggravation of the respiratory distress.

DISCUSSION

Sarcomas are rare malignant tumors of mesenchymal origin, developed from connective tissues. Sarcomas exhibit a wide pathological diversity with more than 70 histological subtypes and a growing number of molecular subtypes. They develop at any age, children included. They can occur anatomically from head to toe and they are of variable aggressiveness even within the same histological subtype. Three categories of sarcomas

corresponding to different clinico-pathological entities of individually specific evolutions and whose treatments are distinct, are distinguished: bone sarcomas, visceral sarcomas and soft part sarcomas (SPS) (Honoré C et al., 2015).

Alveolar soft part sarcoma (ASPS) is a rare tumor that accounts for 0.5 to 1% of soft tissue sarcomas and 5% of non-rhabdomyosarcomatous sarcomas in children. The incidence is higher between ages 15 and 35. ASPS arises from skeletal muscles. The histopathogenesis of this disease is not fully known yet. Previous reports have described ASPS as a potentially fatal sarcoma because distant metastasis often develop either synchronously or metachronously (Marchac A et al., 2007; Ryu J et al., 2006). It occurs preferentially on the limbs and abdomen, and less frequently at the cephalic extremity where it represents approximately 25% of cases and is localized preferentially in the orbit and on the tongue (Marchac A et al., 2007).

The mouth floor is a particular anatomic zone of the oral cavity where specific stomatologic diseases can be located, together with some other lesions of the oral cavity (Bouletreau P et al., 2005). Originally, the occurrence of ASPS within the oral cavity (including the tongue) has been the major pattern of manifestation of this disease within the head and neck regions, especially in infants and children (Ryu J et al., 2006).

The lesion is typically slow growing, and when present in the tongue, patients may have dysphagia, dysarthria, and discomfort. ASPS is often initially misdiagnosed and the proper diagnosis is not made until final surgical pathology. Delay in diagnosis can be devastating as the disease becomes challenging to treat once it metastasizes, lung metastases being present in 65% of cases at diagnosis (Katz AP et al., 2017; Marchac A et al., 2007). For our patient, the diagnosis was done lately, one year after the symptomatology, and had a swelling spanning the mouth floor and the pharynx, a small erythematous papule on the mass which bled upon contact, right jugulo-carotid lymphadenopathy, lung metastasis, with dysphagia, snoring, and stage 3 dyspnea on the NYHA scale requiring emergency tracheotomy.

Developments in molecular biology have made possible the identification of recurring molecular anomalies in certain subgroups of sarcomas, being able to represent diagnostic, prognostic and therapeutic tools. The molecular classification of SPS includes until today 5 main groups of abnormalities: sarcomas with "simple genomic profile" showing reciprocal chromosomal translocations, activating mutation, inhibitive mutation or simple amplification; sarcomas with "complex genomic profile" can include several tens of molecular abnormalities. The development of new targeted therapies is based on the identification of a target specific of a tumor subgroup and involved in carcinogenesis mechanisms and/or tumor growth (Dufresne A et al., 2015).

ASPS is considered resistant to chemotherapy. The main treatment is surgical. It associates the excision of the primary lesion with the resection, sometimes iterative, of possible metastases in the advanced stages. Adjuvant treatment modalities such as radiotherapy and chemotherapy are used very rarely. The choice of adjuvant therapy would appear to depend on the empirical setting: clinical findings such as the ability to use complete surgical excision can be a crucial determinant of whether adjuvant treatment be used. It is accepted generally that adjuvant therapy is unnecessary if a small primary lingual ASPS can be resected completely and the patient does not experience clinical recurrence or distant metastasis. The use of novel targeted therapies as systemic treatment for ASPS has rapidly expanded in recent years following identification of the ASPL TFE3 translocation and overexpression of the MET tyrosine kinase receptor. Lingual alveolar sarcoma can occur at an early age and appears to be chemo-sensitive. Follow-up should be prolonged because metastases can occur more than 10 years after initial treatment (Katz AP et al., 2017; Marchac A et al., 2007; Ryu J et al., 2006). For our case, it was decided to perform the surgical excision and secondly chemotherapy because of relapse the MMT 2005 protocol IVADO treatment course (Vincristine, Actinomycin, Doxorubicin, and Ifosfamide). The child died on Day 7 of the protocol from hemorrhagic shock and aggravation of the respiratory distress. AP. Katz and al. have described a similar 12-month-old case, diagnosed at 2 years of age, treated by glossectomy with favorable outcomes after a 5-year follow-up (Katz AP et al., 2017). A. Marchac and al. have published a case of ASPS of the tongue and the mouth floor diagnosed in a two-year-old girl. She had lung metastasis. She was successfully treated with chemotherapy and surgery (Marchac A et al., 2007).

The 5-year survival rate in children, adolescents and young adults is close to 80% in case of localized disease but poorer in the presence of metastases (10%). Prognostic risk factors in the literature are age (> 10 Years), tumor size (> 5 cm) and presence of metastases (Paillard C et al., 2015; Marchac A et al., 2007). Our patient's prognosis was poor: he was 13 years of age, the tumor size was > 7cm, and he had lung metastasis.

CONCLUSION

Although quite rare, ASPS should be included in the differential diagnosis of head and neck masses in children since early detection and treatment are essential. In particular, lingual alveolar sarcoma diagnosed at an early age seems to be chemo-sensitive.

At this time, surgical resection remains the most effective treatment, but the development of an individualized treatment plan is essential.

Conflict of Interest: None

Consent for publication

Consent was obtained from the patient's parents.

ACKNOWLEDGMENTS

The authors thank the patient's parents, Dr. Klevor Raymond, and the staff of the pediatric hematology and oncology department for their help and support.

REFERENCES

Bouletreau P, Froget N, Gleizal A, Breton P (2005). Affections du plancher de la bouche. EMC-Stomatologie 1:126–40.

Dufresne A, Cassier P, Heudel P, Pissaloux D, Wang Q, and al. (2015). Biologie moléculaire des sarcomes et choix thérapeutiques. Bull Cancer 102:6–16.

Honoré C, Méeus P, Stoeckle E, Bonvalot S (2015). Le sarcome des tissus mous en France en 2014: épidémiologie, classification et structuration de la prise en charge. JCHIRV. 152:224-32.

Katz AP, Chen S, DeNardo BD, Mangray S, Groblewski JC (2017). Lingual alveolar soft part sarcoma in a pediatric patient: Case report and literature review. International Journal of Pediatric Otorhinolaryngology 17:36-9.

Marchac A, Picard A, Landman-Parker J, Larroquet M, Vazquez MP, Franchi G (2007). Sarcome alvéolaire de la langue: un cas pédiatrique. Rev Stomatol Chir Maxillofac. 108:547-50.

Paillard C, Coulomb A, Helfre S, Orbach D (2015). Le sarcome alvéolaire des parties molles de l'enfant et l'adolescent. Bull Cancer 102:792-8.

Ryu J, Kwon Y, Park BK, Jung YS (2006). Lingual alveolar soft part sarcoma treated only by conservative resection. International Journal of Pediatric Otorhinolaryngology 1:243-8.