

## Synovial Sarcoma in the Right Anterior Cervical Region: Case Report

Leniza C Lichtvan<sup>1</sup>, Cassio Henrique Taques<sup>2\*</sup>, Camilla H Tabushi<sup>2</sup>, Carolina Oldoni<sup>2</sup>, Caroline Gevaerd<sup>2</sup> and Cláudia MP Biondo<sup>2</sup>

<sup>1</sup>Professor of Pediatrics, Department of Pediatrics, Universidade Federal do Parana, Universidade Federal do Parana, Setor de Ciencias da Saude, Curitiba, Brazil

<sup>2</sup>Medical Student, Department of Pediatrics, Universidade Federal do Parana, Setor de Ciencias da Saude, Curitiba, Brazil

\*Corresponding Author: Cassio Henrique Taques, Medical Student, Department of Pediatrics, Universidade Federal do Parana, Setor de Ciencias da Saude, Curitiba, Brazil.

Received: July 06, 2018; Published: October 30, 2018

### Abstract

**Objective:** To report a case of a cervical synovial sarcoma in a three-year old child, given the significantly low incidence of this neoplasm in the head and neck region.

**Description:** A three years and three months old female was admitted to the hematology and pediatric oncology department of CHC-UFPR, due to a nodule in the right anterior cervical region, at thyroid gland level. Absence of weight loss, anorexia or night sweats. Bilaterally palpable cervical lymph nodes, in increased number without malignant characteristics. Ultrasound revealed an oval, solid, well delimited, hypoechoic, slightly heterogeneous and vascularized lesion, measuring 48 mm x 40 mm x 15 mm. Histological and immunohistochemical profiles were compatible with biphasic synovial sarcoma after nodule excision. In the impossibility to exclude persistence of residual tumor after the operation, four cycles of chemotherapy were performed. Throughout the treatment the patient presented febrile neutropenia, anemia and thrombocytopenia, with subsequent blood count normalization. Three months after the end of the chemotherapy, magnetic resonance imaging showed a stable condition and the patient maintained good remission of symptoms.

**Comments:** Synovial sarcoma (SS) is a malignant neoplasm of soft tissues, with a more frequent presentation in periarticular regions of the lower limbs in young adult males. The importance of their knowledge by pediatricians is due to the involvement of unusual regions in the child population. Although rare in the cervical region, SS should be considered as a differential diagnosis for cervical masses, and the diagnosis is confirmed through immunohistochemistry and anatomopathological study.

**Keywords:** Synovial Sarcoma; Head and Neck Neoplasms; Pediatrics

### Introduction

Synovial sarcoma is a malignant neoplasm of soft tissues, so named for its histological similarity to synovial cells. Despite this, its cellular origin is unknown, being attributed to pluripotent mesenchymal cells, having no relation with synovial structures. Its most frequent presentation is in periarticular regions of the lower extremities, in young adult male patients [1]. Only 5 to 10% present in the head and neck region [2]. Needless to say finding a cervical synovial sarcoma is not only rare, but even rarer when found in females.

The origin of 90% of these tumors is in the chromosomal translocation t (X; 18) (p11; q11), causing the breaking points to merge different genes, which determines the most frequent histological presentations: monophasic and biphasic [1]. The disease presents in most cases as a painless mass with a progressive increase in volume. Symptoms such as paresthesia and edema may occur, although fever and weight loss are rarely present [1].

The differential diagnosis includes the other causes for cervical masses, with confirmation coming through anatomopathological and immunohistochemistry studies. Epithelial and mesenchymal cell markers characterize an immunochemical study [3]. Cytogenetic analysis, FISH or RT-PCR may aid in the diagnosis and identification of the tumor type, especially in poorly differentiated cases, which may show a better prognosis in the case of monophasic tumors [1].

The treatment indicated is multimodal, initially involving radical surgical excision of the mass, with adjuvant or neoadjuvant radiotherapy. Chemotherapy regimens with Doxorubicin and Isophosphamide may also be used [3].

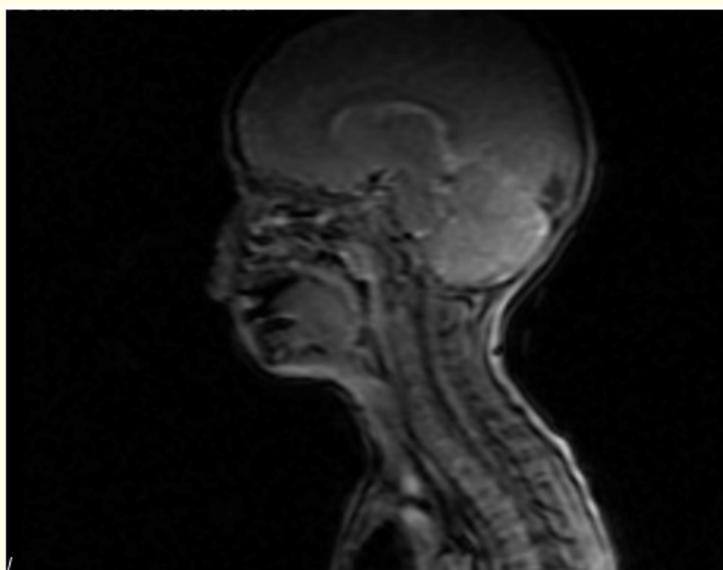
The prognosis involves 45% recurrence in synovial sarcomas of the head and neck, and 33% of distant metastasis [3] for lungs, lymph nodes, and bone marrow [2].

Apart from the already mentioned rarity among females and cervical location, Synovial sarcomas of the head and neck are often confused with other mesenchymal and non-mesenchymal tumors, due to a variety of other findings. Some tumors may show the characteristic monophasic pattern, as well as myxoid, calcifying, and bone forming forms [4]. Thus, it is of the utmost importance for physicians to be able to recognize the importance of properly diagnosing a cervical synovial sarcoma.

### Case Report

A 3-year-old female patient was admitted to the pediatric hematology and oncology department, with a history of nodule appearance in the anterior cervical region a week ago. Parents denied having weight loss, anorexia or night sweats. At the examination, a nodule was observed in the right cervical region, at the level of the thyroid gland, with dimensions of approximately 5 cm x 4 cm, fibroelastic consistency adhered to adjacent structures and painless. Lymph nodes of the cervical region were bilaterally palpable, with no malignancy characteristics. Cervical ultrasonography demonstrated an oval lesion in the anterior cervical and right paramedian areas, cranially located to the thyroid lobe, a solid, well delimited, hyposonic, slightly heterogeneous and vascularized lobe, measuring 48 mm x 40 mm x 15 mm, as well as an increase in the number of cervical lymph nodes, all within typical dimensions. The patient had the nodule excised, with a histological and immunohistochemical profile compatible with biphasic synovial sarcoma.

One month after the nodule excision, a control MRI (Figure 1) demonstrated localized lesion anterolateral to the laryngeal cartilage, without compromising it, with dimensions of 33 x 8 x 21 mm, suggestive of postoperative changes. However, since it was not possible to exclude the presence of a residual tumor, the patient underwent four cycles of chemotherapy with phosphamide 3 g/m<sup>2</sup>/day for three days and doxorubicin 37.5 mg/m<sup>2</sup>/day for two days, associated with mesna drug, to reduce the urinary toxicity of the therapy.

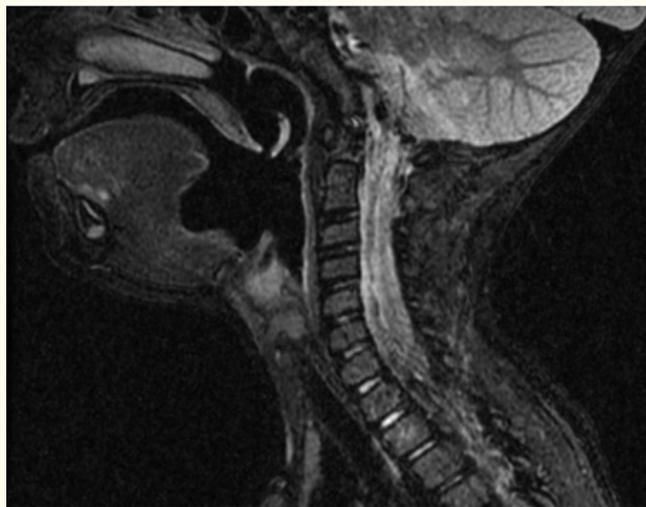


**Figure 1:** Postoperative changes in the subcutaneous region and adjacent molar parts characterized by hypersignals in T1 and T2 in the right lateral cervical, at the level of C3-C5. A search sign in deep planes of the right cervical region, without significant expansive effect, characterized by high signal instead of enhancing after the paramagnetic contrast medium, measuring about 33 x 8 x 21 mm, anterolaterally laryngeal cartilage, with no signs of laryngeal involvement.

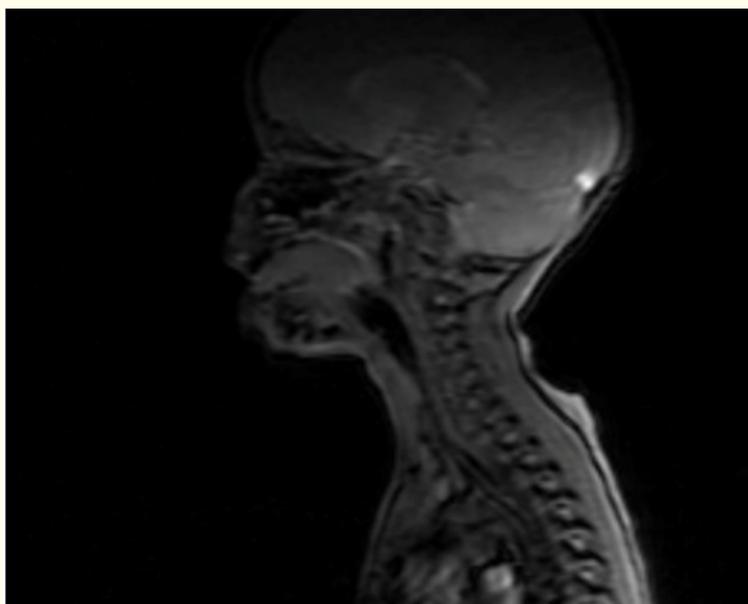
## Synovial Sarcoma in the Right Anterior Cervical Region: Case Report

1120

Throughout the treatment, she presented with two episodes of febrile neutropenia (neutrophils of 26/uL and 0/uL), satisfactorily treated with intravenous Cefepime, with improved immune response and infection control. In addition, she had anemia (hemoglobin levels reached 8.2 g/dL) and thrombocytopenia (96,000/uL). Side effects of chemotherapy were adequately controlled with dexamethasone and diphenhydramine. Red blood cell concentrate was administered to the patient for the correction of anemia. Granulokine was also administered for the prevention and treatment of new episodes of neutropenia. Subsequently, the patient had still three other episodes of neutropenia without infection. After completion of the treatment, serum markers were back within normal limits (hemoglobin 12.2 g/dL, leucocytes 9,590/uL, neutrophils 6,617/uL and platelets 214,000/uL). After chemotherapy termination, MRI (Figure 2) showed a reduction in lesion size compared to the initial examination (19 x 6 x 13 mm). Three months later at the last visit, the MRI (Figure 3) was stable, and the patient maintained a good recovery.



**Figure 2:** Reduction in the lesion dimensions in deep planes of the right cervical region, without significant expansive effect, characterized by high STIR signal, measuring about 19 x 6 x 13 mm, located anterolaterally to the laryngeal cartilage, with no signs of impairment.



**Figure 3:** Reduction of the lesion dimensions, with poor contrast enhancement in the soft parts of the surgical site (right region of the neck) anterolateral to the larynx, measuring about 15 x 6 mm in the non-specific aspect axial plane.

## Discussion

The site of presentation of synovial sarcoma in the described case comprises of a rare finding among the sites of presentation of this tumor [2]. The clinical presentation of the patient, on the other hand, was in accordance to the majority of cases of synovial sarcoma - a painless mass [1]. As shown by Ryan, *et al.* [1], some manifestations that could be found would still be those related to tumor compression, ranging from paresthesias to extremity edema. Consistent with this patient's case, constitutional symptoms, such as unintentional weight loss and fever, are rarely encountered [1]. For the diagnostic confirmation, the importance of cytogenetic analysis [1] is well established - and thus, as it was performed for this patient, it directly lead to the diagnosis of synovial sarcoma.

Surgical excision is, in fact, the initial therapeutic method for synovial sarcomas. After or before this, some studies show the use of radiotherapy or chemotherapy [3]. For this patient, the adjunctive use of chemotherapy cycles with phosphamide was chosen, primarily due to the suspicion of a residual lesion.

The incidence of tumor recurrence in synovial sarcomas, or the residual lesion itself, as in this case, may reach about 45% when in the head or neck region [3], which reiterates the importance of the chemotherapeutic administered to the patient after the excision, given the most likely diagnostic assumption.

Despite the rarity of synovial sarcoma - incidence rates are not well described in the literature - it is the most common malignant soft tissue non-rhabdomyosarcomatous sarcoma in the pediatric population [5]. The prognosis for patients with this type of sarcoma, when located, is usually good. However, for those with metastatic disease, the survival rate is minimal [5]. Thus, it is important to remember this differential diagnosis, especially in the pediatric population, so that the best therapeutic alternative (from excisional, chemotherapeutic or radiotherapeutic) is applied to diminish mortality and recurrence.

## Bibliography

1. Fletcher CDM., *et al.* "World Health Organization. Classification of tumours of soft tissue and bone, 4<sup>th</sup> edition". IARC Press, Lyon (2013).
2. Subtil Magalhães Freire G., *et al.* "Sarcoma sinovial cervical". *Revista Brasileira de Cirurgia de Cabeça e Pescoco* 43.3 (2014): 147-149.
3. Narayanan G., *et al.* "Synovial Sarcoma of the Larynx: Report of a Case and Review of Literature". *Case Reports in Otolaryngology* (2017): 6134845.
4. McCarville M., *et al.* "Synovial Sarcoma in Pediatric Patients". *American Journal of Roentgenology* 179.3 (2002): 797-801.
5. Jang J-W., *et al.* "Synovial Sarcoma of the Posterior Neck : A Case Report and Review of Literature". *Journal of Korean Neurosurgical Society* 47.4 (2010): 306-309.

**Volume 7 Issue 11 November 2018**

**©All rights reserved by Cassio Henrique Taques., *et al.***