



Synovial sarcoma of the hypopharynx in a pediatric patient: Case report

N.H. Alotaibi^{a,*}, Aurélie Bornand^b, Nicolas Dulguerov^a, M. Becker^c, Pavel Dulguerov^a^a Department of Otorhinolaryngology – Head & Neck Surgery, Geneva University Hospital, 4, rue Gabrielle Perret-Gentil, 1204 Geneva, Switzerland^b Department of Clinical Pathology, Geneva University Hospital, 4, rue Gabrielle Perret-Gentil, 1204 Geneva, Switzerland^c Department of Radiology, Geneva University Hospital, 4, rue Gabrielle Perret-Gentil, 1204 Geneva, Switzerland

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ABSTRACT

INTRODUCTION: Synovial sarcoma (SS) is uncommon high grade soft tissue sarcoma, accounting for less than 10% of all head and neck sarcomas. Also, about 10% of SS occur within the Head & Neck. In the pediatric population, SS is an extremely rare head & neck malignancy.

PRESENTATION OF CASE: We present a case of sixteen years old boy diagnosed with SS situated of the hypopharynx treated by surgical excision and post operative radio-chemotherapy.

DISCUSSION: This anatomical location brings additional functional challenges (swallowing, phonation, respiration), especially in the pediatric population. Pre-operative and even post-operative histopathological diagnosis of SS remains difficult. Optimal treatment of Head & Neck SS has to balance functional and oncologic aspects.

CONCLUSION: SS is an extremely rare head & neck malignancy in pediatric population. It has multifaceted challenges including pre and post-operative histopathological diagnosis and optimal modality of treatment. Clinical judgment, especially in the pediatric population, needs to balance tumor free margins and organ preservation in head and neck region.

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1. Introduction

Synovial sarcoma (SS) is uncommon high grade soft tissue sarcoma, accounting for less than 10% of all head and neck sarcomas. Also, about 10% of SS occur within the Head & Neck. In the pediatric population, SS is an extremely rare head & neck malignancy.

2. Presentation of case

A sixteen years old boy was referred for a left neck mass and the remaining clinical history was unremarkable. A firm 4 × 3 cm left neck mass, not mobile, medial to the sternocleidomastoid muscle was appreciated. Flexible endoscopy (Fig. 1) revealed a left sub-mucosal lateral pharyngeal wall mass impinging on the larynx but with normal vocal cords mobility.

The head and neck magnetic resonance imaging studies showed that the mass measured 4.8 × 3.3 × 7.0 cm displacing the great vessels laterally and pushing the left hypopharynx and larynx medially (Fig. 2).

The patient underwent an excision of the mass through a left cervicotomy. During surgery, the tumor was medial and easily dis-

sected from the sternocleidomastoid muscle and carotid sheath. It was covered laterally by a muscular layer and difficult to dissect from the posterior edge of the thyroid cartilage. Complete resection resulted in an elongated hypopharyngeal perforation which was repaired without flap usage. The post-operative course went uneventful, without cranial nerve deficit, and the patient resumed oral feeding on day 7. At 8 months follow up, no recurrence was detected (Fig. 3).

Histology revealed a biphasic Synovial sarcoma(SS), with malignant spindle-cell proliferation (Fig. 4) and focal epithelioid cells, showing vascular invasion. There were 35 mitoses per 10 high-power fields (HPFs) and necrotic foci. The tumor was well-defined but not encapsulated. Immunohistochemical staining was focally positive for EMA (Epithelial Membrane Antigen), pancytokeratins, CD56 and DOG1 (Discovered on GIST-1), and negative for S100, smooth muscle actin, desmin, c-Kit (CD117) and CD34. RT-PCR showed a t(X;18)(p11;q11) translocation, and fluorescence in situ hybridization (FISH) showed a rearrangement of the SS18 gene (Fig. 5).

3. Discussion – synovial sarcoma of the hypopharynx

Synovial sarcoma (SS) is uncommon high grade soft tissue sarcoma, supposedly accounting for less than 10% of all head and neck sarcomas [1]. About 10% of SS occur within the Head & Neck [1], with a median age of presentation in the third decade¹ making SS

Abbreviations: SS, synovial sarcoma.

* Corresponding author.

E-mail address: nlot@hcuge.ch (N.H. Alotaibi).



Fig. 1. Endoscopic view of the larynx and pharynx. A lateral normal mucosa covered mass displaces the larynx.

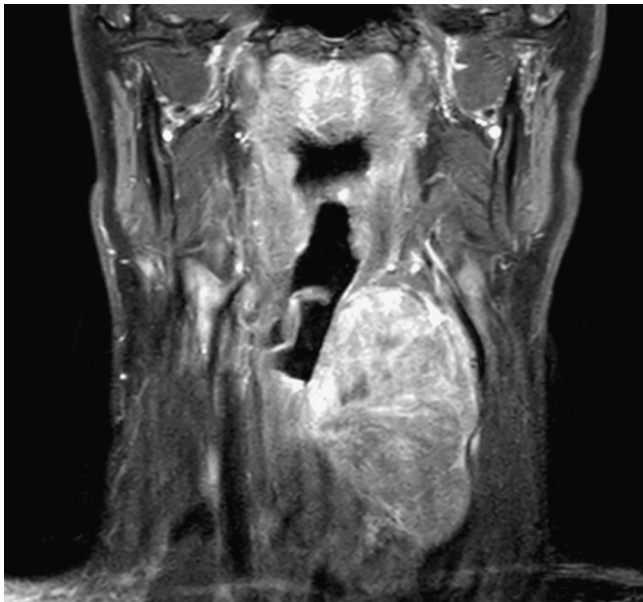


Fig. 2. The mass parenchyma showed T1 weighted signal isointense to skeletal muscle and hyperintense signal on T2 weighted imaging with remarkable enhancement. T1 SPAIR sequence after gadolinium shown.

an extremely rare head and neck malignancy in the pediatric population. About half of head and neck SS arise from the soft tissues of neck, while the remaining half are related to aero-digestive track structures [1]. Therefore, a painless neck mass is the most frequent presenting sign, the remaining symptoms depending on the exact location of the tumor.

Although SS is named because of the histological resemblance to the synovium, joints are rarely involved and the origin has been attributed to pluripotent mesenchymal cells [1]. Histologically two forms are described: monophasic (containing only spindle cells) and biphasic, made off both spindle and epithelioid cells. Spindle cells are arranged in sheets or fascicles with occasional herringbone pattern. Nuclear palisading is rare, unlike leiomyosarcoma and malignant peripheral nerve sheath tumors. Epithelial cells form glandular structure or are arranged in solid nests or cords. There can be some poorly differentiated areas showing an epithelioid pattern, a small-cell pattern and/or a high-grade spindle-cell pattern. Many SS focally show a hemangiopericytic (vascular) pattern [2–4].

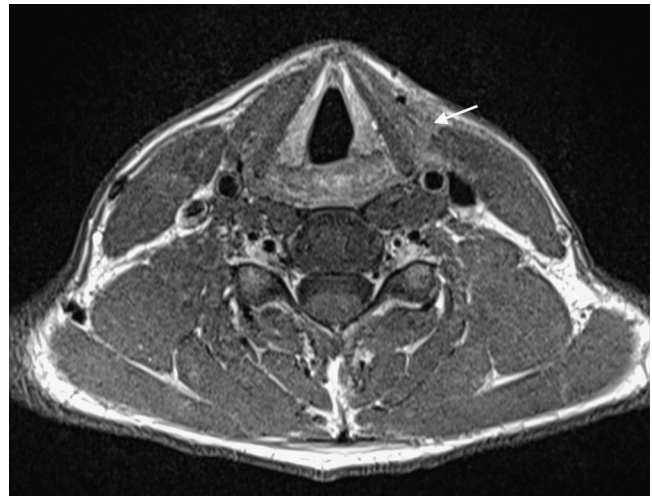


Fig. 3. Gadolinium enhanced T1 weighted image obtained after surgery shows contrast enhancement along the outer border of the thyroid cartilage in the left prelaryngeal strap muscles and along the left sternocleidomastoid muscle due to inflammatory postoperative changes. Follow-up MRI eight months later revealed gradual decrease of contrast enhancement.

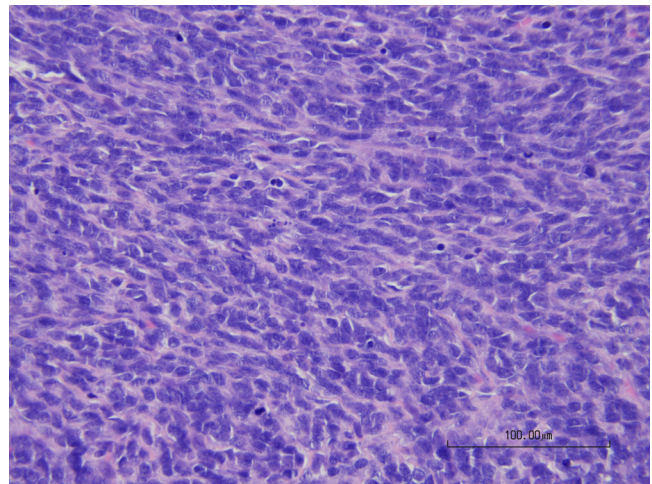


Fig. 4. Malignant spindle-cell proliferation. Hematoxylin-eosin, original magnification, 20×.

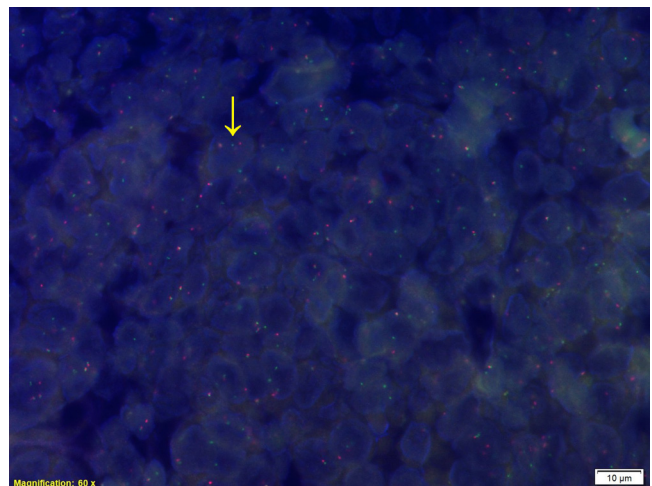


Fig. 5. Break-apart FISH. The red and green signals normally flank the SS18 gene; splitting of these signals indicates a rearrangement (arrow). Magnification 10×.

The differential diagnosis depends on the type of SS. A correct initial histopathologic diagnosis of monophasic SS is often difficult: the differential diagnosis including other spindle-cell tumors such as malignant peripheral nerve sheath tumors, leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma, solitary fibrous tumor, and spindle cell carcinoma. Biphasic SS is easier to identify, with malignant peripheral nerve sheath tumors, carcinosarcoma, and malignant mesothelioma being the differential diagnosis [3,4].

Immunohistochemistry is helpful, with most SS showing focal immunoreactivity for EMA and cytokeratins, contrary to malignant peripheral nerve sheath tumors, leiomyosarcoma and rhabdomyosarcoma [2–4]. SS may be focally positive for CD56 (up to 80%) [4], S100 (up to 40%) [2,4], and DOG1 (15%) [5]. CD34 is generally negative (<5%), in contrast to solitary fibrous tumor [2,4]. Leiomyosarcoma is positive for desmin and smooth muscle actin, while rhabdomyosarcoma is positive for desmin and myogenin [3].

Most (90%) SS exhibit a translocation t(X;18)(p11;q11) between chromosomes 18 (SYT gene) and X (SSX gene), resulting in a SYT-SSX-1 (biphasic SS) or SYT-SSX-2 (monophasic SS) fusion [6]. Generally we use break-apart FISH and RT-PCR to search the translocation. This translocation is specific and diagnostic for SS [2–4].

Preoperative diagnosis of SS is challenging because of its non-specific symptomatology, low clinical morbidity, lack of specific radiologic features, and physicians' unfamiliarity. The most frequent preoperative diagnosis is neurogenic tumors and a recent study of peripheral SS suggests that pain, short symptom duration, peritumoral edema without a definite target sign on MR images should evoke SS [7].

Prognosis of synovial sarcoma has been related to tumor size [1,8], biphasic histology, [1,6,8] SSY-SSX2 gene fusion, and negative resection margins [1]. Since our preoperative diagnosis was a schwannoma, the resection was less radical than possible and the margins were focally positive, a frequent finding with SS. Others have performed extensive resections with total laryngopharyngectomy for margin clearance in similar case [9]. Clinical judgment, especially in the pediatric population, needs to balance tumor free margins and organ preservation. Besides surgery, radiation seems to confer a local control and survival benefit, while the role of chemotherapy is controversial [1].

4. Conclusion

SS is an extremely rare head & neck malignancy in pediatric population. It has multifaceted challenges including pre and post-operative histopathological diagnosis and optimal modality of treatment. Clinical judgment, especially in the pediatric population, needs to balance tumor free margins and organ preservation in head and neck region.

Conflicts of interest

The authors have no conflict of interest to declare.

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Ethical approval

Approval to publish case report is waived by the institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Naif Alotaibi, H, MD: make substantial contributions to conception and design, acquisition of data, analysis and interpretation of data.

Aurélié Bornand, MD: participate in drafting the article and revising it critically for important intellectual content.

Dulguerov Nicolas, MD: participate in drafting the article and revising it critically for important intellectual content.

Becker M, MD: participate in drafting the article and revising it critically for important intellectual content.

Pavel Dulguerov MD: give final approval of the version to be submitted and any revised version.

Guarantor

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