

Synovial Sarcoma of the Left Palatine Tonsil Case Report and Review of Literature

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Abstract

Synovial sarcoma arises primarily in the lower extremities with a predilection for sites in proximity to large joints, such as the knee. It rarely occurs in the head and neck region, and the tonsil is an unusual site for the tumor, with only eight previously published cases in this anatomical site.

6–10 % of all soft tissue tumors, only 3–5 % arise in the head and neck region, and SS arising in the tonsil is a rare finding, with only eight well-documented cases reported in the English language literature. We describe the histopathological findings of a primary synovial sarcoma of the palatine tonsil arising in a 20-year-old male, with presentation of dysphagia and sleep disturbance.

Keywords: synovial sarcoma, palatine tonsil, tonsillectomy, radiotherapy, chemotherapy

Introduction

Synovial sarcomas are rare soft tissue sarcoma that primarily arise from the extremities, especially the lower thigh-knee region [1].

It is a highly malignant soft tissue tumor, accounts for less than 1% of all head and neck malignant neoplasms [6].

The literature has shown that this tumor can be found in unusual sites such as the abdominal wall and the head and neck region which are non-synovium-lined spaces [1].

Although the head and neck are rare primary site for synovial sarcoma, only 3-5% of these neoplasms appear in this region [2]. The parapharyngeal space is the most common location for synovial sarcoma [3, 4] Primary synovial sarcoma of the palatine tonsil is extremely rare [5].

A high clinical suspicion, careful examination, proper imaging and histopathological exam needed to diagnose those kind of condition.

This article is about a rare case of synovial sarcoma which originated in the left palatine tonsil.

Case report

A 20-year-old male patient presented to our otolaryngology clinic with history of difficulty in swallowing, disturbance of sleep and fullness in the throat since more than 2 months.

On and off difficulty in breathing specially during supine position. He did not have other symptoms relating to the ear, nose and throat. He had no history of a loss of weight or appetite. Patient not known to have any medical or surgical issue before, and he is not smoker.

Patient was fully conscious oriented and vitally stable. Oral examination showed left pedunculated purple mucosal arising from right tonsil mass projected just behind the anterior pillar crossing the midline and reaching up to the right tonsil No cervical lymphadenopathy was noted.

Findings on the remainder of the ENT examination were normal.

Revealed the presence of a pedunculated mucosal lesion projecting into oropharynx arising from just behind the posterior pillar of tonsil on left side. No cervical lymphadenopathy was noted. Findings on the remainder of the ENT examination were normal.

CT scan of the head and neck area done and showed large ill-defined

heterogenous enhancing noncalcified soft tissue lesion measured about 4.5* 41*40 mm obliterating the oropharyngeal space compressing the left side of nasopharynx, with sub centimetric cervical lymph node.

The patient underwent excisional biopsy of the mass and the left palatine tonsil and send it to the histopathology.

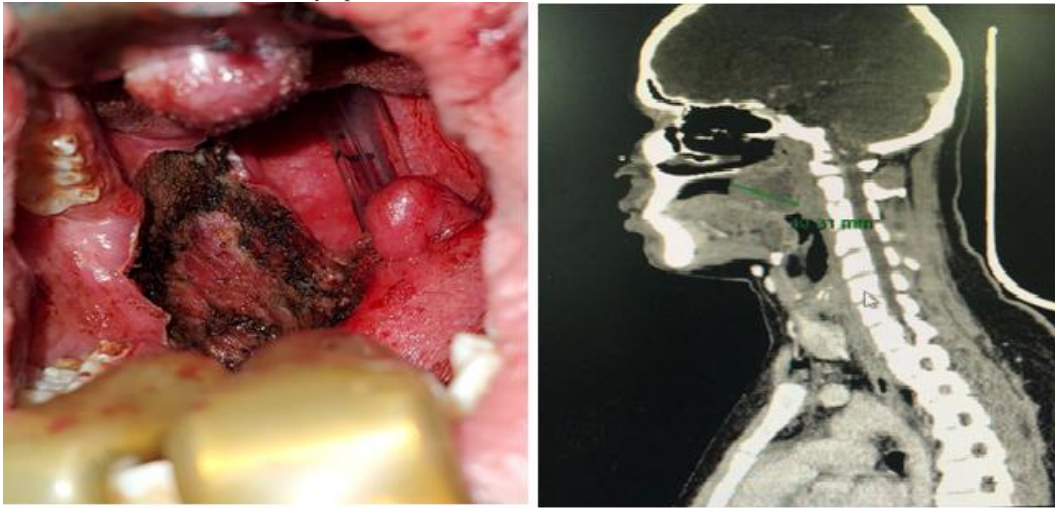


Figure 1





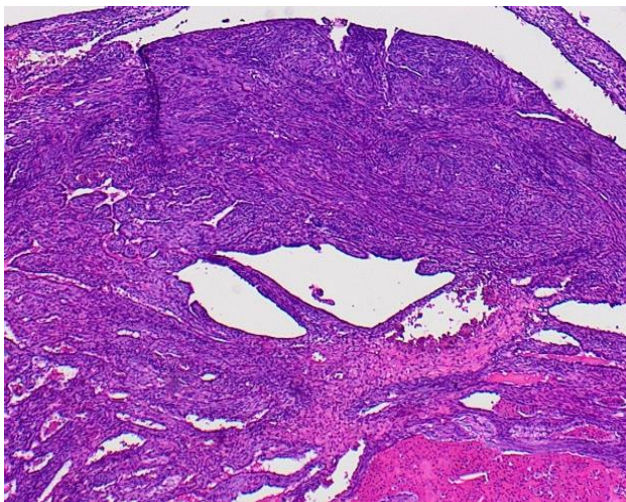
Histopathologic examination showed biphasic tumor with slit like spaces lined by epithelial cells and Spindle cells.

Immunohistochemistry highlighted spindle cells strongly expresses for vimentin, focal weak cytoplasmic positive for bcl-2, pan cytokeratin positive in epithelial cells and CD99 was negative consistent with Synovial sarcoma Carcino sarcoma.

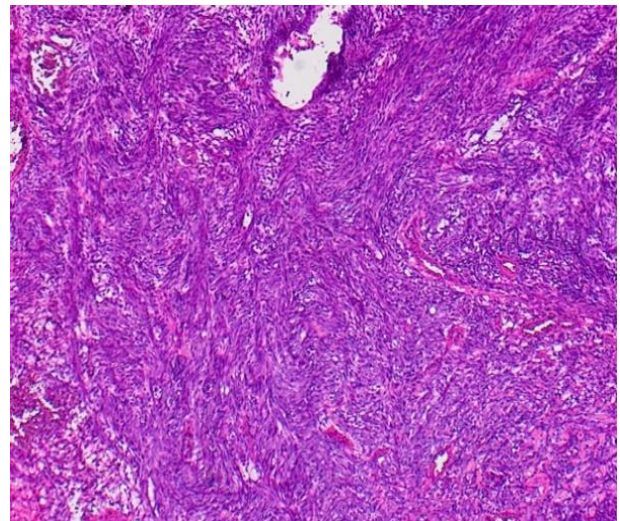
Patient then referred to higher center and received adjuvant radiotherapy.

Case was followed up for 1 years for both local and metastatic work up and is free of any symptoms.

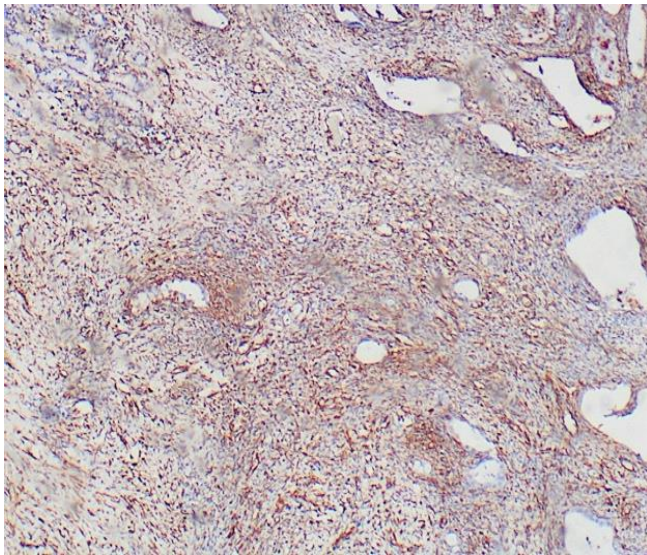
We lost the connection with the patient since then.



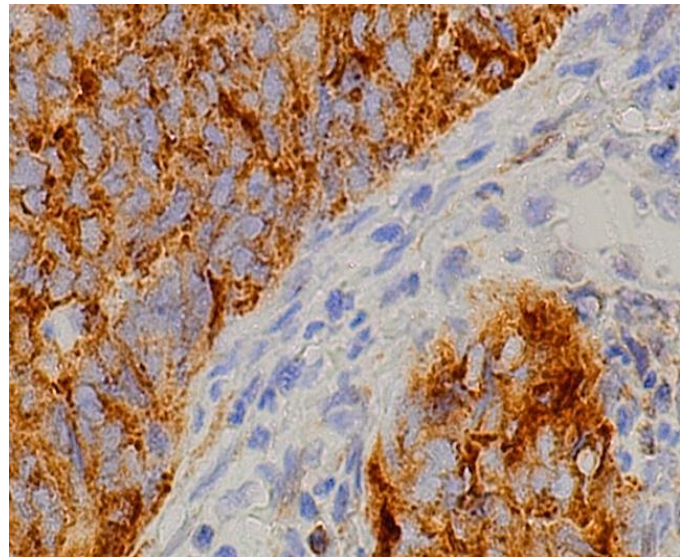
Biphasic tumor



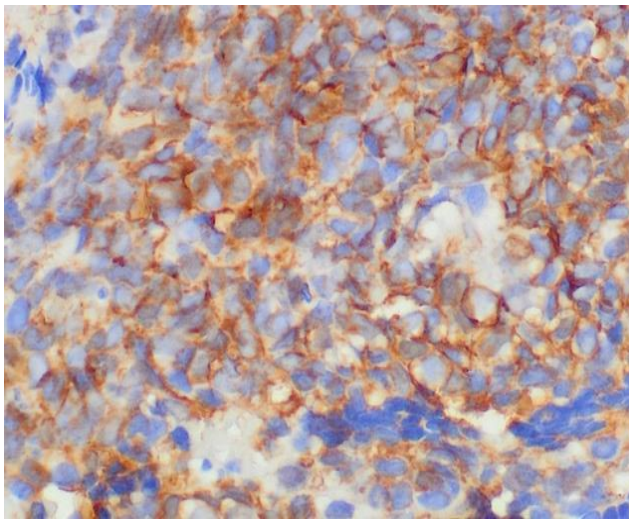
Spindle cell component x10



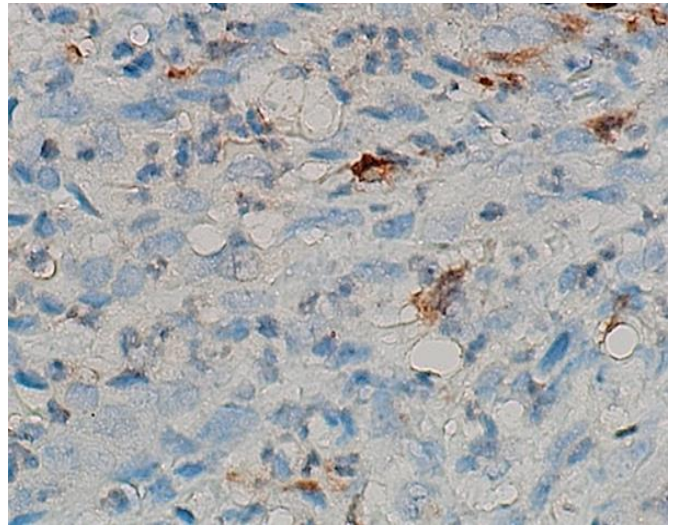
Vimentin positive in spindle cells



bcl-2: Focal positive



CD99: Focally positive



S100: focally positive

Discussion

Among the rare and aggressive malignant neoplasms that occur in the head and neck, the soft tissue sarcomas represent about 1% of all head and neck cancers, and about 4% to 10% of all soft tissue sarcomas in adults. Head and neck site sarcomas occur more frequently in the scalp, the face, and the neck area [6]. Sarcomas of the head and neck tend to recur locally more frequently and have a worse outcome than tumors elsewhere in the body [7].

Synovial sarcoma, a highly malignant soft tissue tumor, accounts for less than 1% of all head and neck malignant neoplasms, according to the site of involvement those kind of malignant neoplasm may present with diffident clinical presentation, in tonsillar area the presentation ranging from mild dysphagia to sleep disturbance, or even more severe airway compression [6].

In the head and neck region, synovial sarcoma most commonly affects the parapharyngeal space. To date, there have been approximately 100 cases of synovial sarcoma of the head and neck region reported in the literature [8].

Prior to treatment clinical and imaging examinations should be considered for all patients with head and neck synovial sarcoma.

The clinical findings of those cases advocate that the extent of the resection have to be decided through preoperative imaging studies, especially CT and MRI, with the cause to pick out top-quality surgical techniques and avoid suboptimal surgical resection. further, a preoperative incisional biopsy for destiny direct surgical planning is tremendously endorsed [9]. Histologically, synovial sarcoma presents as monophasic, biphasic, or uncommonly poorly differentiated subtypes.[9] Synovial sarcoma may cause differential diagnostic challenges with other

soft tissue sarcoma, such as fibrosarcoma, leiomyosarcoma and clear cell sarcoma.

Special immunohistochemical stains are helpful for differentiating synovial sarcoma from other soft tissue sarcomas. However, the precise diagnosis of synovial sarcoma can be difficult with histopathological and immunohistochemical studies alone.

Detection of this specific chromosomal translocation is helpful in confirming the diagnosis of synovial sarcoma, particularly when histological studies are uncertain [9].

Therefore, if the results of histopathological and immunohistochemical studies are inconclusive and there is a high clinical suspicion of synovial sarcoma, molecular genetic testing should be considered for diagnostic confirmation. [9].

According to the general principles of high-grade soft tissue sarcoma, standard local therapy for synovial sarcoma consists of wide surgical resection and adjuvant radiotherapy when appropriate [10].

Synovial sarcoma is one of the most chemo sensitive soft tissue sarcomas, with about 50% response rates to ifosfamide - and doxorubicin-based regimens. In an effort to improve systemic disease control and survival, adjuvant chemotherapy is widely used [10].

Currently, ifosfamide-based regimens with or without doxorubicin are considered the first choice of chemotherapy for patients with locally advanced and metastatic synovial sarcoma. [10-11].

Conclusion

The diagnosis of synovial sarcoma in head and neck is rare and should be kept in mind as one of differential diagnosis even in such unusual sites as the tonsils, especially when patient presented with rapid progression of unilateral tonsillar mass causing dysphagia and compromising the airway.

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Declarations of interest

The Authors declare that there is no conflict of interest.



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