Case Report

# Tonsillar Synovial Sarcoma with Review of Literature

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**Abstract**

Synovial sarcoma is a malignant tumor of mesenchymal origin which can develop at any location, with deep soft tissues of extremities being the commonest site involved, typically in young aged individuals, nevertheless it is uncommon in head and neck region and is enormously unusual in the tonsil. We represent a 30-year-old male with a right tonsillar mass who complained of difficulty in swallowing, odynophagia, right sided otalgia and sore throat for 6-months. The mass was excised and sent for histopathology and immunohistochemical studies that established the diagnosis of biphasic synovial sarcoma of the right tonsil.

***Keywords:*** Biphasic, synovial sarcoma, immunohistochemical studies

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## Introduction

Synovial sarcoma is a malignant tumor of mesenchymal origin that is considered high grade and is very uncommon representing about 5-10% of tumors of soft tissue.[1,2] Synovial sarcoma can arise as a primary tumor at any site, most commonly from the deep soft tissues, particularly of extremities of young aged individuals, however it is uncommon in the region of the head and neck accounting around 3-5% of cases,[3-5] most cases affecting men in their third decade[6], with the hypopharynx being the commonest site.[7-10] Synovial sarcoma of the tonsil is extremely scarce,[1,9,11]and few cases have been mentioned in literature. The diagnosis depends on histopathological examination that is assisted by immunohistochemical and cytogenetic analysis.[12,13] In this article, we represent a case of synovial sarcoma that was initiated in right tonsil of a 30-year-old male patient.

## Case report

A 30-year-old male patient complained of difficulty in swallowing, odynophagia, right sided otalgia and sore throat for 6 months duration during which the patient was diagnosed initially as a case of chronic tonsillitis and was kept on intermittent antibiotic therapy. The patient didn't respond to treatment and eventually consulted an otorhinolaryngologist who, on examination, discovered a right tonsillar mass with no lymph nodes involvement and a normal left tonsil. The patient's medical history was non beneficial such was his family history. Thus, the right tonsil was entirely excised through a transoral approach and sent for histopathology and immunohistochemistry to confirm the diagnosis. The specimen was received in the form

of multiple soft grey pieces of tissue, the largest measuring 1.5 cm length and 1.5 cm width. Microscopical examination of tissue sections stained by the conventional hematoxylin and eosin stains showed the presence of biphasic growth pattern of fascicular hypercellular spindle cells and oval-round cells with hemangiopericytoma-like vascular pattern, suggesting biphasic synovial sarcoma of the right tonsil (figures 1,2 and 3). Immunohistochemical examination revealed that the tumor cells were strongly positive for TLE1 (transducin-like enhancer of split 1), diffusely positive for CD99 (cluster of differentiation molecule 99 (MIC2)), positive for BCL2 (B cell lymphoma 2), CD56 and EMA (epithelial membrane antigen) (figures 4,5,6,7 and 8, respectively), all of which helped to confirm the diagnosis of biphasic synovial sarcoma. Meanwhile, tumor cells showed no expression of cytokeratin (AE1/AE3), CD57 and S100 (figures 9,10 and 11, respectively) that helped to differentiate it from other soft tissue sarcomas.

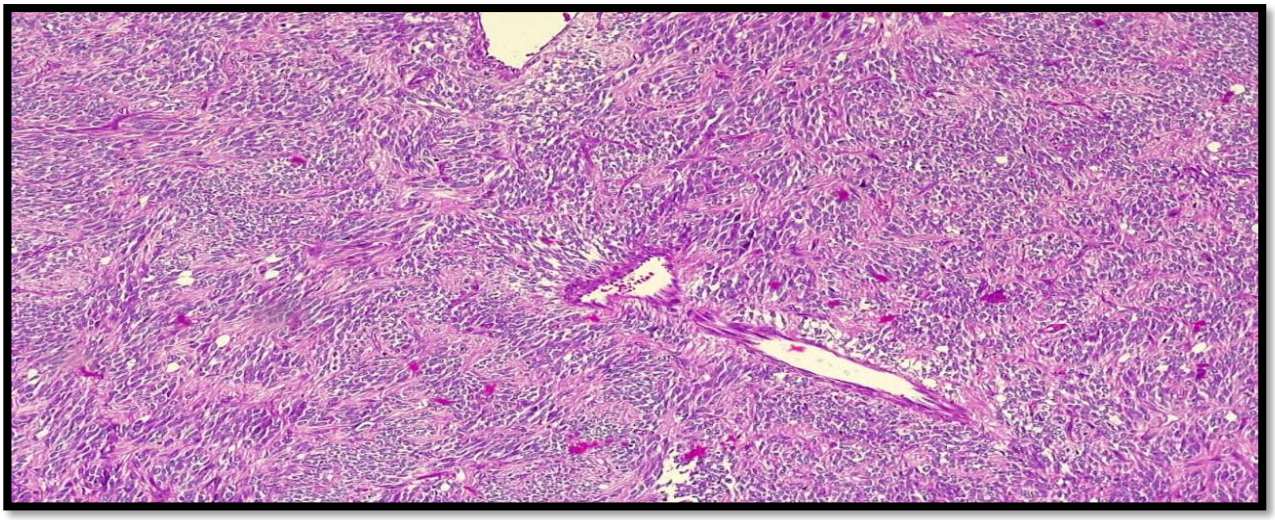

**Figure 1. Biphasic synovial sarcoma showing fascicles of closely packed spindle cells, epithelial cell component with round-oval nuclei and hemangiopericytoma-like vasculature (H&E, ×100)**

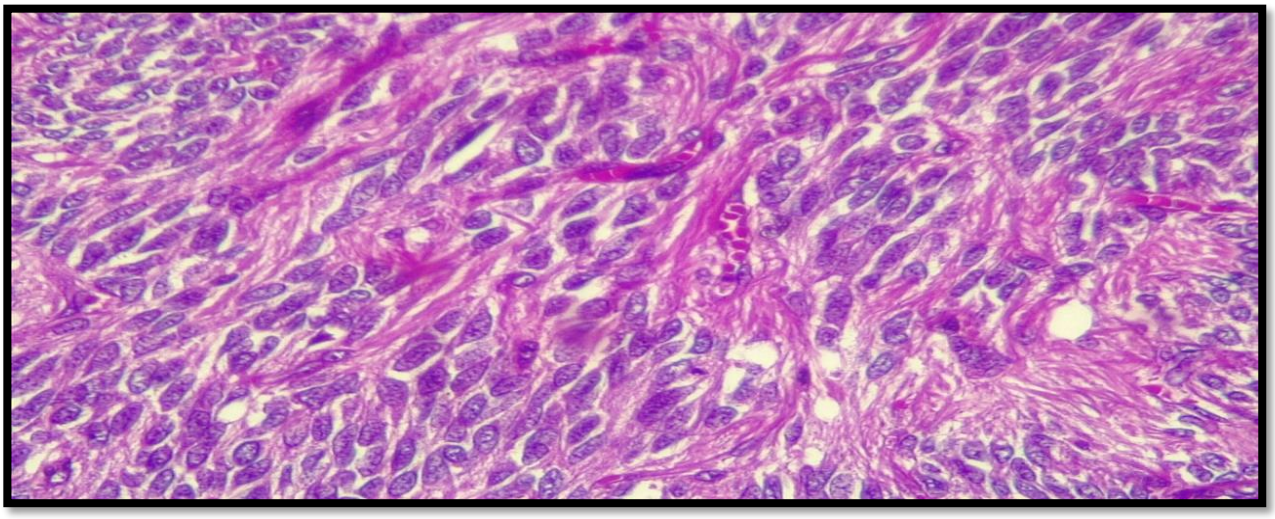

**Figure 2. Biphasic synovial sarcoma showing fascicles of closely packed spindle cells intermingled with epithelial cell component with round-oval nuclei (H&E, ×400)**

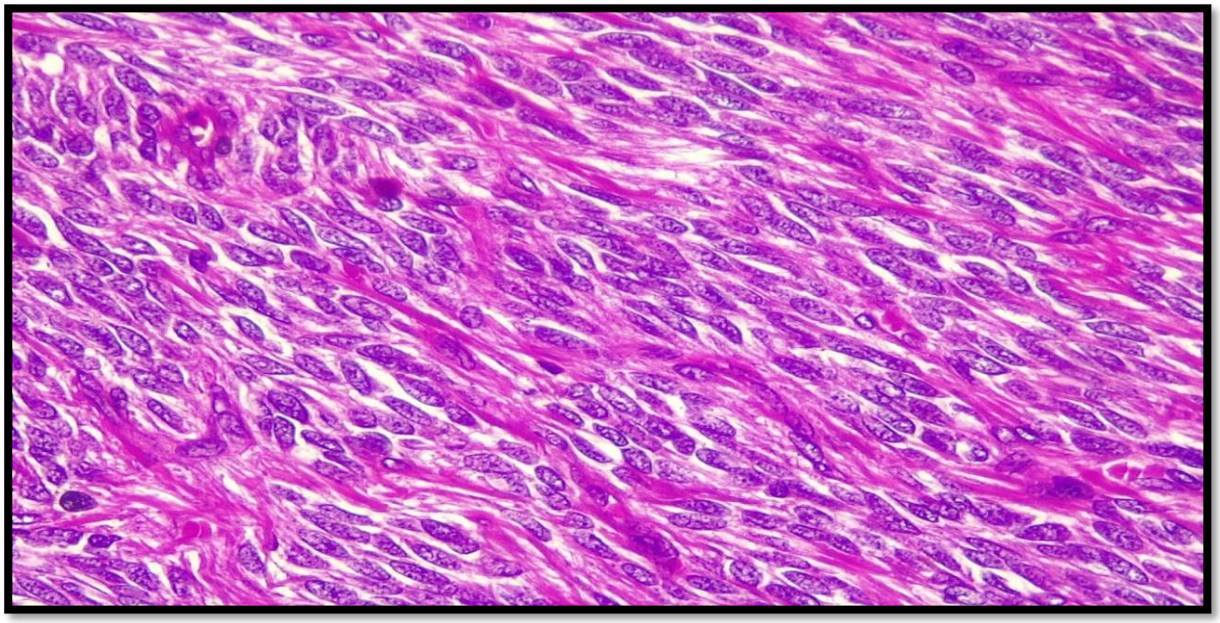

**Figure 3. Biphasic synovial sarcoma mainly consisting of fascicles of closely packed spindle cells (H&E, ×400)**

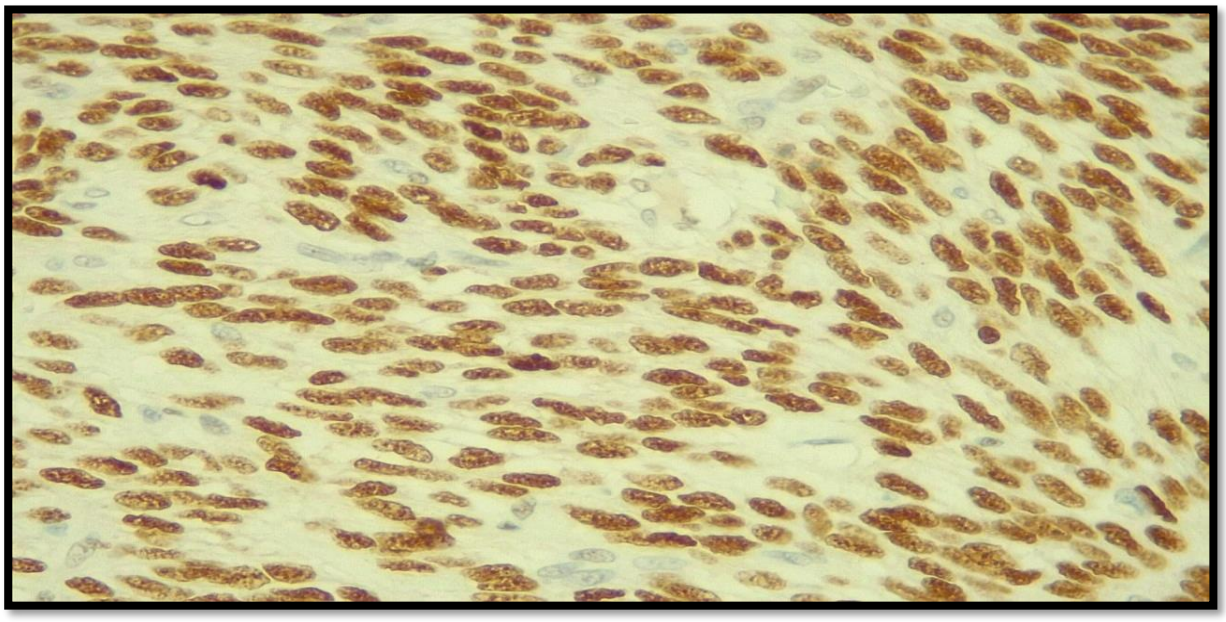

**Figure 4. TLE1 strong, diffuse nuclear expression of malignant cells (IHC, ×400)**

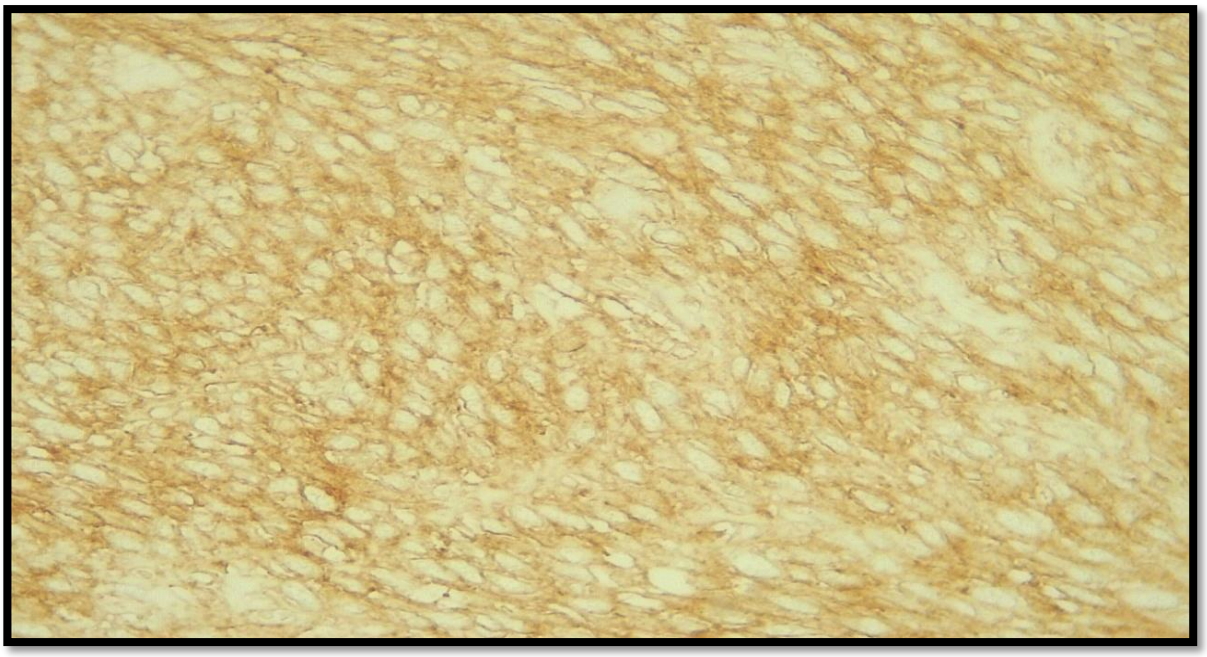

**Figure 5. MIC-2 (CD99) membranous and cytoplasmic expression of the malignant cells (IHC, ×100)**

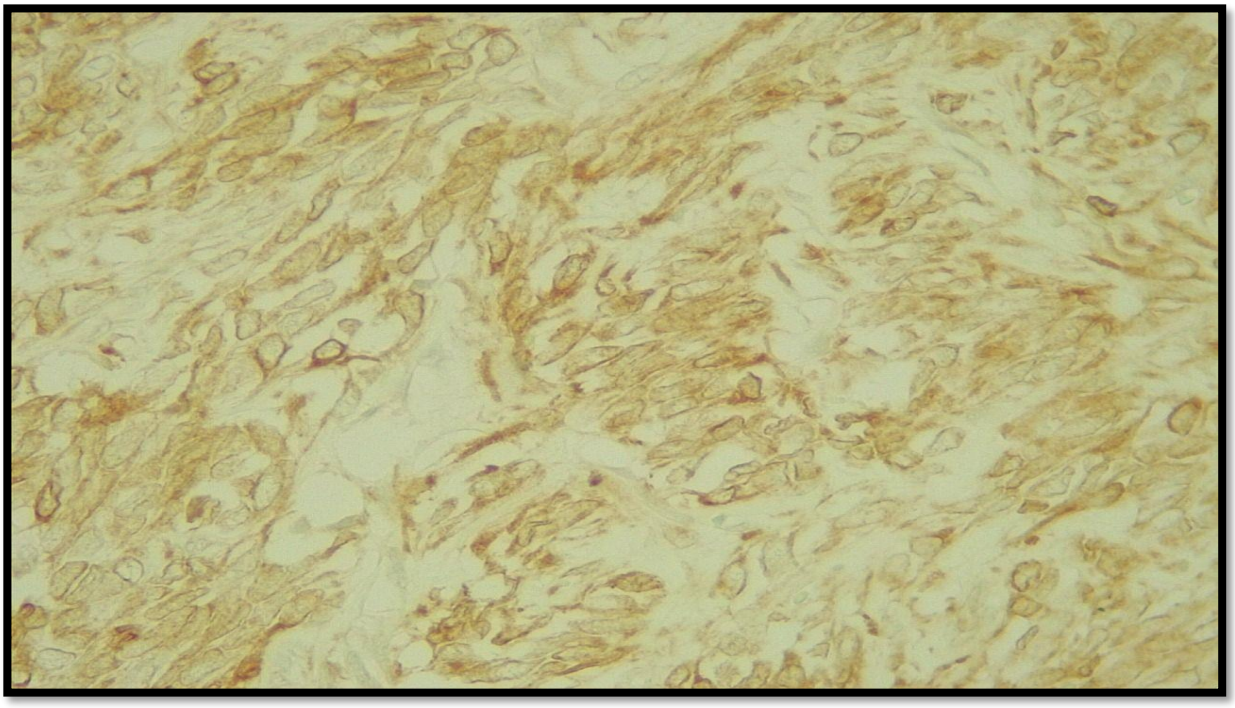

**Figure 6. BCL-2 positive expression of malignant cells (IHC, ×400)**

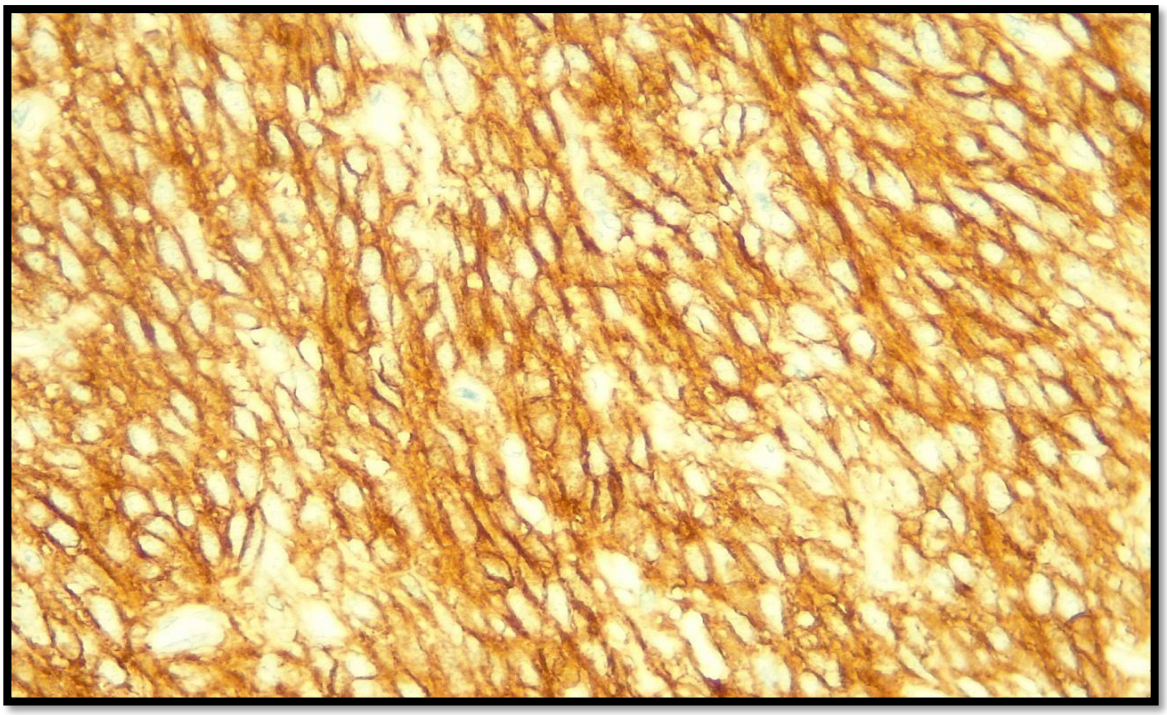

**Figure 7. CD56 positive expression of the malignant cells (IHC, ×400)**

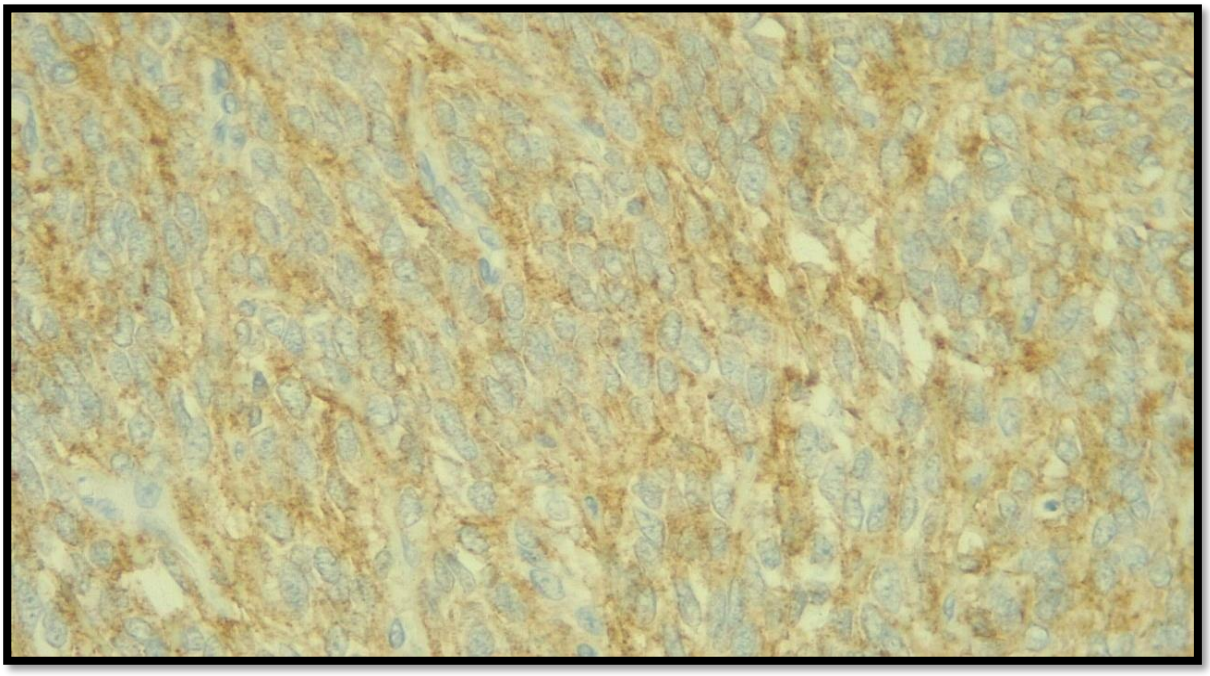

**Figure 8. EMA positive expression of the malignant cells (IHC, ×400)**

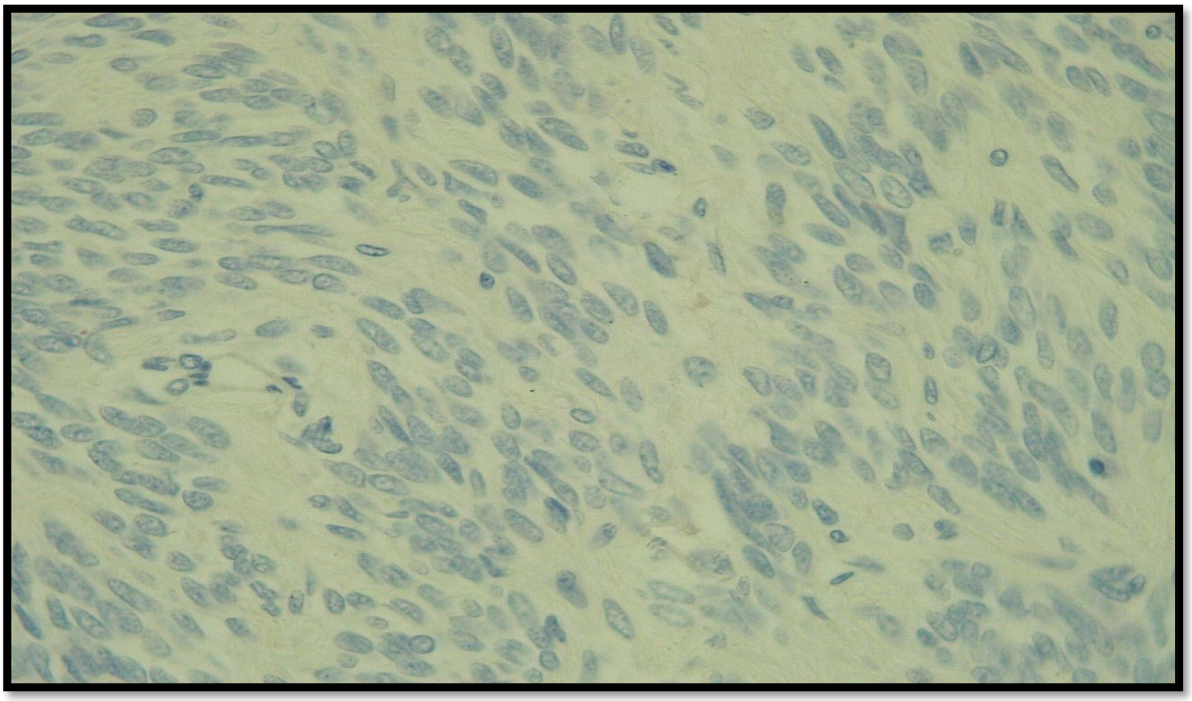

**Figure 9. Negative cytokeratin (AE1/AE3) expression of the malignant cells (IHC, ×400)**

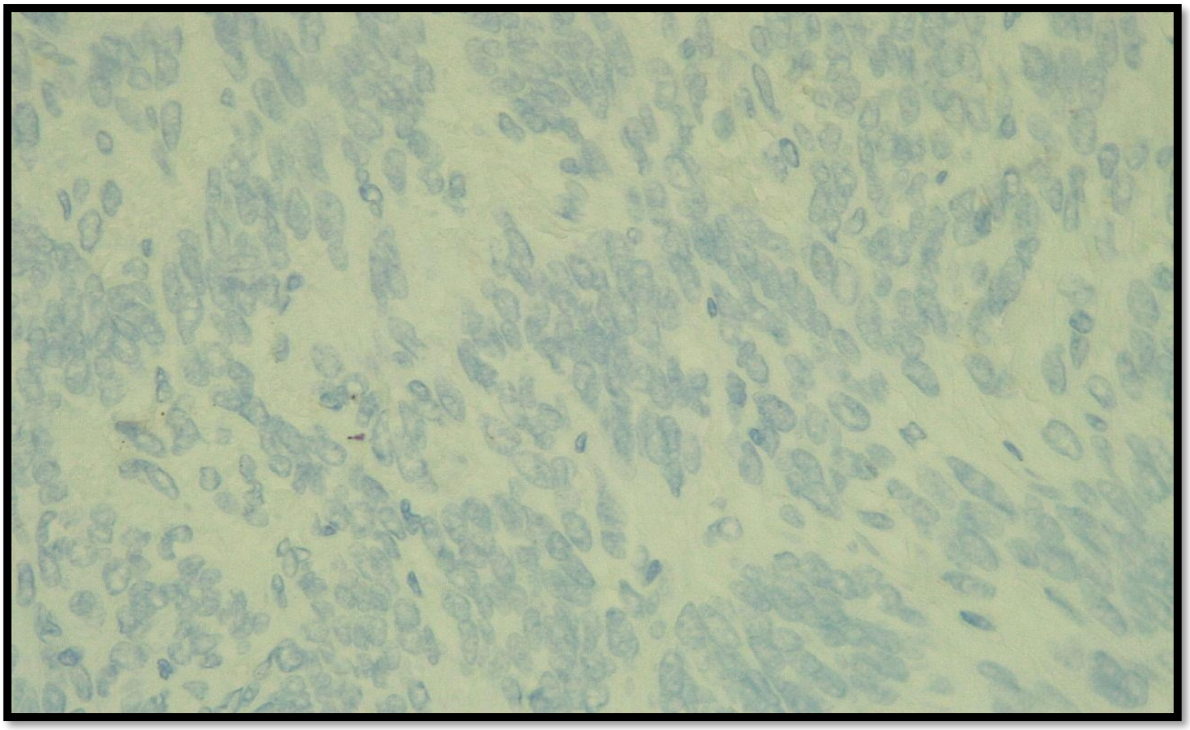

**Figure 10. Negative CD57 expression of the malignant cells (IHC, ×400)**

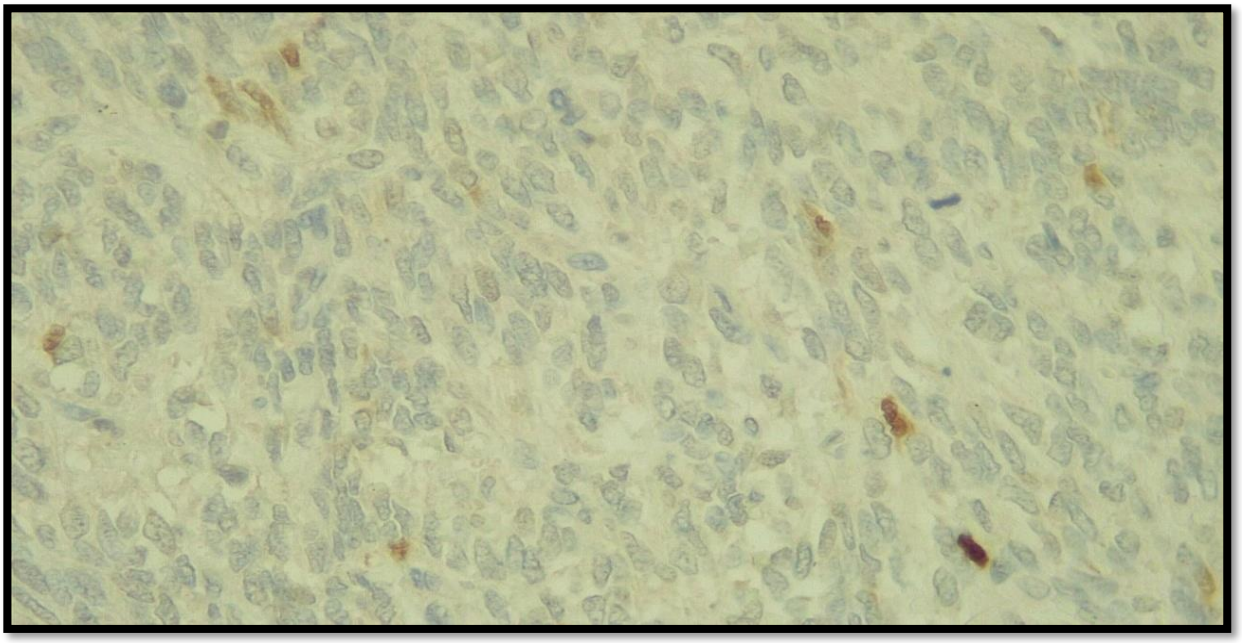

**Figure 11. Negative S100 expression of the malignant cells (IHC, ×400)**

## Discussion

Sarcomas of the head and neck region form a miscellaneous assembly of uncommon hostile malignant tumors, with the scalp, face and neck being the most common primarily involved locations.[3] Head and neck synovial sarcoma is high grade, uncommon malignant mesenchymal tumor representing around 3-5% of cases, [1,7] with the hypopharynx being the commonest site. [1,2,5,8,9,10] This malignant tumor can arise at any age but is predominant in adolescents and young aged individuals. [1,8,11,14,15] Treatment of this sarcoma is still debatable, however, total resection with wide surgical borders represents treatment of choice. [16] Radiotherapy is classically applied to enhance tumor control if excision is insufficient.[16] Till now, around one hundred reports of head and neck synovial sarcomas have been mentioned in literature,[3,17]meanwhile, tonsillar synovial sarcoma remains a very rare entity.[1,9] From a histological point of view, synovial sarcoma can manifest as monophasic (the commonest in the head and neck region)[4,9,14],biphasic or rarely poorly differentiated subtypes.[3,12,18] This malignant tumor can create diagnostic difficulties since the histological features may overlap with other soft tissue sarcomas and non-mesenchymal tumors[3,12] and since synovial sarcomas are regarded substantially more sensitive to chemotherapy than other types of soft tissue sarcomas, [3,19] precise diagnosis is a requisite. Here shines the role of immunohistochemical stains to assist in differentiating synovial sarcoma from other malignant tumors. Recently, several immunohistochemical markers are accessible, like EMA, cytokeratins, specific cytokeratins and vimentin, in addition to BCL-2, MIC-2, S100-P and CD56. [19] BCL-2, S100-P, CD99 and vimentin are expressed in the synovial sarcoma's spindle component.[10]About 90% of the cases express cytokeratin in the epithelial constituent and in a

small number of the spindle constituent of synovial sarcoma.[10] In addition, TLE1 has proven to be a specific and sensitive biomarker for synovial sarcoma that assists to confirm the diagnosis.[12] Despite that, some cases are difficult to diagnose and require confirmation by molecular analysis since most cases show a unique form of chromosomal translocation t(x;18) (p11.2; q11.2).[11,13,16,18,20,21] After reviewing former full-text published studies, we believe that primary tonsillar synovial sarcoma has been documented for only eighteen male patients (table-1). All patients were young adults excluding one case being above 60 years of age, meanwhile the youngest case was a 13-year-old boy. The right tonsil was the most common site while five cases involved the left tonsil and one study didn't mention the site of involvement. Histologically speaking, all cases were biphasic except three being monophasic synovial sarcomas and one study didn't document the histological type. In this study, we presented a 30-year-old male with primary tonsillar synovial sarcoma involving the right tonsil which was diagnosed as biphasic synovial sarcoma and a number of IHC markers were used to confirm the diagnosis and differentiate it from other soft tissue sarcomas and non-mesenchymal tumors. Malignant tumors with spindle cells in a fascicular pattern like malignant peripheral nerve sheath tumor (MPNST) and fibrosarcoma, might sometimes histologically mimic synovial sarcoma. However, MPNST shows focal nuclear S-100 expression, in contrast to our case that showed positive expression of EMA and negative expression of S-100 which was valuable to rule out MPNST[21]. In addition, the malignant cells in this study exhibited strong diffuse expression of TLE1 which is now regarded as a sensitive and specific biomarker for synovial sarcoma[12] (as mentioned earlier) and worked side by side with the conventional histopathological features to exclude other soft tissue sarcomas like fibrosarcoma. In summary, we described a case of a primary tonsillar synovial sarcoma with review of previous literature and although synovial sarcoma seldom involves the tonsil, it should still be kept in mind when evaluating tumors in this site.

**Table 1.** Review of previous cases with tonsillar synovial sarcoma

| The study | Age (years) | Gender | Clinical manifestations | Side of the tumor | Histological type | Immunohistochemical markers |
|---|---|---|---|---|---|---|
| **Shmookler et al, 1982** [22] | 35 | Male | Pediculated tonsillar lesions | Right | Biphasic | |
| **Shmookler et al, 1982** [22] | 34 | Male | Exophytic tonsillar mass with hemoptysis and stridor | | Biphasic | |
| **Engelhardt and Leafstedt, 1983** [23] | 25 | Male | Oral cavity discomfort | Right | Biphasic | |
| **Rangheard et al, 2001**[7] | 21 | Male | Dysphagia and shortness of breath | Left | Biphasic | Cytokeratins and vimentin |
| **Pappo et al, 2005** [9] | 25 | Male | | Right | Biphasic | |
| **Harb et al, 2007** [9] | 35 | Male | | Right | Biphasic | |
| **Harb et al, 2007** [9] | 34 | Male | | Left | Biphasic | |
| **Ishiki et al, 2009** [17] | 19 | Male | Pain on swallowing | Right | Biphasic | Cytokeratins and vimentin |
| **Vogel et al, 2010** [5] | 31 | Male | Left sided sore throat and dysphagia | Left | Biphasic | Cytokeratin (AE1/AE3), EMA, CD99, BCL2, vimentin and calponin |
| **Khademi et al, 2010** [3] | 23 | Male | Sore throat, headache, ear pain and tonsillar mass | Left | Monophasic | Cytokeratin, vimentin, BCL2 and MIC-2 |
| **Khademi et al, 2010** [3] | 26 | Male | Throat discomfort and bleeding | Right | Biphasic | Cytokeratin, vimentin, BCL2 and MIC-2 |
| **Soria-Ce´spedes et al, 2013** [1] | 63 | Male | Progressive dysphagia | Right | Monophasic | CD99, ,OSCAR, EMA, BCL2, PGP 9.5 vimentin and TLE1 |
| **Singh Virk et al, 2013**[4] | 17 | Male | Sore throat and a throat lump | Left | Biphasic | |
| **Crowson et al, 2015** [9] | 23 | Male | | Right | Monophasic | |
| **Crowson et al, 2015** [9] | 26 | Male | | Right | Biphasic | |
| **Van der Graaf et al, 2017**[9] | 19 | Male | | Right | Biphasic | |
| **Alessandro, 2019** [24] | 17 | Male | Sore throat and dysphagia | Right | | |
| **YALÇIN K et al, 2020** [9] | 13 | Male | Dysphagia and snoring | Right | Biphasic | PanCK, CK19, CK18, CK7 and vimentin |
| **Present case** | 30 | Male | Difficulty on swallowing, odynophagia and sore throat | Right | Biphasic | TLE1, CD99, BCL2, CD56, EMA, cytokeratin(AE1/AE3) CD57 and S-100 |

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## الساركوما الزليلية اللوزية مع مراجعة الأدبيات

***الخلاصة:*** الساركوما الزليلية هي ورم لحمي متوسطي خبيث يمكن أن ينشأ في أي موقع، وغالبًا ما يصيب الأنسجة الرخوة العميقة في ألاطراف لدى البالغين الشباب، ومع ذلك فهو غير شائع في منطقة الرأس والرقبة ونادر جدًا في اللوزتين. قمنا بتسجيل حالة رجل يبلغ من العمر ٣٠ عامًا أصيب بكتلة في اللوزه اليمنى بعد ان عانى ٦ أشهر من صعوبة في البلع، الم عند البلع, وآلم في الأذن اليمنى مع التهاب في الحلق. تم استئصال الورم وإرساله لإجراء دراسات التشريح المرضي والكيميائي المناعي التي أكدت تشخيص الساركوما الزليلية ثنائية الطور في اللوزة اليمنى.

***الكلمات المفتاحية:*** الساركوما الزليلية، ثنائية الطور، الدراسات الكيميائية المناعية