# Parapharyngeal Synovial Sarcomas – A Case Report

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## Introduction

Parapharyngeal Space (PPS) tumors are rare entities and mostly benign accounting for 80% of total cases while remaining 20 % are malignant accounting for less than 0.5% of Head and Neck tumors (H&N). Benign lesions from salivary glands or neurogenic cells are most common with pleomorphic adenomas being the commonest one. Synovial sarcomas are further less common pathology among H and N malignancies [1]. Parapharyngeal synovial sarcomas are the least common variants among a total of fewer than 200 cases of PPS tumors that have been reported worldwide.

These tumors are usually diagnosed during their third and fourth decade of life with male predominance. Primary parapharyngeal synovial sarcoma is diagnosed based on histological and immunohistochemical characteristics [2]. Patients are most commonly present with the compressive or infiltrative symptom of slow-growing painless mass. Our presentation aims to describe through a clinical case the clinical, radiological, and histological features of c and to discuss its therapeutic management.

## Mini Review

A 19 yrs old boy presented to the oncology outpatient department with chief complaints of gradually progressive swelling in the right side of the neck for 6 months. On examination, there was a firm 9 x 8 cm mass at the level of right cervical level Ib/II. There was no associated trismus, ankyloglossia, or OSMF. Further evaluation with Contrast-Enhanced (CE) Computed Tomography (CT) scan revealed large heterogeneous soft-tissue attenuation mass with calcification with mild enhancement in the right side of the neck with epicenter at right supra hyoid parapharyngeal space arising from the medial pterygoid muscle with probable infiltration/inferior displacement of the submandibular muscle. Size of lesion was 10.4 x 7.1 x 6.9 cm. The internal jugular vein was not visualized in the proximal neck, secondary to compression/thrombosis [3]. Trucut biopsy revealed cellular tumor comprising of monomorphic spindled cells arranged in fascicles with scant intervening stroma. Cells showed nuclear overlapping at places and possessed elongated to spindled nuclei with evenly distributed chromatin, inconspicuous nucleoli, and scant cytoplasm with ill-defined borders. No evidence of necrosis or increased mitotic activity was seen. Features were suggestive of spindle cell tumors. Immunohistochemistry revealed that tumor cells were immunoreactive for TLE-1 (Strong, nuclear in >90% cells), BCL-2(>50% cells), EMA, and CK (focal, intermediate staining intensity), vimentin (diffuse) and CD34 (focal). Tumor cells were immunonegative for S-100, desmin, and H-caldesmon. Morphologic and Immunoprofile were suggestive of monophasic Synovial sarcoma. Metastatic workup was negative for distant metastasis.

The patient underwent an excision of a neck tumor under general anesthesia. Final post-op histopathology revealed 8.0 x 6.0 x 2.0 cm size tumor with No evidence of necrosis or increased mitotic activity is seen. Tumor cells are immunoreactive for TLE1 (>70%), Bcl2: diffuse, strong while negative for CK, CD34, S100, and SMA. There was no lymph node metastasis out of 20 dissected lymph nodes. Morphology and immune profile were consistent with monophasic synovial sarcoma. The patient was then planned and delivered external beam radiation therapy to a total dose of 63Gy/30fractions@2.1Gy/fraction using a 6 MV photon beam. The patient tolerated the treatment with grade III mucositis which was conservatively managed.

## DISCUSSION

Primary Parapharyngeal Synovial Sarcoma (PPSS) accounts for less than 10% of cases among rare tumors of PPS. PPSS usually present mainly in young adults with median age of 37 years and male preponderance. Analysis of SEER database has revealed that median Overall Survival (OS) at 2, 5, and 10 years was 77%, 66%, and 53%, respectively among 167 cases identified. This analysis also revealed that size >5 cm and stage at presentation are independent determinants of survival include. However, histological subtype, surgical resection and radiation therapy were not found to be independent determinants of survival. Histologic grade is a significant prognostic factor for both disease-specific survival and metastasis-free survival.

Jernstrom first described this entity in hypopharynx at head and neck congress in 1954. PPSS has been described based on histological and immunocytochemical examination. Synovial Sarcomas (SS) most commonly arise adjacent to joints, especially around the knee joint. However, SS does not originate from synovial tissue but from pluripotent mesenchymal cells that might be adjacent or distant from articular surfaces. The origin of SS has been proposed to be from malignant degeneration of these primitive mesenchymal cells.

There are no specific distinguishing clinical or radiological features of PPSS. Patients are asymptomatic unless tumor attains significant size to cause pressure effects like progressive dysphagia or even Dyspnea [4]. Classic SS has two subtypes i.e biphasic (spindle and epithelial cells) and monophasic. The mono-phasic variant is more commonly seen that its biphasic counterpart and is characterized either spindle or less commonly epithelial cell morphology. However, biphasic subtype is usually composed of both distinct epithelial and spindle cell components, hence its nomenclature. Our patient had monophasic variant with single cellular component. Morphological differential diagnosis includes fibrosarcoma, malignant schwannoma or salivary gland tumor. Molecular analysis is useful to conclude definitive diagnosis, especially when the tumor is at unusual location, such as the PPSS. 90% of H&N SS is related to a specific translocation between chromosome X and 18, t(x; 18) (p11.2; q11.2). On CECT scan, these tumors are solid multilocular mass with smooth margins that enhance heterogeneously with contrast. Calcification, if seen is considered as good prognostic factor [5]. However it is seen only on 30%-60% of SS. On MRI, SS has intermediate intensity on T1-weighted sequences and of variable intensity on T2-weighted sequences, with contrast enhancement. Both CT and MRI are helpful to determine any local extent and lymph node involvement.

## Conclusion

Despite high probability of local recurrence approaching 40-80% in adult and 90% in childhood, surgery remains the treatment of choice. Surgery may be an extensive resection requiring bone or nerve sacrifice. Since lymph node metastases are uncommon prophylactic neck dissection is generally not indicated. Adjuvant radiotherapy seems to have improved local control. Chemotherapy has no significant impact on the disease-free interval in H&N synovial sarcomas. The therapeutic

regimen should eventually include wide surgical excision of the tumor to ensure free margins followed by radiation therapy when appropriate, with or without adjuvant chemotherapy. Combined modality therapy yields better results; however, the 5-year survival rate of these patients is poor and ranges from 25% to 55%. High metastatic potential is seen in lung and bones. Local and regional recurrences occur approximately in 80% of cases, with lung metastasis being the usual cause of death in patients with SS of the H&N. Favorable prognostic indicators include an early diagnosis, small tumor size (<5cm) and younger age (<20 years) and a wide surgical excision. Immunohistochemistry also plays a part in diagnosis. SSs are known to have positivity for Epithelial Membrane Antigen (EMA), Bcl-2, CK7, CD 99, and TLE1 and are negative for CD34. BCL-2 is one of the most sensitive tumors marker for SS.

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