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Case Report

Primary biphasic synovial sarcoma in a rare anatomical site in an adolescent: a case report

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ABSTRACT

Synovial sarcoma is a rare malignant soft tissue tumor that predominantly affects adolescents and young adults, commonly arising in the extremities. Head and neck involvement is uncommon, accounting for only 0.7% to 10% of all synovial sarcoma cases. The parapharyngeal space is an especially rare site, representing approximately 0.5% of head and neck tumors. We present the case of an adolescent male with a progressively enlarging mass in the parapharyngeal space. Clinical and radiological evaluation suggested a mesenchymal lesion. Fine-needle aspiration cytology (FNAC), followed by histopathological examination and immunohistochemistry, confirmed the diagnosis of synovial sarcoma. The tumor displayed characteristic biphasic histology and immune profile. The patient underwent surgical resection and was later lost to follow-up. This case highlights the diagnostic challenges posed by mesenchymal tumors in unusual locations such as the parapharyngeal space. A multidisciplinary approach incorporating cytology, histopathology and immunohistochemistry is essential for accurate diagnosis and optimal management. Awareness of such rare presentations is important, especially in adolescents, to avoid misdiagnosis, delayed treatment and recurrence.

Keywords: Adolescent, Case report, Head and neck tumor, Immunohistochemistry, Parapharyngeal space, Synovial sarcoma

INTRODUCTION

Synovial sarcoma (SS) is a rare malignant mesenchymal tumor that accounts for 5–10% of all soft tissue sarcomas. It typically affects adolescents and young adults, usually arising in the deep soft tissues of the extremities near large joints.[1] Head and neck involvement is uncommon, comprising less than 10% of all synovial sarcoma cases.[2,3]

Within this region, the parapharyngeal space is an exceptionally rare site, representing only 0.5% of head and neck tumors.[4] Due to its rarity and nonspecific presentation, primary parapharyngeal synovial sarcoma (PPSS) is often misdiagnosed as more common tumors such as pleomorphic adenoma or schwannoma.[5] Parapharyngeal space (PPS) tumors are rare, constituting approximately 0.5% of all head and neck neoplasms and presenting a diagnostic and therapeutic challenge due to their deep location and proximity to vital structures.[5,6] These tumors can be benign or malignant, with salivary gland and neurogenic tumors being the most common histological types.[2] Radiological imaging plays a crucial role in assessing tumor size, extent and anatomical relationships but is usually insufficient for definitive diagnosis.

Histopathological evaluation, along with immunohistochemical (IHC) analysis, is essential for accurate identification and subtyping of synovial sarcoma. Immunoprofiling typically includes positivity for epithelial markers (e.g., cytokeratin, EMA) and mesenchymal markers (e.g., vimentin), with molecular

confirmation via detection of the SYT-SSX gene fusion being diagnostic in such cases.[3,6] In this report, we present a rare case of primary synovial sarcoma arising in the parapharyngeal space of an adolescent male.

This case underscores the need for high clinical suspicion, especially when encountering unusual presentations of neck masses in young individuals and highlights the critical role of cytology, histopathology and immunohistochemistry in confirming the diagnosis.

CASE REPORT

A 15-years-old male presented with complaints of a progressively enlarging intraoral mass extending into the right lateral neck, associated with pain, dysphagia, intermittent fever and trismus for one month. The patient denied any history of trauma, prior infections or systemic illness.

On clinical examination, a firm to hard mass measuring approximately 2×2 cm was palpated in the right lateral neck region. The swelling was mildly tender and exhibited limited mobility, particularly with neck movement. Intraorally, the mass was palpable in the right parapharyngeal space, causing mild medial displacement of the oropharyngeal structures.

Contrast-enhanced computed tomography (CT) of the neck revealed a well-defined, heterogeneous soft tissue lesion measuring 4.5×4.5×1.8 cm. The lesion showed mild post-contrast enhancement and was noted to involve the medial pterygoid muscle, extending medially into the parapharyngeal space. No obvious bony erosion or regional lymphadenopathy was identified.

Fine-needle aspiration cytology (FNAC) was performed, revealing a highly cellular smear composed of dispersed and clustered spindle to oval cells with mild pleomorphism, raising suspicion of a mesenchymal neoplasm (Figure 1a, b). Further histopathological evaluation was advised for definitive diagnosis.

Histological examination of tissue obtained via core needle biopsy revealed a biphasic tumor composed of both epithelial and spindle cell components. The epithelial component was arranged in solid nests and sheets of round to oval cells with moderate anisonucleosis and pale to eosinophilic cytoplasm. The spindle cell areas showed elongated cells arranged in interlacing fascicles with moderate nuclear pleomorphism and indistinct cytoplasmic borders (Figure 1c, 1d). Mitotic activity was sparse and focal areas of necrosis were noted. IHC studies were performed which showed cytoplasmic positivity for cytokeratin, EMA along with nuclear positivity for TLE1 and Bcl-2. It was negative for S-100 and melanocytic markers. Based on the histological and immunohistochemical findings, a diagnosis of biphasic synovial sarcoma of the parapharyngeal space was confirmed.

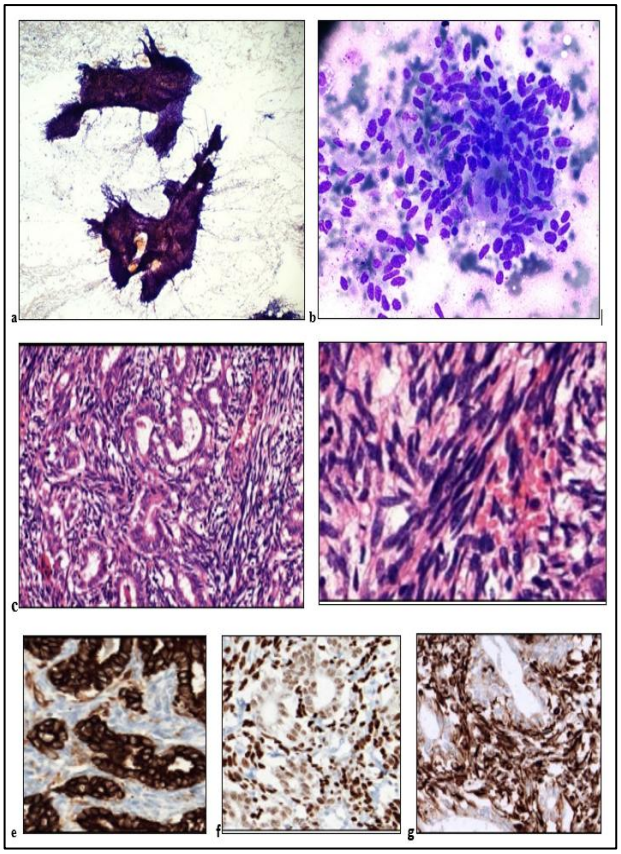

Figure 1 (a): PAP-stained smear right from neck swelling are highly cellular arranged in sheets and clusters (Scanner view). (b) MGG stained smear showing oval to elongated spindle cells with hyperchromatic nuclei (40X). (c) H&E-stained section show biphasic component with prominent round to oval epithelial cells with moderate anisonucleosis (20X). (d) H&E-stained section show spindle cell with fascicular growth pattern (40X). (f) Epithelial component showing cytokeratin positivity. (g) Spindle cell component are positive for vimentin. (h) Diffuse and uniform positivity for TLE-1.

DISCUSSION

Synovial sarcoma is a rare, aggressive soft tissue sarcoma that typically arises near large joints of the extremities in adolescents and young adults. While it accounts for 5–10% of all soft tissue sarcomas, its occurrence in the head and neck region is distinctly uncommon, constituting less than 10% of all synovial sarcoma cases.[1,3] Among head and neck sites, the parapharyngeal space (PPS) is an exceptionally rare location, with very few cases of primary parapharyngeal synovial sarcoma (PPSS) reported in the literature.[1,5]

The PPS is a deep anatomical compartment of the neck, bordered by critical neurovascular structures, making tumors in this region difficult to detect early and challenging to manage surgically. Most PPS tumors are benign, with salivary gland and neurogenic origin being

the most common.[2] As such, malignant mesenchymal neoplasms like synovial sarcoma in this space may be misdiagnosed clinically and radiologically. In this case, the adolescent patient presented with a firm, tender neck mass with accompanying symptoms of dysphagia, trismus and fever features that are nonspecific and could mimic infectious or benign neoplastic conditions.

Imaging studies such as contrast-enhanced CT or MRI are essential for determining tumor size, anatomical extent and surgical planning. However, imaging alone cannot establish a definitive diagnosis. FNAC may suggest a mesenchymal tumor, but histopathological examination with immunohistochemistry (IHC) remains the gold standard for diagnosis. In this case, histology showed a biphasic pattern with both epithelial and spindle cell components typical of biphasic synovial sarcoma, which accounts for approximately 20–30% of all synovial sarcomas.[6]

Immunohistochemistry is essential for distinguishing synovial sarcoma from other spindle cell neoplasms. Typical markers include positive staining for cytokeratin, epithelial membrane antigen (EMA), Bcl-2 and TLE1, along with negative staining for S-100, desmin and CD34, which helps rule out neural, myogenic and vascular tumors.[7,8] Molecular studies detecting the SYT-SSX1 or SYT-SSX2 fusion transcripts resulting from the t(X;18)(p11.2;q11.2) translocation are considered diagnostic and may aid in cases with ambiguous morphology.[9] Treatment typically involves wide surgical excision with or without adjuvant radiotherapy. However, achieving complete resection with negative margins in the PPS is often challenging due to anatomical constraints. Chemotherapy may be considered in high-grade or unresectable cases, although its role remains controversial.[10] Long-term follow-up is essential, as synovial sarcoma has a high risk of local recurrence and distant metastasis, particularly to the lungs.[11]

This case highlights the importance of considering rare sarcomas such as synovial sarcoma in the differential diagnosis of PPS tumors, especially in adolescents. Early biopsy, histopathological analysis and multidisciplinary management are critical to ensuring timely diagnosis and appropriate treatment.

CONCLUSION

Synovial sarcoma is an aggressive tumor, thus accurate diagnosis depends on morphologic and immunohistochemical examination along with proper molecular analysis (RT-PCR). One should keep it as possible differential even at young age or unusual site, in cases diagnosed as mesenchymal lesion on cytopathology. Eventually biphasic morphology should prompt pathologist for immunohistochemical or molecular confirmation. Like other sarcomas, definite diagnosis and tumor radical surgical resection at an early stage remain the mainstay of therapy. Thus a definitive diagnosis requires a multidisciplinary approach, integrating clinical, radiological, histopathological and immunohistochemical findings. Early recognition and accurate diagnosis are essential for timely management and improved outcomes.

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