ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 9, Issue – 5, October – 2024 , Page No. : 22 – 26

Out of the Ordinary Case of Primary and Recurrent Synovial Sarcoma of the Head and Neck: Two Case Reports

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Citation this Article: Victor Mondal, N. Priyadarshini, Manickavasagam, Ravichandran, "Out of the Ordinary Case of Primary and Recurrent Synovial Sarcoma of the Head and Neck: Two Case Reports", IJMSIR - October - 2024, Vol – 9, Issue - 5, P. No. 22 – 26.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Synovial sarcoma is a rare and aggressive form of cancer, infrequently occurring in the head and neck region. This article aims to present an in-depth review of the current literature on primary synovial sarcoma affecting the head and neck. By analyzing existing studies, this review seeks to provide insights into its clinical features, diagnosis, management, and outcomes, helping to enhance understanding of this uncommon but serious malignancy in an anatomically complex and critical area.

Keywords: synovial sarcoma, head and neck, cd 99, TLE, Neoadjuvant chemo.

Introduction

Indeed, sarcomas represent a rare subset of malignancies, constituting approximately 1% of all cancer cases. Among sarcomas, around 80% originate from soft tissues, while the remaining 20% arise from bone. Soft tissue sarcomas develop from mesenchymal pluripotent cells, which include muscle, fat, fibrous tissue, blood vessels, and other supportive tissues of the body

Synovial sarcomas account for approximately 8% to 10% of all soft tissue sarcomas. Among these sarcomas, 3% to 10% occur in the head and neck region, with soft palate synovial sarcomas representing only 1% of head and neck cases. In terms of the male-to-female ratio for synovial sarcoma occurrence in the soft palate specifically, there isn't a specific ratio due to rarity that is widely reported in medical literature specific to India. The most frequent presentation is an asymptomatic, painless mass. We present two rare cases of synovial sarcoma of head and neck.

Case Report 1

A 54-year-old male presented to the otorhinolaryngology department with a swelling in his soft palate. The swelling had been present for 6 months and had progressively increased in size over the past two months. Difficulty in swallowing food. Notably, there was no history of pain associated with the swelling. Upon physical examination, the swelling was approximately 2 x 4 cm mass noted in the junction of the soft palate and hard palate. It had a firm consistency and was non-tender to palpation. However, the margins of the swelling were

not well-defined. Mucosa appears pink and indurated. Extending anteriorly 3 cm from the carina of teeth, rally up to left molar, medially up to midline. The patient's mouth opening was adequate. Bilateral level II cervical lymphadenopathy. MRI (fig1case1) revealed heterogeneously enhancing complex solid cystic lesion, predominently involving the left half of soft palate and adjacent hard palate, with lobulated and irregular margins, measuring ~4.0x2.0x1.6 mm (APXTRXCC). PET CT showed FDG avid lesion in the left side of soft palate with no invasion of adjacent structures. Bilateral level II cervical lymphadenopathy (FIG1:CASE1). Anteriorly – The lesion is noted to extend into the hard palate. Posteriorly Extraneous indentation over the oropharyngeal lumen causing mild luminal narrowing, lumen measuring ~ 5 x 6 mm (AP x TR). o Medially the lesion is seen to extend beyond the midline into the right. o Laterally – extending into the left retromolar trigone.

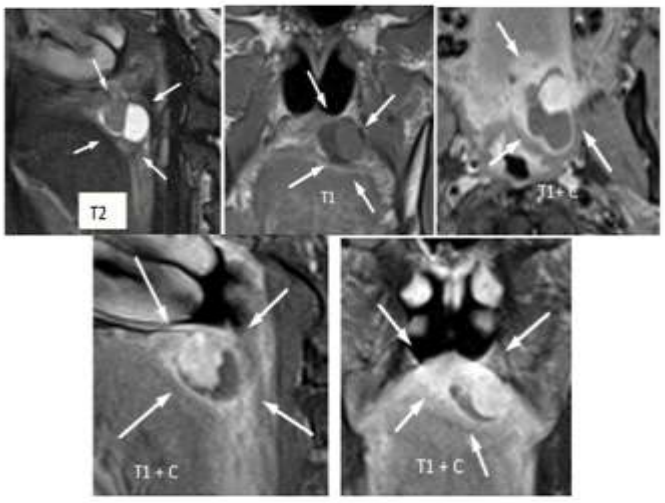


Figure 1:

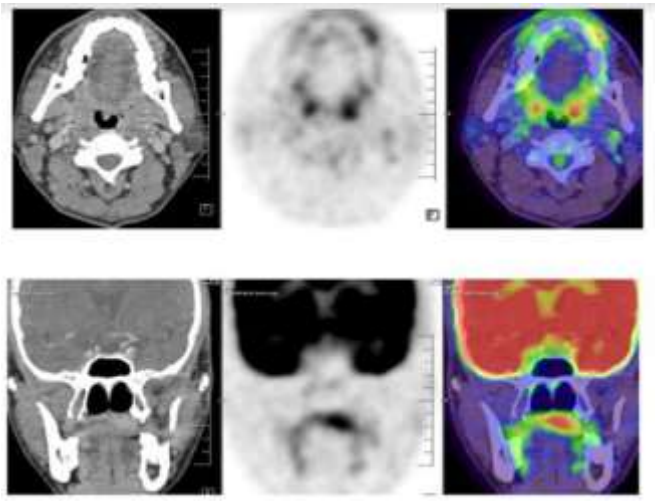
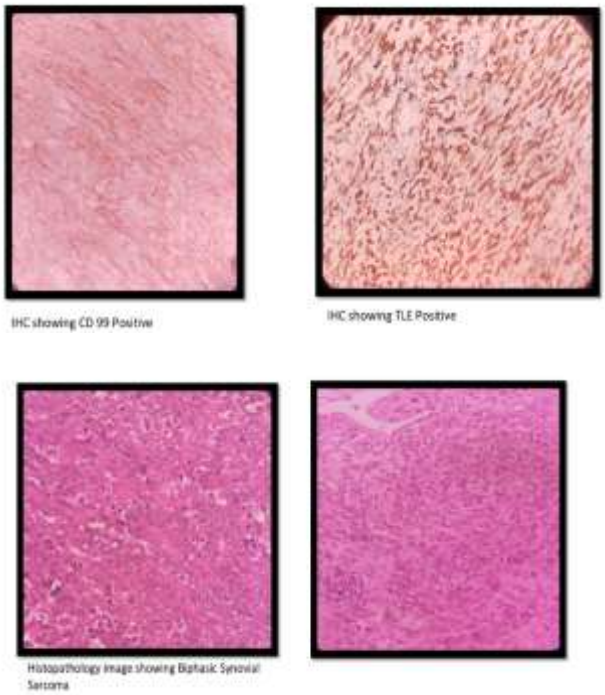


Figure 2:

PET CT showed (Figure 2 case 1) FDG avid lesion in the left side of soft palate with no invasion of adjacent structures. Bilateral level II cervical lymphadenopathy.

Patient underwent biopsy and the histopathological examination revealed spindle cells arranged in sheets and fascicles, characterized by round to oval nuclei with vesicular chromatin patterns, prominent nucleoli, and scanty cytoplasm. The tumor exhibited high cellularity, small areas of necrosis, and atypical mitotic activity. Overall, the histomorphology favours a diagnosis of malignant mesenchymal tumor. Immunohistochemistry studies demonstrated positive expression for vimentin, CD99 and TLE1, focally positive for desmin and negative for SOX 10, CD34, MyoD1, P63 and CK. Ki 67, proliferative index, is 30%. The diagnosis of high-grade spindle cell sarcoma possibly synovial sarcoma was given. The patient was started on neo-adjuvant chemotherapy with 6 cycles of Inj ADRIAMYCIN, Inj IFOSFOMIDE followed by surgery.



Case Report 2

51year old man is known case of carcinoma of base of tongue in 2019, cT4aN2M0. Patient had received CCRT (6Gray 33 fraction +5 dose of Cisplatin). Completed on 11/11/2019. Patient was noted to be clinically and radiologically disease- free post therapy. Disease free status for 4 years presented with swelling over right lateral border of tongue. For evaluation of the same with MRI Head and Neck (Fig3 case2) was done showed relatively well defined T2 and STIR hyperintense lesion showing restricted diffusion measuring 1.5x1.7x1.9 cm. PET CT image showed -FDG avid lesion 1.9x1.4x1.8 cm (figure 4case2)

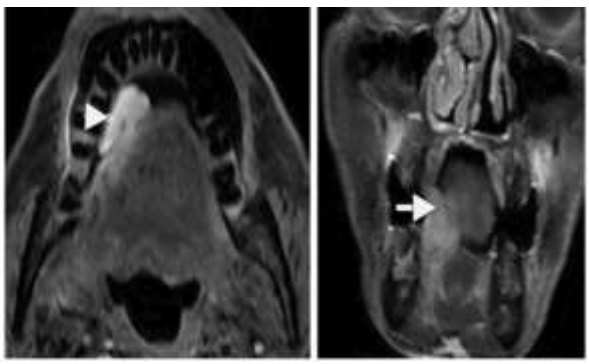


Figure 3

Biopsy was done histopathology revealed pleomorphic sarcoma. IHC was strongly positive for vimentin and negative to PANCK and p40 (Fig 5 case2)

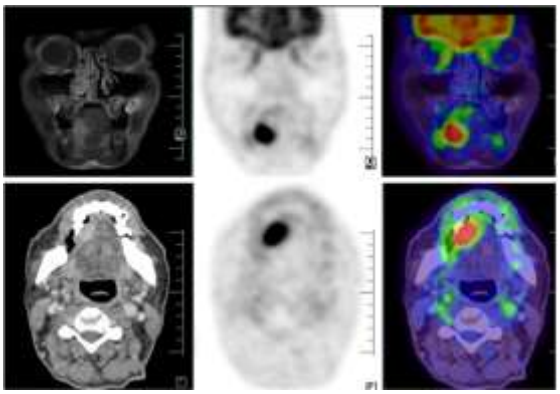


Figure 4

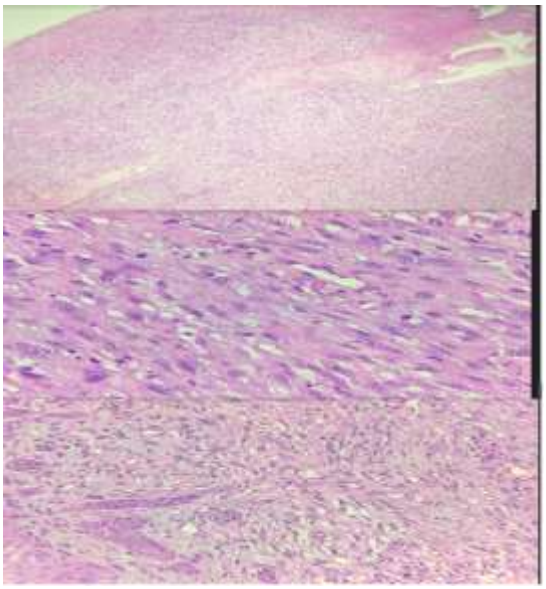


Figure 5

Patient was undergone 3 cycle of neoadjuvant chemo with Inj DOXORUBICIN and Inj IFOSFAMIDE, Inj MESNA. Response evaluation done with Whole body PET CT.

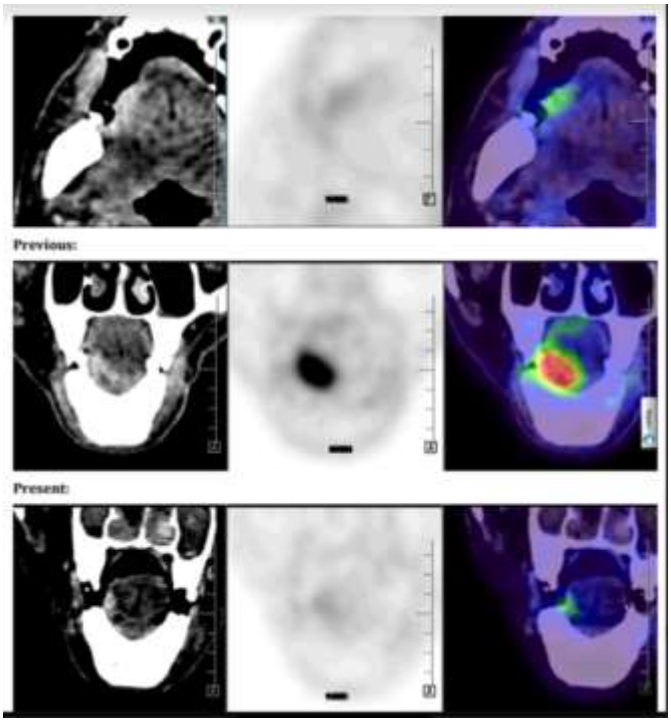


Figure 6

Pet Ct Impression

- Significant resolution of the relatively ill -defined malignant lesion in right lateral tongue.
- No significant regional lymphadenopathy
- No distant metastasis

Patient was taken for surgery. Composite resection of right lateral border of tongue was done. No evidence of residual tumor. Resection margin is free from tumor.

Discussion

Soft tissue sarcomas in the head and neck regions account for approximately 3 to 10% of cases. These tumors can occur in various locations, including the prevertebral space, from the base of the skull to the hypopharynx, retropharyngeal and parapharyngeal spaces, anterior neck along the edges of the sternocleidomastoid muscle, and sites within the oropharynx and larynx.

Histopathologically, synovial sarcoma can be classified into four types:

1. Biphasic type: This type exhibits both epithelial and spindle cell components, which are present in various patterns and extents.
2. Monophasic epithelial type: Here, only the epithelial component is observed.
3. Monophasic spindle cell type: This subtype consists solely of spindle cells.
4. Poorly differentiated type: These tumors lack clear differentiation.

Immunohistochemically, both epithelial and spindle cells in synovial sarcoma stain positive for epithelial membrane antigen (EMA) and cytokeratin. However, only the spindle cells express vimentin. CD99 is seen in approximately 67% of all cases.

The tumor is associated with high recurrence rates, typically ranging from 60% to 90%, occurring within 2 years despite therapeutic interventions. Most metastases originate from hematogenous dissemination, although up to 20% can spread via the lymphatics to regional lymph nodes, leading to clinically enlarged lymph nodes. Routine cervical lymph node dissection is not performed unless enlarged lymph nodes are present. Ultimately, hematogenous dissemination is the primary cause of mortality.

Conclusion

Although synovial sarcomas exhibit relatively slow growth, the 5-year survival rate remains poor, even when they occur in the head and neck region. Prognosis is influenced by several factors, with tumor size and deep extension at the time of primary treatment being the most significant. Survival tends to decrease as these two factors. Given the rarity and aggressive nature of synovial sarcoma in the head and neck region, early and accurate diagnosis is critical for optimizing patient outcomes. Multimodal treatment approaches, including surgery and radiotherapy, remain the cornerstone of

management. Advances in molecular genetics may provide more targeted treatment options in the future, further improving survival rates and quality of life."

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