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 – 6, November – 2024 , Page No. : 120 – 125

Decoding The Enigma: A Case Study on Oropharyngeal Synovial Sarcoma

[1]Dr. Sai Vivek, M.B.B.S, M.D., Post Graduate Student, Resident, Department of Radiodiagnosis, Sree Balaji Medical College and Hospital, Chennai

[2]Dr. Piyoosh Priyadarshee, M.B.B.S, M.D., Post Graduate Student, Department of Radiodiagnosis, Resident, Sree Balaji Medical College and Hospital, Chennai

[3]Dr.Somya Nath, M.B.B.S, M.D., Post Graduate Student, Department of Radiodiagnosis, Resident, Sree Balaji Medical College and Hospital, Chennai

Corresponding Author: Dr. Sai Vivek, M.B.B.S, M.D., Post Graduate Student, Resident, Department of Radiodiagnosis, Sree Balaji Medical College and Hospital, Chennai

Citation this Article: Dr. Sai Vivek, Dr. Piyoosh Priyadarshee, Dr. Somya Nath, "Decoding The Enigma: A Case Study on Oropharyngeal Synovial Sarcoma", IJMSIR - November - 2024, Vol – 9, Issue - 6, P. No. 120 – 125.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Synovial sarcoma, a rare and aggressive subtype of sarcoma originating from mesenchymal cells, presents significant diagnostic and therapeutic challenges. This case report details an uncommon instance of synovial sarcoma located in the soft palate, a site with limited previous documentation. A 47-year-old male presented with a painless mass on the soft palate, initially suspected to be an abscess. This case underscores the necessity of considering malignancy in persistent or progressive soft tissue masses, even in atypical anatomical sites. Comprehensive diagnostic evaluation, including maxillofacial computed tomography (CT) and positron emission tomography-fluorodeoxyglucose (PET-FDG) scans, was pivotal in confirming the diagnosis and evaluating disease extent. The standard treatment involves complete tumor excision, yet establishing standardized margins for head and neck tumors remains challenging. Radiotherapy is crucial, especially for tumors larger than 5 cm or those with positive margins, although the role of chemotherapy is still debated despite its potential benefits. Effective management requires timely referral and a multidisciplinary approach to improve patient outcomes. While synovial sarcoma poses substantial diagnostic and treatment difficulties, advances in diagnostic technologies and personalized medicine hold promise for enhancing prognosis and treatment efficacy.

Keywords: Synovial sarcoma, head and neck, neck mass, oropharyngeal mass, malignant mass, surgical resection of synovial sarcoma, otolaryngology, oncology.

Introduction

Sarcomas, which originate from mesenchymal cells, are a rare and diverse group of malignant tumors, constituting less than 1% of all adult cancers [1,2]. The most prevalent subtypes include liposarcoma, leiomyosarcoma, undifferentiated pleomorphic sarcoma, and gastrointestinal stromal tumors [3]. The cell of origin for synovial sarcoma is still unknown, and it can be categorized into two morphological subtypes:

monophasic and biphasic [4]. While the etiology of most sarcomas remains unclear, several associated or predisposing factors have been identified. In addition to well-known genetic predisposition syndromes like Li-Fraumeni syndrome and neurofibromatosis type I, there is increasing recognition that a significant proportion of sarcoma patients may carry pathogenic germline variants. Other predisposing factors include retinoblastoma, exposure to radiation therapy, chemotherapy, chemical carcinogens, chronic irritation, lymphedema, and the involvement of human immunodeficiency virus (HIV) and human herpesvirus 8 in the pathogenesis of Kaposi sarcoma [5]. Lifestyle factors typically linked to cancer, such as smoking, diet, or exercise, are not considered risk factors for soft tissue sarcoma [6].

Sarcomas typically manifest as painless, slowly enlarging soft tissue tumors in young adults [7]. In uncommon instances, patients might display constitutional symptoms like fever or unexplained weight loss [8]. Specifically, for synovial sarcomas, the average duration of symptoms before achieving an accurate diagnosis is around two years. However, for sarcomas occurring in the head and neck region, the diagnosis is generally made about 20 months earlier, resulting in an approximate four-month interval between the onset of initial symptoms and diagnosis [9]. Sarcomas can arise in any anatomical location, but they predominantly affect the extremities, with only 9% of cases occurring in the head and neck area. Within the oral cavity, most reported instances involve the jaw bones and tongue [7]. According to Kadapa et al., only about ten cases were documented in the literature up to 2014 [7].

The differential diagnosis encompasses a range of conditions, including benign soft tissue tumors like lipoma and malignant tumors such as sarcoma, metastatic carcinoma, melanoma, or lymphoma. To aid in the timely identification of concerning soft tissue lesions, the United Kingdom Department of Health has outlined specific criteria for urgent referral. These criteria include the presence of a soft tissue mass exceeding 5 cm in diameter, a painful lump, a lump that is increasing in size, a lump of any size located deep to the muscle fascia, or the recurrence of a lump following previous excision [10]. These guidelines help ensure that potentially serious conditions are promptly evaluated and managed. When it comes to treating sarcomas, further research is needed to establish definitive protocols. The primary treatment involves the complete surgical removal of the tumor; however, specific margin guidelines for head and neck sarcomas have not been standardized. Radiotherapy can also be crucial, particularly in postoperative care for patients with positive or close margins or tumors exceeding 5 cm in size. Despite its importance, the optimal radiation dose has yet to be standardized [11]. Chemotherapy may offer a survival benefit, but its overall effectiveness continues to be a subject of debate and controversy [12]. This highlights the need for ongoing research to refine treatment strategies and improve patient outcomes. This clinical case, highlighting the exceptional rarity of oropharyngeal synovial sarcoma as evidenced by its limited presence in medical literature, underscores an essential point: the importance of raising awareness among clinicians regarding its distinctive clinical presentation and diagnostic complexities, as well as the critical need for early referral. The infrequency of this condition calls for increased clinical vigilance, prompt evaluation, and proper management. These measures are vital, as they can significantly influence patient outcomes and survival rates, underscoring the impact of timely and accurate medical intervention.

Case Discussion

In August 2021, a 47-year-old male visited his family doctor with a painless mass on the right side of his soft palate, which had been present for three weeks. He had no significant medical history, regular medications, allergies, or habits of smoking and drinking. He also did not have a fever or any other related symptoms. During the physical examination, the mass was observed to be about 1.5 cm in diameter, with intact mucosa, mild signs of inflammation, and a soft consistency upon palpation. The patient was afebrile. A week later, due to the absence of improvement, an increase in the size of the lesion, and new symptoms including intermittent shortness of breath and painful swallowing, he was referred to the emergency department (Figure 1).

Routine blood tests were normal. A maxillofacial CECT scan was done in which a well-defined hypodense lesion measuring 39 x 40 x 30 mm (AP x TV x CC) noted in the left palatine tonsil. On contrast, the lesion exhibited heterogenous enhancement. Clinical and laboratory findings suggested the possibility of a granuloma resembling an abscess, while the consideration of a tumor was based solely on direct observation. Reactive lymph nodes with typical bilateral central hilum were also present. No other notable pathological changes were detected (Figures 2-3). Following these observations, the patient was referred to the Otorhinolaryngology (ENT) department at the nearest hospital for further evaluation.

After the initial evaluation by the ENT department, a biopsy was planned. The biopsy was taken and a weeks later, pathology results confirmed a diagnosis of Synovial Sarcoma of the Biphasic Subtype (FNCLCC score 3). At this stage, the mass had expanded beyond the soft palate, appeared more concerning, and was covered with fibrin, taking on a vegetative look. Chemotherapy was started at this stage. The patient was prescribed tramadol, metoclopramide, and metronidazole, and further imaging with positron emission tomography-fluorodeoxyglucose (PET-FDG) scans was arranged. Two days later, the patient returned to the emergency department due to additional growth of the mass. Upon examination, a lesion was observed occupying the soft palate, covered with fibrin, and exhibiting a vegetative appearance, extending past the midline and reaching towards the tongue. It did not appear to involve the palatine tonsil.

A PET-FDG scan was performed, indicating the presence of an active malignant tumor. Bilateral superior lateral cervical lymph node formations, measuring less than a centimeter in short-axis and showing minimal uptake, raised concerns about potential lymph node involvement. Additionally, a CT-SPN scan report noted an increase in the size of the previously identified expansive lesion centered on the right/paramedian right side of the soft palate, which now extended to the uvula, appearing edematous and enlarged. The conclusion, based on the histological diagnosis of synovial sarcoma, was a staging of T3 N0. After this, the patient was posted for complete surgical resection of the mass along with sentinel lymph nodes. After the extensive resection and conservative cervical lymph node dissection, radiotherapy was given. Post treatment, all the symptoms were alleviated, and he was asked for follow-up visit every 6 months.

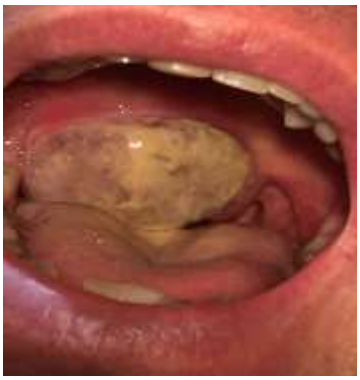

Figure 1: Oropharyngeal mass when the patient was sent to the emergency room

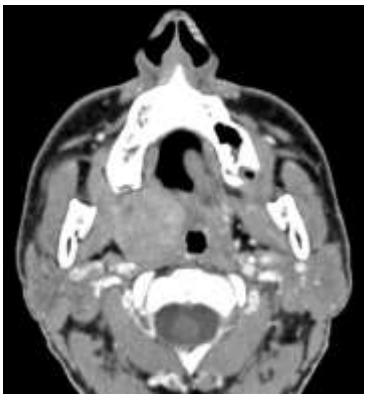

Figure 2: Plain CT Scan image, axial section showing a well-defined oropharyngeal mass

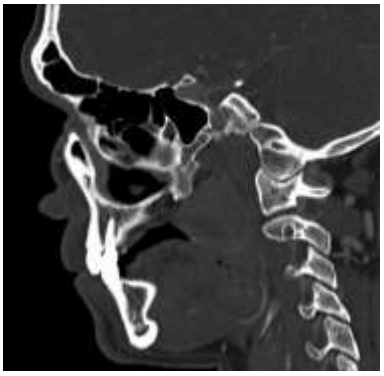

Figure 3: Plain CT Scan image, sagittal section showing the mass

Discussion

Synovial sarcoma is a rare and highly aggressive form of sarcoma that originates from mesenchymal cells [1,10]. Although it can develop in various parts of the body, its presence in the soft palate is exceptionally uncommon, with only a handful of cases documented in the literature [7,11].

The patient's initial symptoms included a painless mass on the posterior soft palate, which initially suggested a peritonsillar abscess due to the brief duration of symptoms, mild signs of inflammation, and the absence of systemic symptoms. While odynophagia is commonly linked with peritonsillar abscesses, its absence does not rule out the condition [13]. Literature indicates that a painless mass is the most frequent presentation of head and neck synovial sarcoma [2,7]. Despite this, the more common presentation of an abscess led to it being considered the primary diagnosis. However, the unusual presentation highlighted the need for early reassessment. During the follow-up, an increase in mass size was observed, along with no improvement and the emergence of new symptoms such as dyspnea and odynophagia. These observations necessitated an emergency room referral, which enabled the diagnosis to be made earlier than typically reported in the literature [9]. As demonstrated by Algargaz et al., delays in diagnosing sarcomas are common due to their rarity and variable clinical presentations [10].

This underscores the critical need to consider malignancy in the differential diagnosis of persistent or progressive soft tissue masses, even when they occur in atypical locations.

Regarding treatment, after histological confirmation of the diagnosis, the patient in this case received five cycles of chemotherapy, followed by surgery that included extensive resection and conservative cervical lymph node dissection. Given the presence of positive margins in the surgical histological report, adjuvant radiotherapy was recommended. The standard treatment for such cases generally involves complete tumor excision, with radiotherapy being crucial when positive or marginal margins are present [11]. However, since there are no universally accepted standards for margins or radiation doses, varied approaches have been employed. For instance, Kouhen et al. described cases where surgery was followed by radiotherapy with differing radiation doses, and chemotherapy was not administered to either patient [9]. This variation highlights the need for further research to establish standardized treatment protocols.

For family physicians, it is essential to maintain a high level of awareness of rare differential diagnoses to ensure timely referral to specialized care. This was particularly

relevant in the case presented, where the patient's medical history did not clearly indicate a risk for synovial sarcoma. Nonetheless, investigating potential underlying genetic factors or environmental exposures that might contribute to sarcoma development remains crucial.

The diagnostic process was greatly supported by advanced imaging techniques such as maxillofacial CT scans, MRI, and PET-FDG scans. These imaging modalities were essential in defining the characteristics of the lesion, assessing its extent, and identifying any potential metastatic spread [14]. Prompt referral was crucial for initiating timely treatment. Ongoing follow-up and monitoring are essential for detecting any recurrence of the disease and managing any possible late effects of the treatment.

Family doctors play a key role in providing patient-centered care, which involves thorough and integrated management of the patient's condition. Regular follow-up ensures timely reassessment and early detection of any changes, aligning with the comprehensive needs of the patient and ensuring consistent monitoring.

Raising awareness about the diverse presentations of synovial sarcoma, including its rare occurrence in sites like the soft palate, is crucial. This awareness significantly enhances patient outcomes by facilitating early detection and precise diagnosis—critical components of effective management. This case highlights the importance of increased awareness in reducing diagnostic delays, optimizing care, and improving the prognosis for synovial sarcoma.

By combining healthcare provider vigilance with patient-centered approaches and a focus on heightened awareness, the medical community can greatly impact the management and outcomes of complex cases like synovial sarcoma, demonstrating the potential for significant improvements in patient care.

Conclusion

This case underscores the critical importance of vigilance and awareness in the diagnosis and management of rare malignancies such as oropharyngeal synovial sarcoma. Despite its rarity, this condition can present in unexpected locations like the soft palate, necessitating a high degree of clinical suspicion and thorough diagnostic evaluation. Advanced imaging modalities, including maxillofacial CT and PET-FDG scans, are indispensable for accurate lesion characterization and assessment of disease extent, while timely referral and patient-centered care play crucial roles in optimizing treatment outcomes.

The evolution of this case from initial misdiagnosis to accurate identification and treatment highlights the necessity for ongoing education and awareness among healthcare providers. The variability in presentation and the potential for aggressive disease progression underscore the need for heightened clinical suspicion, especially in cases with atypical presentations.

Ongoing follow-up and surveillance are vital for managing disease recurrence and addressing any long-term treatment effects. By fostering an environment of heightened awareness and comprehensive care, healthcare professionals can significantly improve outcomes for patients with synovial sarcoma and other rare malignancies. This case exemplifies how early detection, precise diagnosis, and an integrated approach to patient care can lead to better prognoses and enhanced patient management.

References

1. Siegel RL, Miller KD, Wagle NS, Jemal A: Cancer statistics, 2023. CA Cancer J Clin. 2023, 73:17-48. 10.3322/caac.21763
2. Lockey MW: Rare tumors of the ear, nose and throat: synovial sarcoma of the head and neck. South Med J.

1976, 69:316-20. 10.1097/00007611-197603000-00021

3. Zagars GK, Ballo MT, Pisters PW, Pollock RE, Patel SR, Benjamin RS, Evans HL: Prognostic factors for patients with localized soft-tissue sarcoma treated with conservation surgery and radiation therapy: an analysis of 1225 patients. Cancer. 2003, 97:2530-43. 10.1002/cncr.11365
4. Sbaraglia M, Bellan E, Dei Tos AP: The 2020 WHO Classification of Soft Tissue Tumours: News and Perspectives. Pathologica. Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F (ed): IARC, 2021. 113:70-84. 10.32074/1591-951X-213
5. Ballinger ML, Goode DL, Ray-Coquard I, et al.: Monogenic and polygenic determinants of sarcoma risk: an international genetic study. Lancet Oncol. 2016, 17:1261. 10.1016/S1470-2045(16)30147-4
6. American Cancer Society medical and editorial content team. Risk Factors for Soft Tissue Sarcomas. Cancer.org. Last Revised: April 6, 2018. (2018). Accessed: 31/08/2023: https://www.cancer.org/cancer/types/soft-tissue-sarcoma/causes-risks-prevention/risk-factors.html.
7. Kadapa NP, Reddy LS, Swamy R, Kumuda, Reddy MV, Rao LM: Synovial sarcoma oropharynx - a case report and review of literature. Indian J Surg Oncol. 2014, 5:75-7. 10.1007/s13193-013-0285-0
8. Sinha S, Peach AH: Diagnosis and management of soft tissue sarcoma. BMJ. 2010, 341:c7170. 10.1136/bmj.c7170
9. Kouhen F, Afif M, Benhmidou N, et al.: Head and neck synovial sarcoma: a rare location: report of two cases. Pan Afr Med J. 2015, 20:232. 10.11604/pamj.2015.20.232.6225
10. Algargaz W, Abushukair HM, Barakat F, Mohamad I: Transcervical approach to oropharyngeal synovial sarcoma: a case report. Future Sci OA. 2022, 8:FSO815. 10.2144/fsoa-2021-0118
11. Lawrence W Jr, Donegan WL, Natarajan N, Mettlin C, Beart R, Winchester D: Adult soft tissue sarcomas. A pattern of care survey of the American College of Surgeons. Ann Surg. 1987, 205:349-59. 10.1097/00000658-198704000-00003
12. Fonseca AS, Azevedo AC, Magalhães FM, Andrade NA: Synovial sarcoma in head and neck: a case report. IntArch Otorhinolaryngol. 2014, 18:87-9. 10.1055/s-0033-1361081
13. Peritonsillar Cellulitis and Abscess. (2023). Accessed: September 5, 2023: https:// www.uptodate.com/contents/peritonsillar-cellulitis-and-abscess.
14. Christie-Large M, James SL, Tiessen L, Davies AM, Grimer RJ: Imaging strategy for detecting lung metastases at presentation in patients with soft tissue sarcomas. Eur J
15. Cancer. 2008, 44:1841-5. 10.1016/j.ejca.2008.06.004