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Low-grade Spindle Cell Sarcoma of the Tonsil: A Rare Entity

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Abstract

Background: Spindle cell sarcoma of the tonsil is an extremely rare malignancy in the head and neck region. The initial clinical presentation can be an asymptomatic mass lesion, and the biopsy of this growing mass should be conducted to exclude a malignant process. In this case report, we described a patient with spindle cell sarcoma of the tonsil with good clinical outcome after surgery alone.

Case presentation: A 38-year-old male presented with foreign body sensation of the throat and clinical examination revealed asymmetrical enlargement of the right tonsil. Tonsillectomy was performed, and histopathological examination and immunohistochemistry analysis revealed a low-grade spindle cell sarcoma of the tonsil. He developed local recurrence at twelve months and underwent wide local excision. The patient remained well without any local recurrences and metastases eight years after diagnosis.

Conclusion: We reported this case study in an effort to increase the awareness of spindle cell sarcoma of the tonsil and to emphasize the challenges in the diagnosis of this sarcoma. Surgical extirpation with a clear margin appeared to be an adequate treatment for the low-grade spindle cell sarcoma of the tonsil.

Keywords: Soft tissue sarcoma, Spindle cell sarcoma (SCS), Tonsil

1. Background

Primary spindle cell sarcoma is an exceptionally infrequent entity. Spindle cell lesions of the head and neck are heterogeneous in terms of clinicopathologic presentation and tumor biology (1-3). In adults, among the spindle cell malignancies in the head and neck, spindle cell carcinoma and melanoma are by far more common than sarcomas (4). Most malignancies of tonsils are epithelial, with the commonest being squamous cell carcinoma. The clinical presentation of spindle cell sarcoma can be similar to benign lesions at the early stage (5). Clinicians must be aware that intraoral sarcoma is very rare and early diagnosis is of significant importance. The objective of our study was to report this unusual neoplasm that poses diagnostic and management challenges to pathologists and clinicians due to its heterogeneity in clinicopathologic presentation. We described a case of spindle cell sarcoma of the tonsil with a better outcome after surgery alone.

2. Case Presentation

A 38-year-old male presented with intermittent foreign body sensation over the throat for 6 months. On examination, the patient had unilateral right tonsillar enlargement with a normal mucosal membrane. There was no palpable cervical lymphadenopathy. The patient underwent bilateral tonsillectomy, and intra-operatively, a mass measuring 2×1 cm was found at the lower pole of the right tonsil with no clinical invasion of adjacent structures. Histopathology examination demonstrated quite uniform spindle cells with mild atypical nuclei and amphophilic cytoplasm. Extensive immunohistochemistry analysis revealed that the tumor cells were diffusely positive for smooth muscle actin and vimentin but negative for other markers, such as CD21, CD31, CD34, CD35, EMA, D2-40, CKMNF 116, DESMIN, S-100 protein, h-Caldesmon, pan-keratin, and p63 [Figure.1]. A diagnosis of Spindle cell sarcoma was established. The clinicopathologic staging according to the AJCC-UICC system was stage IA, G1T1N0M0. 18 fluorodeoxyglucose (FDG) positron emission tomography-computed tomography (PET-CT) at a later date confirmed that there were no significant abnormal findings. Although the patient had been referred to the oncology department, he refused any oncology treatment. He was on regular follow-up and one year later, he developed swelling at the right side of the vallecula. Magnetic resonance imaging (MRI) demonstrated a well-defined nodular lesion at the right vallecula region that was hyperintense on T2W and hypointense on T1W [Figure 2]. It appeared that there was a clear separation between this lesion and the base of the tongue anteriorly and epiglottis posteriorly for which he underwent wide excision. The histopathological analysis was confirmed as recurrent spindle cell sarcoma. He had been referred to the Oncology Department; however, he refused

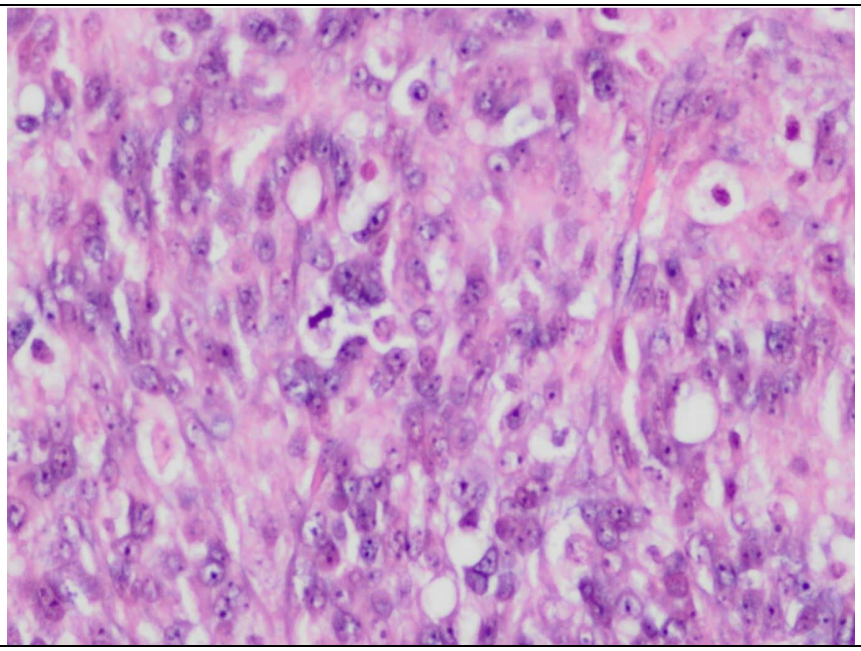

Figure 1. Histopathology revealed quite uniform spindle cells with mild atypical nuclei and amphophilic cytoplasm. The tumor cells are immunopositive for smooth muscle actin (diffuse) but negative for CD21, CD35, EMA, D2-40, Desmin, S-100 protein, pan-keratin, p63, and CD34. (Hematoxylin & Eosin stain x400)

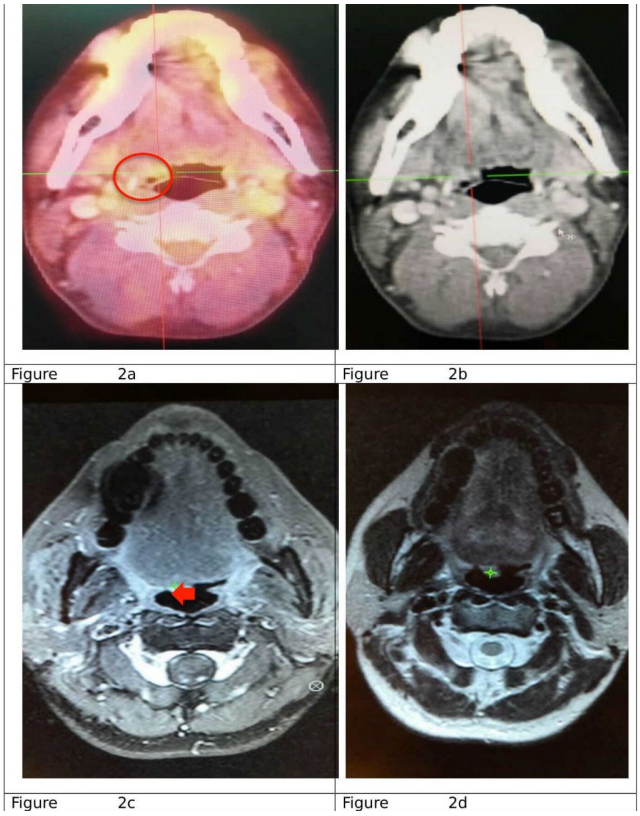

Figure 2a & 2b. Axial Fused 18F FDG PET-CT (left) & CT (right) images show an FDG-avid lesion (SUVmax: 3.23 ng/l, size: 1.0 cm). The round marker shows the involvement of the right side of the vallecula. The correlated contrasted CT (2b) shows that the lesion is hypodense with minimal enhancement.
Figure 2c & 2d. Axial contrasted T1 post-Gadolinium MRI and T1-weighted images show an enhanced soft tissue low-intensity lesion in the right side of the vallecula with an indistinct margin (arrow)

any further oncology intervention. The patient remained well without any more local recurrences or metastasis eight years after diagnosis.

3. Discussion

Spindle cell lesions of head and neck mucosal sites

are diverse in clinicopathologic presentation and diagnostically challenging. The most common spindle cell lesion affecting the upper aerodigestive tract is spindle cell carcinoma. Spindle cell carcinoma is a variant of squamous cell carcinoma that simulates a true sarcoma but is epithelial in nature (**2**).

Primary spindle cell sarcoma is an extremely unusual malignancy and one of the least reported entities. Primary undifferentiated sarcomas are rare mesenchymal tumors that more commonly affect the musculoskeletal system. There are only very few cases of primary spindle cell sarcoma arising from various body parts that have been reported around the world (1). The etiology of tonsillar sarcoma is unknown. The clinical behavior of tonsil sarcoma is poorly understood due to the rarity of the cases.

Unilateral tonsillar enlargement is considered to be a sign of malignancy (6). The most prevalent malignancies of the tonsils are squamous cell carcinoma and non-Hodgkin lymphoma (7). Spindle cell sarcoma should be included in the differential diagnosis of tonsillar masses because interpretation as undifferentiated carcinoma or lymphoma may lead to different lines of treatment.

Feng et al. in their SEER population-based analysis of spindle cell sarcoma, reported that the mean age of diagnosis of patients was 61 years and the incidence peaked during the seventh decade of life (1). In contrast to this finding, our patient was much younger compared to the study population. Their study showed no gender predilection (1).

There are no specific criteria for sarcoma in the diagnostic imaging methods of computed tomography CT or MRI. Computed tomography and magnetic resonance imaging are useful in determining tumor extent in planning the surgical approach (8). PET-CT imaging can be useful in the detection of primary lesions and the metastases of soft tissue sarcoma due to its metabolic activity (5).

Accurate diagnosis of soft tissue sarcoma is reached by histopathology and immunohistochemical analysis (9). Fine Needle Aspiration Cytology should not be used as a replacement for an excision biopsy. Immunocytochemistry on aspirates may not help to distinguish a low-grade sarcoma from a benign spindle cell lesion (10). The histological diagnosis of soft tissue sarcomas is difficult due to its rarity, complexity, and heterogeneity.

Spindle cell sarcomas pose a challenge to the treating clinician due to inconsistent management outcomes (9). The diversity of clinical behavior, the paucity of patients, variable metastatic potential, and the rarity of cases makes it difficult to develop standardized treatment protocol (8). Surgery is the most reliable treatment for sarcomas of the oral and maxillofacial region. Adequate excision with a safe surgical margin as the initial therapy is important for better survival (11). Surgery alone appears to be an adequate treatment for small, low-grade soft-tissue sarcomas with negative surgical margins. Patients with high-grade tumors or with incomplete resection should receive aggressive treatment, which includes surgery and radiotherapy with or without chemotherapy (12).

Distant metastasis is very rare and only occurs in high-grade primary lesions (9). En bloc resection in the head and neck region is difficult because of closely adjacent neurovascular structures and the inherent functional and cosmetic deformities associated with the sacrifice of these structures (8). This may be the reason why there is a higher local recurrence rate and worse disease-specific survival in head and neck sarcomas compared to other sites (13).

4. Conclusion

We presented a unique case of spindle cell sarcoma of the tonsil. Although Spindle cell sarcoma of the oropharynx is a rare tumor, we should keep in mind the possibility of such presentation which poses challenges to pathologists as well as to clinicians. Surgical extirpation with a clear margin is believed to be the most appropriate treatment, the suggestion that was confirmed in our case.

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Footnotes

Author's contributions: Study concept, design, drafting the article and revising the manuscript: Sethu Thakachy Subha. Study design, Data collection and final approval of the manuscript -Mohamad Doi . Data analysis and interpretation and final approval of the manuscript :Fathinul Fikri Ahmad Saad .Data analysis and interpretation and final approval of the manuscript: Nor Yatizah Mohd Yatim
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