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A Narrative Review of Myxoid Solitary Fibrous Tumor: A Rare Benign Maxillofacial Tumor

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ABSTRACT

Solitary fibrous tumor (SFT) is a relatively rare mesenchymal neoplasm characterized by spindle cells exhibiting fibroblastic differentiation. The neoplasm was initially identified in the pleura but had later been recognized in various human anatomical areas. In addition, it typically features disorganized spindle cells, varying cell density, collagen-rich stroma, and slender branching blood vessels. Several studies have shown that myxoid SFT with a significant myxoid stroma is exceedingly rare, with less than 15 cases reported. Approximately 20% of SFT often occurs in the head and neck, including the meninges. The extracranial head and neck area have been reported to be the most common areas, particularly the sinonasal tract, oral cavity, and deep soft tissues, such as the orbit. Therefore, this narrative review aims to describe the distinctive features of SFT to assist clinicians in identifying their rarity.

Keywords: Solitary Fibrous Tumor, Head and Neck Neoplasm, Immunohistochemistry, Soft Tissue Neoplasm

ABSTRAK

Tumor fibrosa soliter adalah neoplasma mesenkim yang jarang ditandai dengan sel gelendong yang menunjukkan diferensasi fibrotik. Awalnya diidentifikasikan di pleura, tumor ini telah dikenali di berbagai lokasi anatomi manusia. Secara histologis, ia menampilkan sel gelendong yang tidak tersusun, kepadatan sel yang bervariasi, stroma yang kaya kolagen, dan pembuluh darah bercabang yang ramping. Tumor fibrosa soliter (TFS) miksoid dengan stroma miksoid yang signifikan sangat jarang terjadi, dengan kurang 15 kasus yang dilaporkan dalam literatur. Sekitar 20% TSF terjadi di daerah kepala dan leher, termasuk meningen, dengan daerah kepada dan leher ekstrakranial menjadi lokasi paling umum, khususnya saluran sinonasal, rongga mulut dan jaringan lunak dalam seperti orbita. Tinjauan literatur ini menjelaskan ciri khas SFT untuk membantu dokter gigi dalam mengidentifikasikannya di sebalik kelangkaannya.

Kata kunci: Tumor Fibrosa Soliter, Neoplasma Kepala dan Leher, Immunohistokimia, Neoplasma Tisu Lembut

1. Introduction

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm composed of spindle cells with fibroblastic differentiation. Initially, it was recognized as a lesion in the pleura but has now been accepted as a neoplasm that can develop in several anatomical areas. The histological traits of SFT include disorganized spindle cells, areas of differing cell density, a stroma abundant in collagen, and thin branched blood vessels [1]. Although localized myxoid alterations are not rare in SFT, cases featuring significant myxoid stroma are uncommon. Several reports have shown that less than 15 cases of myxoid SFT have been recorded in the medical literature [2–6] .

Approximately 20% of SFTs are found in the head and neck area, including the meninges. In the extracranial head and neck area, it typically arises in the sinonasal tract, oral cavity, and deep soft tissues, such as the orbit. In addition, it is highly rare for SFT to originate primarily in superficial soft tissues, such as the dermis [7–11]. SFT that develops in the extracranial head represents a total of 10% of cases and typically emerges initially as a small, symptomatic tumor, with the sinonasal tract and orbit being the most frequently affected areas. In orbit, the clinical presentation features a growing mass in the eyelid, orbit, epiphora, or proptosis [11].

SFT can arise in the oral cavity beneath the buccal mucosa, on the tongue, and in the lower lip. Tumors that develop in the deep soft tissues of the cheek or neck usually appear as a painless mass. While the majority display benign characteristics, there have been instances of malignant behavior. Local recurrence often occurs in 40% of cases when tumors arise in difficult-to-reach areas of the orbit or sinonasal tract, where complete, en-bloc resection is not feasible [12].

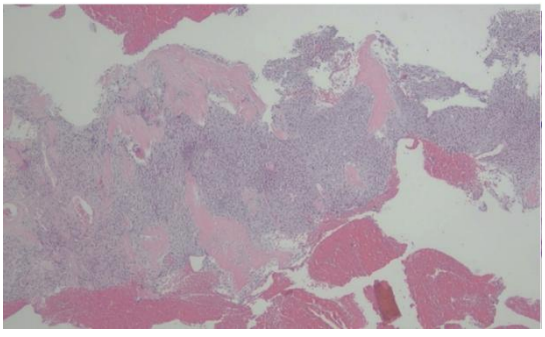
2. Subtopics

2.1 Clinical Presentation

SFT appears well-delineated, which is a slow-growing, painless mass that could be asymptomatic or cause variable compression symptoms depending on the size and anatomical areas [13].

2.2 Histological Features

After gross examination in this study, the mass appeared encapsulated. The sliced surface was yellowish white, with a soft myxoid and rubbery fibrous area. Microscopically, the lesion comprised elongated spindle cells dispersed in the heterogeneous stromal matrix with myxoid and collagenous areas. Mitotic figures, cellular pleomorphism, or nuclear aplasia were often absent. The immunohistochemical analysis showed intense positivity to CD34 but no immunoreactivity with staining and actin, desmin, S-100, c-kit protein, or neurofilament (6). Figure 1 illustrates the prominent features of tumors in this study.



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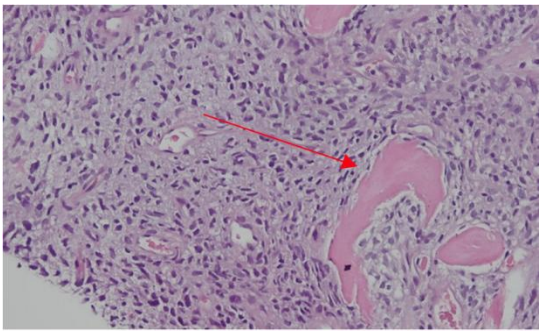

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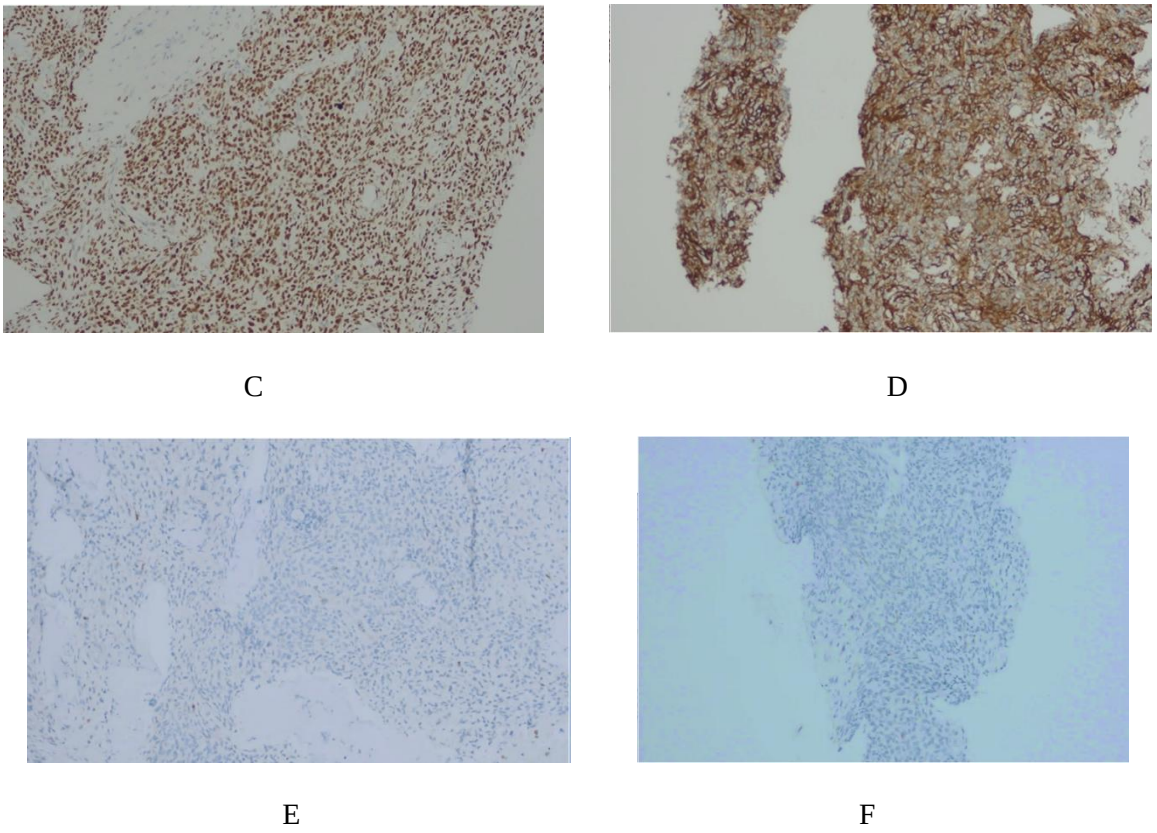

Figure 1. Histopathological results (A) Hematoxylin and eosin (H&E) staining at low magnification (·4) demonstrated lesional tissue composed of fusiform cells in an irregular pattern embedded in myxoid stroma. (B) At higher magnification (×20), spindle-shaped cells embedded in a prominent myxoid stroma with staghorn-pattern blood vessels were observed, consistent with the characteristic histological architecture of SFT. (C) Immunohistochemistry showed diffuse nuclear positivity for STAT 6), (D) revealed intense positivity to CD 34, and tumor cells were negative for S100 (E, ×10) and CD 117 (F, ×10).

These results confirmed the diagnosis of SFT, consistent with its immunophenotypic profile. The presence of STAT6 positivity was a highly specific marker for SFT, and the lack of S100 and CD117 expression further assisted in differentiating SFT from histologically similar spindle cell neoplasms. Image courtesy of the Histopathology Unit, Department of Pathology, Hospital Tengku Ampuan Rahimah (HTAR), Klang,

2.3 Radiological Features

SFT, including myxoid variants, frequently displayed diverse radiologic features based on their areas and biological activity. Therefore, acquiring adequate radiological images was essential for diagnosis and treatment as several modalities offered valuable insights.

Studies shown that SFT is diverse and typically nonspecific in radiological imaging. Colour ultrasonography reveals a hypoechoic mass with strong blood flow signals, indicating a large blood supply due to their dense vascular network. While on computed tomography (CT) scan, SFT appears as an oval, well-defined mass isodense to the skeletal musculature with prominent avid blood vessels. In instances where masses were larger or malignant, a more heterogeneous appearance due to fibrosis, hemorrhage, necrosis, myxoid and cystic degeneration, or calcifications was presented [14]. However, on Magnetic Resonance Imaging (MRI), T1-weighted images appears as isodense to grey matter, and T2-weighted images exhibited a heterogenous isodense or hypodense quality [15].

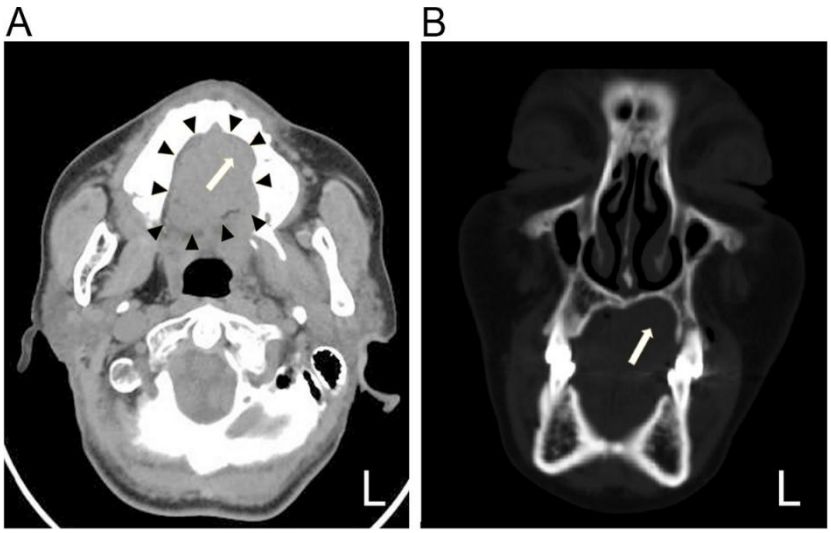

Figure 2. The CT images exhibited a mass (with black arrows) locating on the left palate with bone resorption (white arrow). **A**: Transverse plane, **B.** Coronal plane, L: left Image reused from Wang C, Wang B, He J. Solitary fibrous tumors of the oral and maxillofacial region: a case series from a single-center. BMC Oral Health. 2024 Nov 27; 24(1):1444. Available from https://bmcoralhealth.biomedcentral.com/articles/10.1186/s12903-024-05241-2

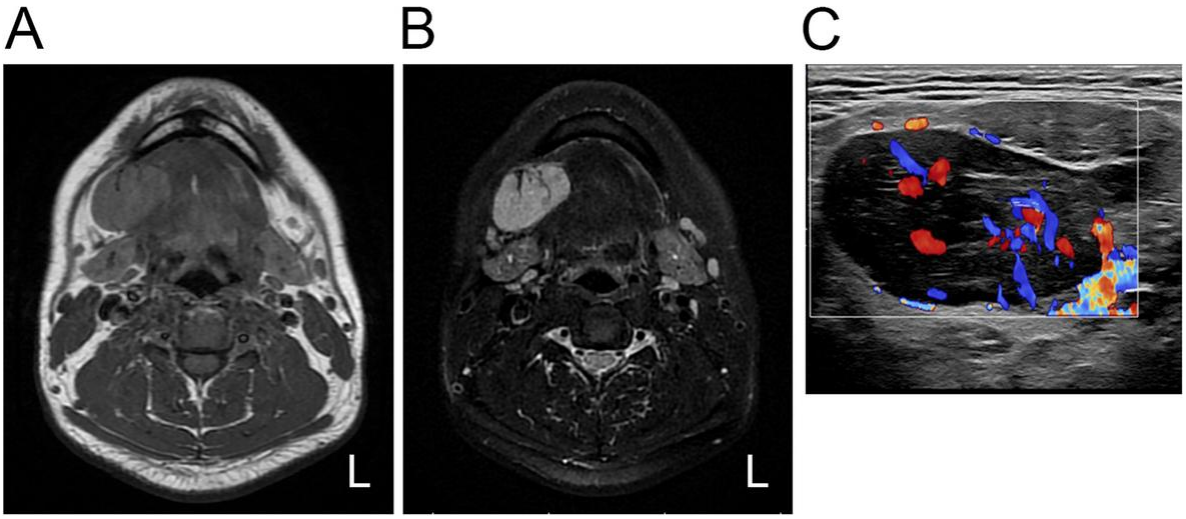

Figure 3. The MRI results reveals an elliptical abnormal signal located in the right submandibular region, with isointensity on T1WI **(A)** and hyperintensity on T2WI **(B). C** Color ultrasonography reveals abundant blood flow signals in the mass L: Left. Image reused from Wang C, Wang B, He J. Solitary fibrous tumors of the oral and maxillofacial region: a case series from a single-center. BMC Oral Health. 2024 Nov 27; 24(1):1444. Available from https://bmcoralhealth.biomedcentral.com/articles/10.1186/s12903-024-05241-2

2.4 Current Treatment

The primary treatment for maxillofacial SFTs is surgical excision. The goal is to complete an en bloc resection with negative margins. The transoral technique is excellent for improved outcomes and significantly faster recovery, but it only provides limited exposure ([16].

A typical method begins with clinical palpation to precisely localize the mass, which can then be further marked using a surgical marker to aid with intraoperative localization. Following localization, an incision is typically planned in the centre of the lesion in order to allow direct access. To reduce intraoperative bleeding, a vasoconstrictor is given before the incision. Dissection is performed in a blunt manner, allowing for meticulous advancement towards the tumor margin while preserving the surrounding tissues. When the tumour is well encapsulated but friable, as is common in many soft tissue neoplasms, removal can be hindered by tissue fragility

and a proclivity to rupture. In such cases, a sequential dissection technique is suggested to guarantee full excision while preserving the lesion's integrity to the greatest extent possible [17].

There is a reported study of a rare instance of SFT in the buccal area in which a transoral technique was used for tumour resection, including coronoidectomy to increase visibility and give enough working space for the transoral excision. It is a feasible surgical method for accessing and eliminating tumours in challenging anatomical locations within head and neck region [18].

Surgical treatment with adequate margins and extended follow-ups remained the standard of care in managing patients with SFT. This was with a recurrence-free survival surpassing 90% for completely resected cases and overall recurrence or metastatic rates ranging from 5% to 10%, according to early studies. In certain situations, neoadjuvant radiotherapy aiming at tumor shrinkage was used to control local symptoms or facilitate surgical excision [13].

2.5 Prognosis

According to Wushou et al,the prognosis of SFT can be affected by a variety of demographic, clinical and pathological characteristics, as evidenced in a comprehensive population-based analysis that included 804 cases from the SEER database, which indicates a gradual decline in long-term survival among SFT patients. Table 1 illustrates the survival rates for SFT.

On the other hand, increasing age was associated with poorer outcomes, with patients over 51 years exhibiting a higher hazard of recurrence and mortality. Similarly, tumours with distant metastases at diagnosis, as defined by SEER staging, were strongly predictive of unfavourable prognosis. High histological grade (Grade III/ IV) also emerged as a critical determinant of prognosis, significantly increasing the risk of both recurrence and death.

Notably, the type of treatment received had a significant impact on patient outcomes. When compared to combination surgery and radiation, surgical resection alone was independently linked with superior DFS; however, this benefit was not statistically significant in terms of OS. Therefore, the prognosis of SFT is complex, with age, metastatic status, tumour grade and treatment modality all playing pivotal roles. These prognostic indicators should be addressed in clinical decision-making to optimise long-term outcomes for patients with SFT. Table 2 illustrates the prognostic factors for DFS and OS in SFT [19].

Table 1. Survival Rates for SFT (DFS and OS)

Time point	Disease-freesurvival (DFS) (%)	Overall survival (OS) (%)
3-year	73.3	71.9
5-year	65.7	63.3
10- year	53.3	47.3

Table 2. Prognostic Factors for Disease-Free Survival (DFS) and Overall Survival (OS) in Solitary Fibrous Tumour (SFT)
Multivariate Cox Regression Analysis from SEER Database Study (Wushou et al 2015)

	Prognosic Factor	Hazard Ratio (DFS)	P-value (DFS)	Hazard Ratio (OS)	P-value (OS)
1	Age > 51 years	1.851	0.024	1.652	0.033
2	SEER Stage : Metastatized tumor	4.269	0.000	2.905	0.028
3	Pathologic Grade III/IV	2.734	0.001	2.585	0.000
4	Surgery Alone (vs surgery + Radiotherapy)	0.217	0.045	0.811	0.476

Note: Hazard Ratios (HR) greater than 1 indicate worse prognosis; values less than 1 suggest a protective effect. Only surgery alone showed a statistically significant improvement in DFS, but not in OS.

3. Discussion

Every SFT had some degree of myxoid change, but SFT exhibiting predominantly myxoid stroma was rarely encountered [20]. A slow-growing lesion with a well-defined growth pattern did not adhere to surrounding tissues and the absence of cranial nerve involvement or lymphadenopathy often suggested a benign condition. In addition, the lack of erythema, pain or warmth, and normal white blood cell count implied that the process was neither infectious nor inflammatory. Other common differential diagnoses for maxillofacial tumors include pleomorphic adenoma, subcutaneous lipoma, Warthin's tumor, monomorphic adenoma, and malignant tumors of the salivary glands [21]. Other alternative diagnoses included vascular anomalies, such as lymphangioma, and vascular neoplasms such as haemangioma, angioleiomyoma, and juvenile nasal angiofibroma [22].

Cross-sectional imaging revealed distinctive radiographic findings, though not pathognomonic. Tumors showed enhancement following contrast administration, but this was either homogenous or heterogeneous. On MRI, SFT typically displayed low T1 signal intensity and differing T2 signal. Meanwhile, on CT, extracranial SFT of the head and neck appeared as solitary well-circumscribed masses that could be isointense to muscle in studies without contrast. The most frequent radiographic osseous result was the regressive remodeling of adjacent bone due to the long-standing pressure effects on the slow-growing mass [23]. Radiographic evidence of bone destruction could be observed, but this did not necessarily correlate with histologic characteristics of malignancy [24].

Histological examination played a crucial role in diagnosing SFT in this study. A sufficient tissue sample was assessed for typical morphologic features alongside a distinctive immunophenotype. In addition, a strong nuclear expression of STAT6 was a highly sensitive and specific immunohistochemical marker for SFT [25].

Finally, long-term follow-up was essential for patients with SFT because the clinical behavior did not match the histopathological appearance at all times. A small but statistically significant increased risk of local recurrence in extra-thoracic SFT existed [26].

The literature on maxillofacial SFT was scarce, with several key gaps noted. Initially, information regarding the clinical behavior, prognosis, and recurrence rates of SFT in the maxillofacial area was quite limited when compared to other anatomical areas. Moreover, there is a lack of comprehensive multicenter studies and randomized controlled trials evaluating treatment protocols, particularly regarding long-term outcomes following surgical resection. The available studies primarily consisted of case reports or small case series, which restricted the capacity to generalize conclusions or create treatment protocols. In addition, studies on the effectiveness of adjuvant treatments, such as radiation or chemotherapy in preventing recurrence or metastasis in the framework of maxillofacial SFT were insufficient.

4. Declaration of Competing Interest

The authors declare no conflicts of interest.

5. Acknowledgments

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