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Spindle Cell Sarcoma in the Oral Cavity as a Gingival Mass: A Case Report with Literature Review

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Case Report

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Abstract

Spindle cell sarcoma is an extremely rare tumor, with only a few cases reported worldwide. Although it has been reported in different parts of the body, occurrences in the oral cavity are exceedingly uncommon. Here, we report a 34-year-old Iranian woman who was referred with the chief complaint of bleeding from a mass inside her mouth. On physical examination, a firm, exophytic, and pediculated mass measuring 2×2 cm with a smooth surface was observed on the lingual side of the right mandibular ridge. The mass was removed and sent to the laboratory, which revealed spindle cell carcinoma with a high proliferative index and focal muscle differentiation. Cone beam computed tomography and radiography were requested for the patient. She underwent mandibulectomy and mandibular reconstruction using a free flap, taken from the patients' fibula, and was discharged in good condition. Reporting this case can enrich the literature and raise awareness among physicians regarding this rare disease.

Key Clinical Message

Spindle cell sarcoma may present as a benign-appearing gingival mass in the oral cavity. Rapid recurrence after excision should prompt immediate histopathologic reassessment and definitive surgical management. Early recognition and wide resection are essential to prevent progression and improve outcome in this rare but aggressive malignancy.

Introduction

Spindle cell sarcoma (SCS) is a rare neoplasm and one of the least frequently reported tumors. According to the largest study based on the Surveillance, Epidemiology, and End Results (SEER) database (1973–2017), 3299 cases have been documented; about 80% were white with a median age of 57 years (ranging from birth to 103 years) and no sex predominance (1). Considering the limited number of published case reports, both diagnosis and treatment of SCS are still a challenge, and patient survival depends on factors such as age, TNM stage, tumor site, surgical intervention, and pathological grade (1).

As a connective tissue tumor, SCS mainly originates within the layers of connective tissue. However, it can be observed not only in the soft tissue but also in the bone or visceral organs, including synovia, nerve sheets, leiomyoma (2), the abdomen (3), and even in the heart (4). The oral cavity is a rare site for SCS, with only a few cases reported in this region (5) and the maxillofacial area (6). Also, less than 1% (0.7%) of non-odontogenic oral spindle cell neoplasms are classified as soft tissue SCS (7).

Considering the scarcity of this type of tumor and its unusual manifestation in the oral cavity, here, we report a SCS of the oral cavity in a young woman, describing her symptoms, pathological findings, imaging results, treatment approach, and the patient's outcome.

Case presentation

A 34-year-old Iranian woman was referred to the Oral Medicine Department of Shiraz Dental School, Shiraz, Iran, with the chief complaint of bleeding from the mass inside her mouth, which had been enlarging over the past week. On physical examination, the physician observed an exophytic mass on the right mandibular gingiva adjacent to tooth #6 (first molar). The mass had a smooth surface, was pedunculated, bluish in color, and exhibited a firm consistency. It measured approximately 2×2 cm on the lingual side of the gingiva. As it was a soft tissue pedunculated mass in the oral cavity, we considered reactive lesions, including pyogenic granuloma and peripheral giant cell granuloma (as more common causes) for primary differential diagnoses. On second thought, we considered other mesenchymal tumors (considering the soft surface). A definitive diagnosis required pathological examination; therefore, a biopsy sample was taken.

An excisional biopsy was performed, during which the mass was completely removed and sent to the laboratory for pathological examination. During the surgery, bleeding was controlled using cautery. The pathological report showed a creamy-brown, dome-shaped elastic mass covered by ulcerated mucosa. In microscopic examination, the spindle cell carcinoma was detected (Fig. 1), characterized by a high proliferative index and focal muscle differentiation, based on immunohistochemical staining (IHC) results (Pathology No. DP-140-03). The tumor was negative for CD34, p63, CD56, and myogenin; exhibited high Ki-67 activity, and was positive for SMA and Desmin. At 1-month follow-up after the IHC analysis, the lesion had quick recurrence with a similar size. The results of the physical examination before and after the excision are shown in Fig. 2 (A: before excision; B: 1 month post-excision).

Accordingly, cone beam computed tomography (CBCT) was requested for the patient. The results of which (Fig. 3) revealed discontinuity of the lingual cortex and destruction of the crestal bone, as well as decreased alveolar height extending from the distal side of the right mandibular canine to the mesial aspect of the ipsilateral second molar beneath the soft tissue lesion on the right side of the mandible. An ill-defined radiolucent lesion was detected at the mesial of tooth #7 to the distal of tooth #7, involving the furcation area of the first molar tooth. Non-centric band-like periodontal ligament (PDL) widening was seen along both roots of tooth #6, indicating involvement of the tooth by the lesion (lack of PDL widening, alveolar crest rejected periodontal disease). Concentric widening of PDL was seen around the root apices of tooth #7, compatible with an inflammatory response. No periosteal reaction was detected over the cortical surfaces of the mandible. Teeth #4, #5, and #6 showed grade I mobility. Additionally, rarefying osteitis with surrounding sclerosis was evident in the periapical region of tooth #6.

The patient was referred to an oncologist, who requested plain radiography of the body, including the neck, chest, and abdominopelvic regions. A venous blood sample was taken from the patient and a complete blood count (CBC) was measured, revealing reduced reticulocyte count (3.91×10^{6}/μL), hemoglobin level (10.1 g/dL), and hematocrit (31.6%). Other CBC parameters, along with blood urea nitrogen, creatinine, and liver enzyme levels, were within normal range.

Subsequently, the patient was referred to a fellowship of surgery and underwent mandibulectomy and mandibular reconstruction using a free flap, taken from the patients' fibula (Fig. 4), connecting the vessels as well. The patient was hospitalized for two weeks and was discharged in good condition. At final follow-up 6 months after surgery, she had no complications. Since the lesion was completely excised and the margins were free, there was no need for follow-up with radiotherapy, and the patient has resumed her normal life, including returning to work.

As shown in Table 1, we found only three other cases reporting SCS in the oral cavity, apart from our own report, two of which were men and one woman. Overall, SCS typically presents with a mass in any part of the body, generally progressive (10). Considering SCS in the oral cavity, all cases reported a progressive nature of the tumor, the duration of disease and the extent of progression were different. In case #1, the patient had a small tumor on his tongue for 2 years (9), while in case #2, the mass enlarged excessively within 2 months, not only with intra-oral mass, involving the teeth (26, 27, 28), but also with facial swelling, obliterating the buccal vestibule and perforating the left maxillary buccal cortical plate (5). Interestingly, in case #3, a large maxillary lesion progressively enlarged during 2 years, making it difficult for the patient to speak and swallow, and leading to anemia and weight loss (6). In our case, we had excised the lesion completely and sent it for pathological examination. However, when the patient returned 1 month later, the lesion had regrown to its original size and was excised again. The rapid regrowth within just 1 month showed the progressive nature of the mass.

Table 1
A summary of the cases presented with spindle cell sarcoma of the oral cavity in the literature

1	Reference number	Age	Gender	Duration of mass	Mass site	Mass size	Medical history	Physical examination	Histologic view	IHC	Treatment
	(9)	**54**	**M**	**2 years**	**tongue**	**8*7mm**	**None**	**Hard well-circumscribed mass without lymphadenopathy**	**High-grade, undifferentiated spindle-cell sarcoma.**	**Not mentioned**	**Surgery (hemiglossectom + adjuvant chemotherapy**
2	(5)	48	M	1 month	Left side of face	4*5 cm	Type II diabetes, smoking	Non-reducible well-defined immobile swelling	Spindle to round cells with pleomorphism, mitotic stage IVA	vimentin	Chemotherapy
3	(6)	42	F	2 years	Right maxilla			Firm exophytic mass with ulceration involving molars	Cellular atypia and mitotic, stage IVA	s-100, CD34, ki67(5–10%),	Wide surgical resection

Differential diagnosis, investigations and treatment

Diagnosis

Similar to other studies, the final diagnosis in our case was established based on the results of the pathological report, whereas imaging revealed the extent of the mass. In our case, we performed an excisional biopsy and sent it for pathological examination, while all of the other cases were performed with an incisional biopsy (5, 6, 9). In our study, IHC detected a high positivity of ki-67, indicating high proliferation, high-grade characteristics, and the presence of muscle differentiation markers. In case #3, ki-67 was positive in 5–10%, which was similar to the present study, while other IHC markers differed. Nonetheless, these findings also point to the nature of SCS as a connective tissue neoplasm (Table 1). Two of the cases reported the tumor stage as IVA, while lymphadenopathy and lymph involvement were not reported in any of the cases.

Treatment

According to the SEER report, more than 60% of all SCS cases were treated with surgery alone (1), similar to our patient. Given the aggressive nature of this type of tumor, the surgeon decided to perform mandibulectomy with reconstruction surgery. In contrast, chemotherapy was recommended by the physicians for the other reported cases, though some patients accepted the treatment while others declined. In case #3, the patient underwent maxillectomy with obturator placement as reconstruction surgery, while she refused adjuvant therapy (6). Also, in case #1, treatment involved surgery with hemiglossectomy along with adjuvant chemotherapy (the details were not provided) (9). Neoadjuvant chemotherapy was also started for the other patient (case #2), followed by left maxillectomy, lymph node dissection, radiotherapy, and prosthetic reconstruction; however, the treatment was discontinued after a few days because of the patient's death (5). Accordingly, we stand by the point that surgery is the most important treatment for patients with oral SCS; non-surgical treatments have been reported to be associated with worse prognosis in the overall SCS cases (1), and we speculate that this may also apply to tumors in the oral and maxillary regions. It is important to consider that this cancer has a progressive nature, and the treatment should be applied as soon as possible. Patients must be thoroughly informed about the potential risks associated with delayed intervention. Awareness of general and triage physicians about this condition is also necessary for the on-time referral of such patients.

Outcome (follow-up)

The patient presented here could fortunately continue normal life after two excisions, with no signs of recurrence. Other cases also reported no recurrence after about a year (6, 9). Unfortunately, the tumor had progressed extensively in case #2, which caused the patient's death before the treatments could influence the tumor. This underlines the critical importance of timely and appropriate diagnosis and treatment. As mentioned in the SEER report, the mortality

rate is not negligible, with 2,115 deaths among 3,229 recorded SCS cases (1); therefore, it is important to pay greater attention to this tumor and treat it before disease progression becomes irreversible.

Some studies have addressed the genetic mutations observed in SCS, like NUTM1 rearrangements (NUTM1:MGA fusion), in SCS of the lung (11). However, we did not perform genetic analysis for this case, due to the high costs of genetic testing in our country. Future studies on the genetic profile may be able to diagnose patients before the clinical presentations occur, so that the patient and the physician become aware of the susceptibility of that patient to this type of aggressive tumor and diagnose it at early stages, when it has a better prognosis. A strength of this case report is the detailed clinical, histopathologic, immunohistochemical, radiographic, surgical, and follow-up documentation of a very rare oral presentation of spindle cell sarcoma. The main limitation is the absence of molecular or genetic testing, primarily because of cost-related limitations.

Conclusion

SCS is a very rare but aggressive tumor that can present at any site of the body, rarely in the oral cavity. According to our review, the case we presented here was the fourth case reported in the literature with oral (and maxillary) SCS. Fortunately, our patient underwent timely and extensive surgical treatment just 1 month after diagnosis and remains in good condition to date. Considering the scarcity of this phenomenon, especially in this specific site, presenting this case can help physicians become aware of this type of tumor. This case points to the significance of excising oral exophytic lesions with pathological examination, keeping in mind that a malignant lesion could begin with a clinical appearance similar to an irrigational lesion. In such cases, timely diagnosis and treatment are the keys to extending the patient's life expectancy and improving their quality of life.

Discussion

SCS is an exceptionally rare malignancy, with only 3299 cases reported worldwide since 2018 (1). As reported by the SEER study, the mean age of diagnosis is 61 years, with incidence peaking in the seventh decade of life, mainly in white people (about 81%), followed by 11.4% in Black, and < 8% in Indian, Asian, and Pacific Islander races, with no sex preference reported. According to this dataset, primary SCS can be found at almost any site of the body, mainly in superficial sites (the head and neck, limbs, trunk, abdomen, and pelvis) rather than internal organs, such as the digestive, respiratory, reproductive, locomotor, urinary, nervous, endocrine, and circulatory systems, or the peritoneum and spleen (1). The oral cavity, categorized either in the digestive system or in the head and neck, is a very rare site for SCS, reported in only one case in the SEER dataset, accounting for less than 1% of tumors reported in the oral and maxillofacial region (1). A review of spindle cell neoplasms in the oral cavity, traced to epithelium, mesenchyme, and odontogenic components, also indicates SCS as a very rare tumor (8). Additionally, another review of 307 cases with spindle cell neoplasm reported none in the oral cavity (7). Therefore, reporting this case can attract the physician's attention to considering this and other types of rare tumors when they examine an exophytic lesion in the oral cavity, as well. As the physical examination revealed a mass in the oral cavity with a soft appearance, we did not speculate on SCS in the first place, because it is a very rare tumor in this site. Instead, more prevalent diagnoses like pyogenic granuloma or peripheral giant cell granuloma were considered, along with other mesenchymal tumors. Ultimately, the results of pathological examination revealed the definite diagnosis and helped us to treat the patient appropriately.

There are a few cases available in the literature reporting SCS in the oral cavity. For a better comparison of these cases, the details of each case are summarized in Table 1. In the following, we discuss the different dimensions of our case in comparison with similar cases.

Declarations

Ethics approval and consent to participate

Not Applicable

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Data Availability Statement

The supporting the findings of this study are available from the corresponding author upon reasonable request.

Conflicts of Interest

None

Funding

None

Author Contribution:

Sara Maroufi: Conceptualization; Data curation; Writing - original draft.

Amirmohammad Roldar: Investigation; Data curation.

Fahimeh Rezazadeh: Supervision; Validation; Writing - review and editing.

Fatemeh Daryaei: Investigation; Resources.

Seyede Asiyeh Sadrpoor: Pathological assessment and interpretation; Methodology; Visualization; Writing - review and editing.

Kiarash Fereidouni: Writing - review and editing.

Iman Deilami: Resources; Supervision.

Human Arianpour: Resources; Project administration; Supervision.

Azadeh Andisheh Tadbir: Pathological assessment and interpretation; Validation; Writing - review and editing.

All authors reviewed and approved the final manuscript.

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Figures

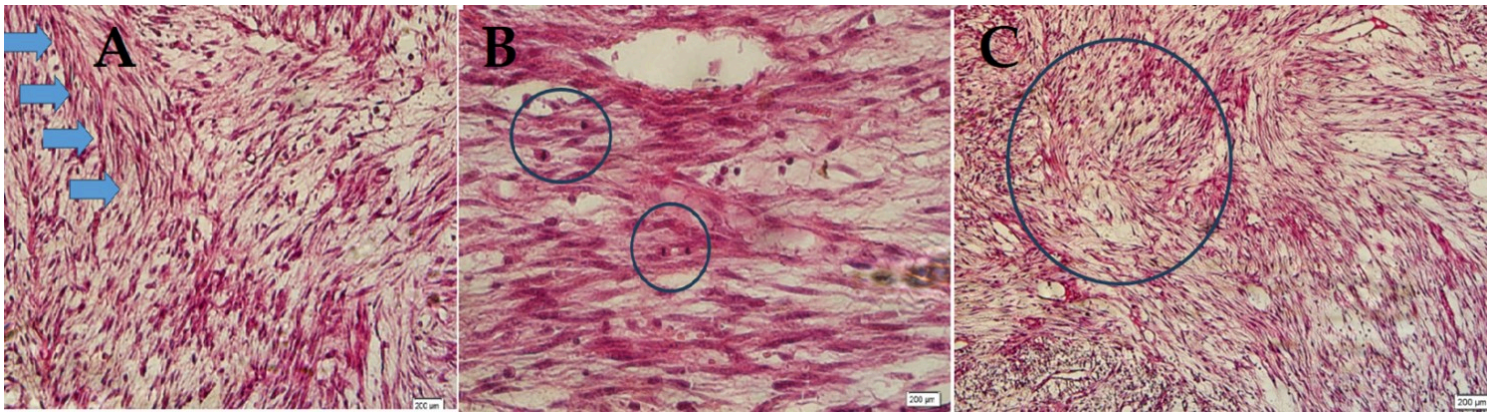


Figure 1

Pathological result of the lesion, A) herring bone bundles, B) Mitotic figures, and C) storiform pattern by small cell bundles

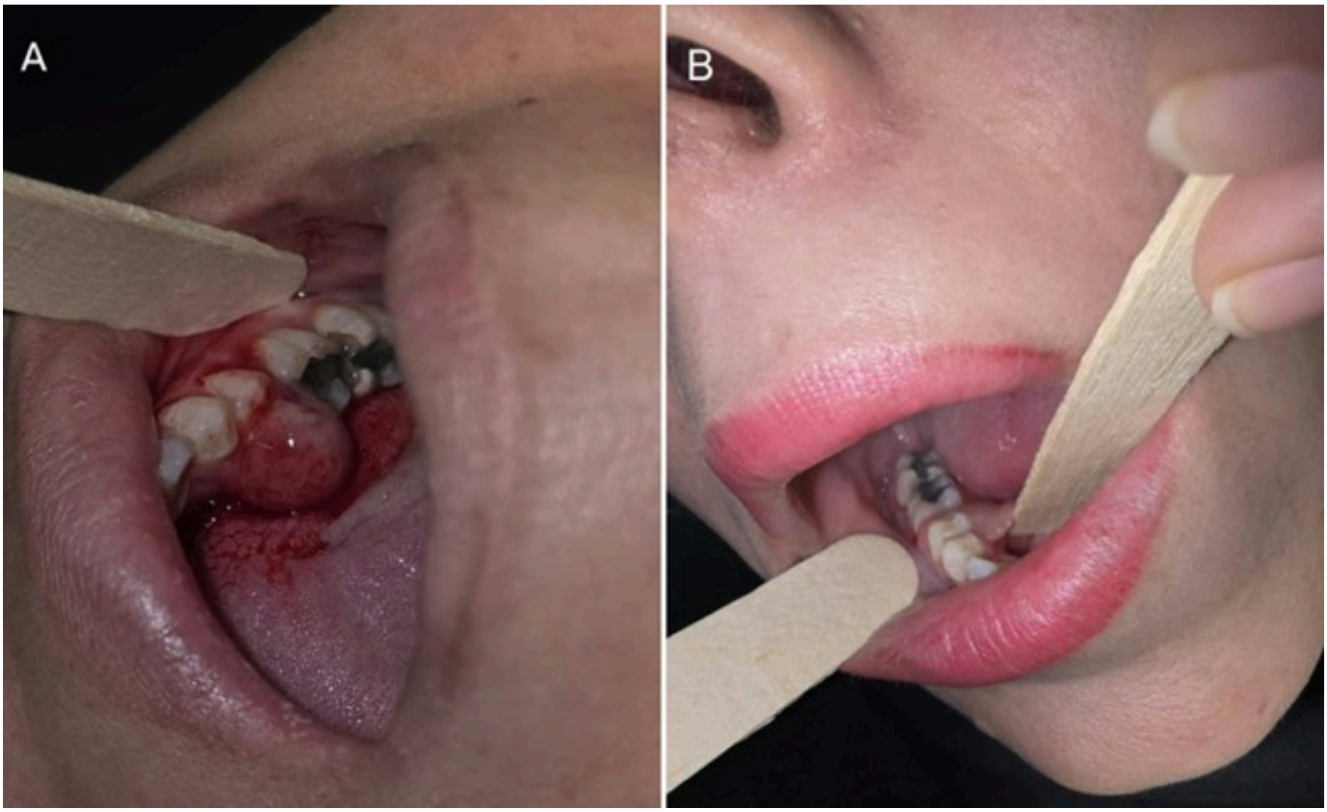


Figure 2

Clinical presentation of the lesion A) before excisional biopsy and B) 1 month after excision, based on physical examination findings

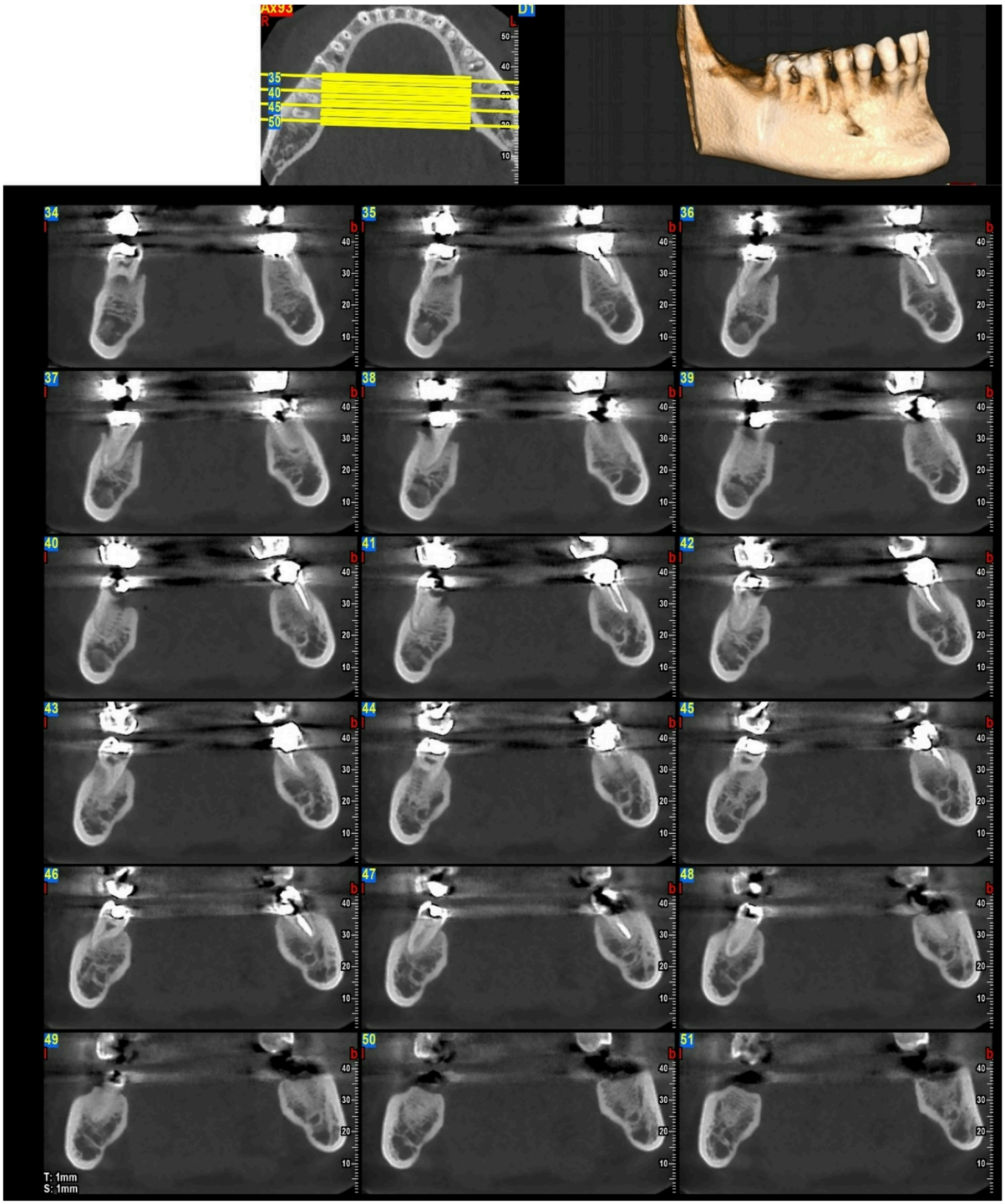


Figure 3

CBCT view of the right side of the mandible shows Loss of continuity of lingual cortex, ill-defind radiolucency distal of tooth #7, PDL band-like widening of tooth #6

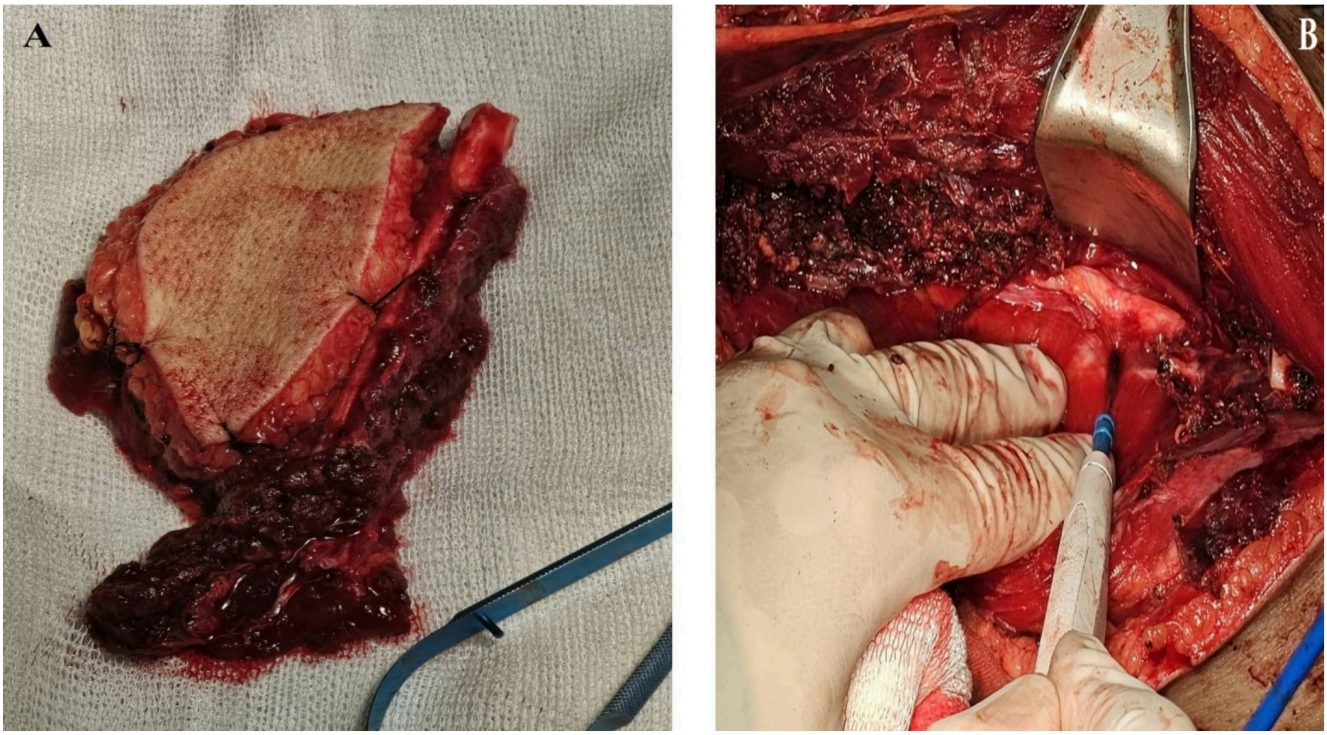


Figure 4

A) Image of the fibular flap, B) Image taken during surgery