Solitary Fibrous Tumor of the Mandible

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Abstract

A 41-year-old woman presented with a facial asymmetry in the mental region and a painful, well-circumscribed, tender mass in the right lower buccal vestibule, associated with extensive ill-defined bone rarefaction with subtle cortical bone resorption. Microscopically, a proliferation of bland spindle cells interspersed with collagen fibers and prominent staghorn-like blood vessels was observed. Immunohistochemical analysis revealed strong positivity for CD34, Bcl-2, CD99, and STAT-6, confirming the diagnosis of Solitary Fibrous Tumor (SFT). Conservative surgical enucleation was performed, and 4 years later, recurrence was observed with extensive bone involvement and moth-eaten margins resembling a malignant tumor. SFT is a distinctive spindle cell tumor of fibroblastic differentiation, characterized by prominent branching staghorn-like vessels and a specific NAB2::STAT6 gene fusion. We herein contribute with a central SFT of the mandible with recurrent behavior and radiographic appearance suggesting malignancy.

Keywords: Solitary Fibrous Tumors; Oral; Recurrent; Mandible

A 41-year-old woman was referred for evaluation of facial asymmetry in the mental region of the mandible of unknown duration. Intraoral examination revealed a painful, well-circumscribed, tender mass in the right lower buccal vestibule measuring 6 cm, associated with extensive ill-defined bone rarefaction with subtle cortical bone resorption in imaging studies. Following an incisional biopsy, a microscopic examination showed a proliferation of bland spindle cells interspersed with collagen fibers and prominent staghorn-like blood vessels. Immunohistochemical analysis revealed strong positivity for CD34, Bcl-2, CD99, and STAT-6, confirming the diagnosis of solitary fibrous tumor (SFT). The patient underwent conservative surgical enucleation, and the tumor showed positive margins microscopically. The patient was lost on follow-up over four years when she returned for evaluation of a slow-growing tumor at the same site. Imaging exams revealed a 5.5 cm hypodense lesion with irregular, moth-eaten margins in the right mandibular body, resembling a malignant tumor. Under the clinical hypothesis of recurrent SFT, an enucleation followed by curettage of bone cortices was performed. Macroscopically, the specimen was a fibrous tumor forming lobules with invaginated superficial vascular channels. The microscopic and immunohistochemical features were consistent with those observed four years earlier. Hypercellularity was evident but marked pleomorphism and necrosis were absent. Mitoses numbered 0 per 10 high-power fields and the Ki-67 index was less than 1% (Fig. 1). The final diagnosis was recurrent SFT of the mandible. There were no signs of recurrence or metastasis one year after the surgery, and long-term follow-up will be offered. SFT is a distinctive spindle cell tumor of fibroblastic differentiation, characterized by prominent branching staghorn-like vessels and a specific *NAB2::STAT6* gene fusion [1]. In the oral cavity, SFT typically presents as a painless, well-defined nodule, most commonly affecting the buccal mucosa in middle-aged adults, and is usually treated with conservative surgical removal [1–2]. Some cases may involve the lower vestibule as a well-defined, highly vascular mass closely associated with the mental neurovascular plexus [2]. The features considered in risk assessment models for extra-meningeal SFT include the age of the patient, tumor size, tumor site, necrosis, nuclear atypia, hypercellularity, mitoses per 10 high-power fields, lack of circumscription, and invasiveness [3–4], which are rarely reported in oral SFT, though they can occasionally reach large sizes and show bone location [1–2, 5]. The present intra-osseous SFT showed recurrent behavior, compromised margins, hypercellularity, and a size greater than 5 cm; the lack of pleomorphism, necrosis, or mitotic figures excluded high-grade or dedifferentiated SFT. We herein contribute with a central SFT of the mandible with recurrent behavior and radiographic appearance suggesting malignancy.

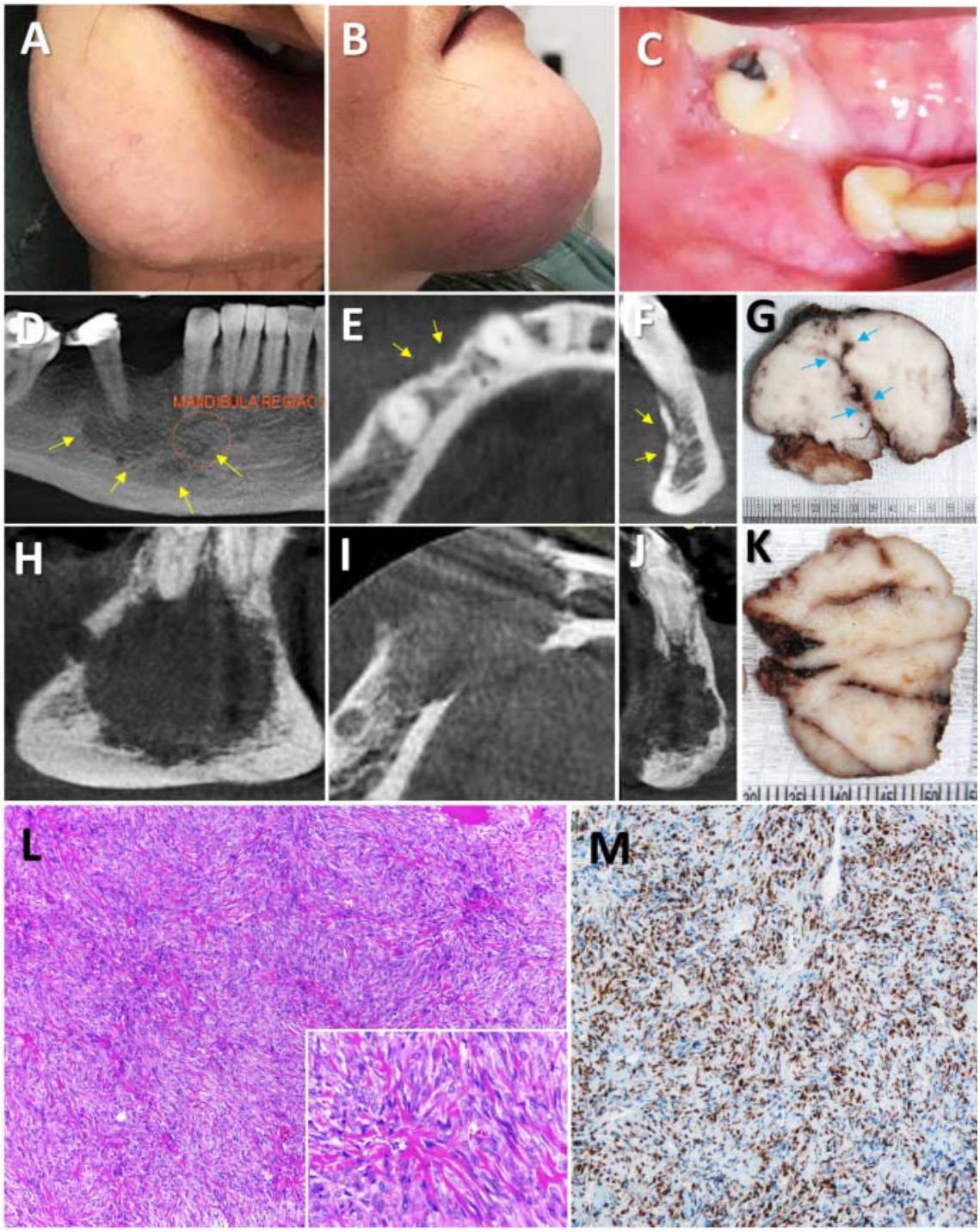

Fig. 1. A-C: Initial clinical presentation of exuberant facial asymmetry caused by a well-circumscribed swelling of smooth surface, located in the right lower buccal vestibule. Cutaneous superficial telangiectatic blood vessels covered the lesion (**A-B**, extraoral examination; **C**, intraoral examination). **D-F.** The tumor exhibited an ill-defined bone rarefaction, leading to slight buccal bone cortical resorption (yellow arrows). **G.** Grossly, the tumor appeared as an incompletely removed soft tissue fragment of fibrous consistency and ill-defined irregular base; a central staghorn blood vessel is evident (blue arrows). (**D-F**, Cone-beam computerized tomography; **G**, macroscopic examination) **H-J.** Four years later, a large hypodense lesion of irregular margins was noted causing exuberant bone destruction and cortical disruption. **K.** the gross appearance of the persistent tumor was of an ill-defined and lobular fibrous tumor. (**H-J**, Cone-beam computerized tomography; **K**, macroscopic examination) **L-M.**

Microscopic evaluation showing haphazardly arranged spindle to ovoid cells with indistinct eosinophilic cytoplasm and bland ovoid nuclei within a collagenous stroma were admixed with hyalinized staghorn blood vessels, and diffusely positive for STAT-6 in nuclear pattern (**L**, H&E, 100x, [Insert 400x]; **M**, Immunoperoxidase, 100x)

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Contributions

TCF, MA, MJR, and ACA wrote the manuscript and prepared the Fig. 1, JVP provided clinical information and images, ALOCR, WFPvH, MJR, and ACA reviewed the manuscript. All authors approved the images.

Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Conflict of Interest

The authors declare that they have no conflict of interest.

Data Availability

No datasets were generated or analysed during the current study.

Code Availability

There is no software or custom code associated with this paper.

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