A mixed image in the maxillary sinus

Flávia Sirotheau Corrêa Pontes, DDS, PhD,[a] Felipe Paiva Fonseca, DDS, PhD,[b]
Beatriz Helena Rodrigues e Silva, DDS,[a] Eduardo Rodrigues Fregnani, DDS, PhD,[c]
Sâmia Cordovil de Almeida, DDS,[a] Anderson Costa Sousa, DDS,[a] Marcondes Sena-Filho, DDS, PhD,[d]
Oslei Paes de Almeida, DDS, PhD,[b] and Hélder Antônio Rebelo Pontes, DDS, PhD[a]

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CLINICAL PRESENTATION

A 19-year-old male patient complaining of a diffuse swelling of 4 months' duration on the right side of his face was referred to our department. No palpable cervical lymph node was found, and patient's past medical history was noncontributory. Intraoral examination did not reveal any significant visual alteration in the right maxillary mucosa; however, a slight swelling in the maxilla, extending from the first to the third molars, could be noted during local palpation, demonstrating a hard consistency suggestive of bone cortical expansion (Figure 1A). There was no tooth mobility. Panoramic radiography revealed an irregular, ill-defined radiolucent image containing radiopaque foci on the right side of the maxilla and invading the maxillary sinus (Figure 1B). Computed tomography (CT) demonstrated a hypodense image containing small hyperdense foci. Axial sections revealed destruction of the posterior aspect of the maxillary sinus and of the lateral lamina of the pterygoid process, and coronal sections showed an infiltrative growth in the nasal cavity and the sagittal sections revealed orbital floor disruption (Figures 1C-E).

DIFFERENTIAL DIAGNOSIS

The nonspecific clinical features and the aggressive mixed radiographic presentation of the lesion led us to consider a broad range of lesions. Although central ossifying fibroma and a benign or malignant odontogenic tumor with hard tissue production were initially considered, malignant neoplasms, such as synovial sarcoma (SS), Ewing sarcoma family of tumors (ESFT), and osteosarcoma or chondrosarcoma, were considered more likely, given the destructive growth observed on the CT scan.

[a]Service of Oral Pathology, João de Barros Barreto University Hospital, Federal University of Pará, Belém, Brazil.
[b]Piracicaba Dental School, Oral Diagnosis Department, University of Campinas, Piracicaba, Brazil.
[c]Department of Oral Medicine, Sírio-Libanês Hospital, São Paulo, Brazil.
[d]School of Dentistry, UniEVANGÉLICA University Center, Anápolis, Brazil.

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Central ossifying fibroma is a benign neoplasm containing a fibrocellular stroma and variable amounts of mineralized structures and accounts for approximately 15.4% of all benign fibro-osseous lesions.[1] Although it can rarely present as multiple lesions, it is more frequently diagnosed as a solitary tumor, predominantly affecting females in their second to fourth decades of life and causing an asymptomatic swelling in the posterior region of the mandible.[1-3] In the current report, because of the painless growth of a lesion with a mixed radiographic appearance, central ossifying fibroma was initially considered. However, the infiltrative and destructive growth pattern of the tumor was not consistent with the well-demarcated features typically seen in central ossifying fibromas.

Similarly, the destructive growth of the tumor carrying a mixed radiographic appearance (feature used to exclude ameloblastoma and myxoma) made a benign odontogenic tumor highly unlikely, and we initially decided to consider only a malignant odontogenic tumor, such as ameloblastic fibrosarcoma (AFS) and odontogenic ghost cell carcinoma (OGCC), as a diagnostic option. OGCC more frequently involves the maxilla, whereas AFS is more common in the posterior region of the mandible, and both tumors may extend toward the maxillary sinus.[4,5] Adult males are the most affected, but some cases have been described in younger patients. A long-term persistent swelling followed by a rapid, painful growth is the most frequently described finding.[6-8] Radiographically, these tumors may appear as poorly defined mixed lesions, depending on the degree of dystrophic calcifications (i.e., AFS) and deposition of dentinoid material (i.e., OGCC). Because malignant odontogenic tumors can demonstrate an aggressive clinical course, we considered them a possibility.

SS is an aggressive high-grade neoplasm derived from undifferentiated mesenchymal cells and carries the specific t(X;18)(p11.2;q11.2) chromosome translocation.[9,10] SS is more frequently diagnosed in the extremities, whereas the head and neck region is affected in 3% to 10% of the cases, usually affecting males in their third to fifth decades of life.[10] Head and neck tumors usually present nonspecific clinical signs and symptoms, such as a progressive painless growth,[11] with calcifications being found in some cases.[12,13] Thus, because SS has been described in the

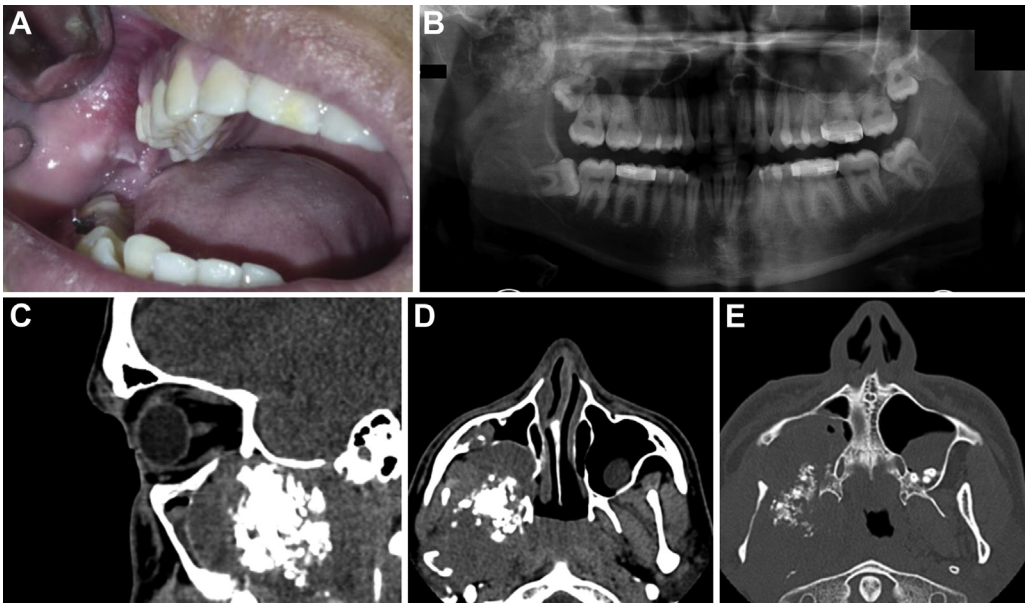

Fig. 1. Clinical and imaging features observed in this case. **A,** Intraoral examination did not show any evident alteration in the maxillary mucosa of the patient. However, palpation of the affected maxillary area demonstrated a slight cortical bone expansion. **B,** Panoramic radiography showed an ill-defined radiolucent image, with radiopaque foci invading the maxillary sinus. **C,** Soft-tissue window sagittal plane of computed tomography (CT) revealed the presence of hyperdense material inside the hypodense lesion that extensively obliterated the right maxillary sinus. Soft tissue **(D)** and hard tissue **(E)** window axial plane of CT scan showing the destruction of the posterior wall of the maxillary sinus, which is almost completely involved by the tumor.

paranasal sinuses,[12,14] it was also considered a diagnostic possibility in this case.

ESFT represents the third most common primary bone neoplasm, harboring the t(11;22) chromosomal translocation involving the *EWS* and *FLI-1* genes. Males are more affected in their first two decades of life and usually exhibit a painful swelling. The long bones, pelvis, and ribs are the most affected locations, but gnathic bone involvement is uncommon. Radiographically, a destructive radiolucent process with poorly defined borders is the main characteristic of this entity.[15,16] Although ESFT diagnosis in this case was supported by the patients' age, when gnathic bones are involved, the mandible is the most affected site, with the neoplasm only rarely affecting the maxilla and the maxillary sinus[17]; moreover, calcifications are not commonly found in ESFT.

Finally, osteosarcoma and chondrosarcoma are the two most common primary malignant bone tumors (excluding hematologic malignancies), and the involvement of the head and neck region has widely been documented.[18] Adult males are the most affected patients, with a rapidly growing painless swelling. Radiographically, osteosarcoma and chondrosarcoma usually cause cortical bone destruction, ranging from ill-defined radiolucent to variably mixed images, occasionally presenting the so-called "sun-ray" and "Codman triangle" findings.[18,19] The aggressive mixed radiographic features of this case led us to consider osteosarcoma and chondrosarcoma as the most likely diagnostic possibilities.

DIAGNOSIS

An incisional biopsy under local anesthesia was done, and microscopic examination revealed a malignant neoplasm comprising two cellular components. Epithelial cells were arranged in small foci and exhibited abundant eosinophilic cytoplasm with indistinct cell borders and round-to-ovoid nuclei. These epithelial nests were surrounded by pleomorphic spindle cells containing scarce cytoplasm and hyperchromatic nuclei organized in short bundles that predominated in the histologic sample. Scattered mitotic figures could be found, but necrosis was absent (Figure 2). Considering the two cellular components observed, microscopic diagnosis was highly suggestive of SS, but the immunohistochemical study was done to exclude other less likely possibilities, such as hemangioendothelioma, sinonasal hemangiopericytoma, rhabdomyosarcoma, ESFT, lymphoma, osteosarcoma, and melanoma. Diffuse positivity for vimentin was found in spindle cells, for TLE1, Bcl-2, and CD99 in both spindle and epithelial cells, and for AE1/AE3 and EMA in epithelial cells (Figure 3). The Ki67 proliferative index was higher than 40%, and reactions against S100, LCA, desmin,