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Case Report

Ossifying fibroma of the maxilla: A case report with literature review ☆

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ABSTRACT

Ossifying fibroma is a benign fibro-osseous lesion arising from the periodontal ligament cells. The lesion may progressively enlarge with the mass affecting the mandible or maxilla, resulting in facial deformities and tooth displacement despite its benign nature. Here, we presented a case of an 18-year-old female with ossifying fibroma in the maxilla extending to the maxillary sinus, infraorbital area, and skull base, resulting in considerable facial asymmetry. Since the primary treatment of ossifying fibroma is surgical resection, it is essential to determine the areas where the lesion has expanded, where a 3-dimensional computed tomography scan could play a critical role in providing such information. A complete surgical excision and histopathologic examination in treating this patient are crucial, made possible by a meticulous preoperative radio imaging technique.

Introduction

Ossifying fibroma (OF) is defined as a benign fibro-osseous, expansile lesion with a circumscribed margin arising from the cells of the periodontal ligament. It is characterized by the replacement of typical bone architecture with either fibroblasts or collagen fibers containing a variable amount of bone or cementum-like tissue or both [1]. OF has been known for its progressive, slow-growing behavior; however, this characteristic may vary broadly. Some lesions may eventually become massive, entailing significant aesthetic problems and functional deformities such as facial asymmetry and tooth displacement. Ossifying fibroma, cemento-ossifying fibroma, and cementifying fibroma are often used to describe these entities due to their bone and cementum-like tissue content in ossifying fibromas [2].

Ossifying fibroma is frequently diagnosed among the population in their second to fourth decade with a male-to-female ratio of 1:5, seldom occurring in children, adolescents, and the elderly. Facial bones, particularly the posterior region of the mandible, are the repeated site of emergence for 75% [3,4]. Moreover, evidence has described their development in the paranasal sinuses, frontal-ethmoid-sphenoid bone, and orbital floor [5]. However, the involvement of the sinonasal tract is relatively uncommon [6].

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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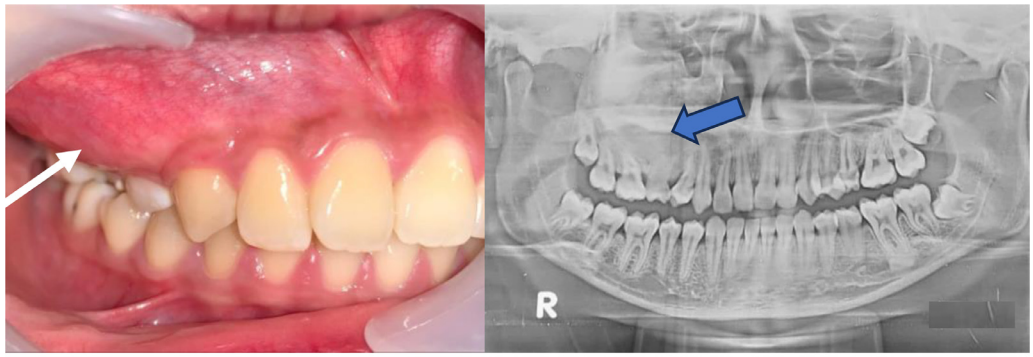

Fig. 1 – Ossifying fibroma. (A) The intraoral picture shows a lump in the right maxillary region, (b) orthopantomography.

Surgical eradication is a modality of choice to manage OF. Enucleation and curettage can excise a small and well-defined lesion, whereas a sizeable expanding lesion requires radical excision within a normal margin followed by aesthetic reconstruction. The recurrence rate ranges between 0% and 28% of cases in literature; therefore, some authors have advised entire surgical resection at the earliest stage possible [7,8].

Here, we aimed to present a case report of an 18-year-old female diagnosed with ossifying fibroma with minor symptoms involving the right maxilla extending to the sphenoid and pterygoid bone, discovered by radiography and confirmed by histopathologic investigation.

Case presentation

A female, 18 years old, was referred to our center with an asymptomatic lump that had been growing for over 7 years. She denied having a history of any toothache or trauma in the jaw. Superficial examination showed swelling on the right maxilla region, extending to the infraorbital region, displacing the nasal base to the left side, leading to facial asymmetry, albeit no discoloration compared to the surrounding area. Intraoral examination showed a lump that was solitary, solid, not ulcerated, and firm in consistency from maxillary region 13-17 expansion to the palatal area of tooth 17; no midline shift was seen.

Investigations/imaging findings

Orthopantomography (OPG) of the maxilla showed a 4 cm mixed bone density lesion with a diffused margin around the root of teeth 14, 15, 16, 17, and 18 with tooth resorption (Fig. 1B, arrow), extending from the alveolar process into the maxillary sinus. Non-contrast computed tomography (CT) scan examination revealed a solid hyperdense mass, extensive bone expansion with intact cortex and corticomedullary differentiation in the right sphenoid bone, pterygoid bone to the alveolar process with well-defined margin, measuring approximately 6.4 × 4.0 × 4.5 cm. The mass filled the right maxillary and sphenoid sinus, causing erosion of the palatine bone and pushing the right nasal concha to the left side. An image of mixed density was observed inside the lesion, as shown in Figure 2.

Three-dimensional CT used for extensive mass excision management was planned. Although the essential data of the tumor and the adjacent anatomy have been readily provided by 2-dimensional images, a more detailed structure of the tumor was still necessary to assist in surgical planning and was achieved by a 3-dimensional printed model (Fig. 3). The 3D anatomical model was used to evaluate the size and extent of the pathology in Figure 4. CT scan aids clinicians in determining the necessary surgical management, with complete resection reducing the chance of recurrence of this mass.

Differential diagnosis

Initial CT scan findings led to the suspicion of fibrous dysplasia with a differential diagnosis of ossifying fibroma. However, a further histopathologic investigation showed islands of trabeculae of mature bone, characterized by prominent osteoblastic rims and highly cellular fibrous connective tissue matrix surrounded with oval spindle cells, consistent with the diagnosis of ossifying fibroma.

Discussion

World Health Organization (WHO) classified oral and maxillofacial benign fibro-osseous lesions into 3 classifications: ossifying fibroma, fibrous dysplasia, and osseous dysplasia [2,9]. The first person to use the ossifying fibroma term was Montgomery in 1927. Furthermore, the first representation of ossifying fibroma was described by Menzel in 1872 in a 35-year-old female with a large nodule in the mandible [7]. Since then, the nomenclature surrounding OF has been diverse. WHO published its first histological typing of Odontogenic tumors in 1971, separating tumors into 3 main categories:

1. Neoplasm and other tumors related to odontogenic apparatus
2. Neoplasm and other tumors related to the bone
3. Epithelial cyst

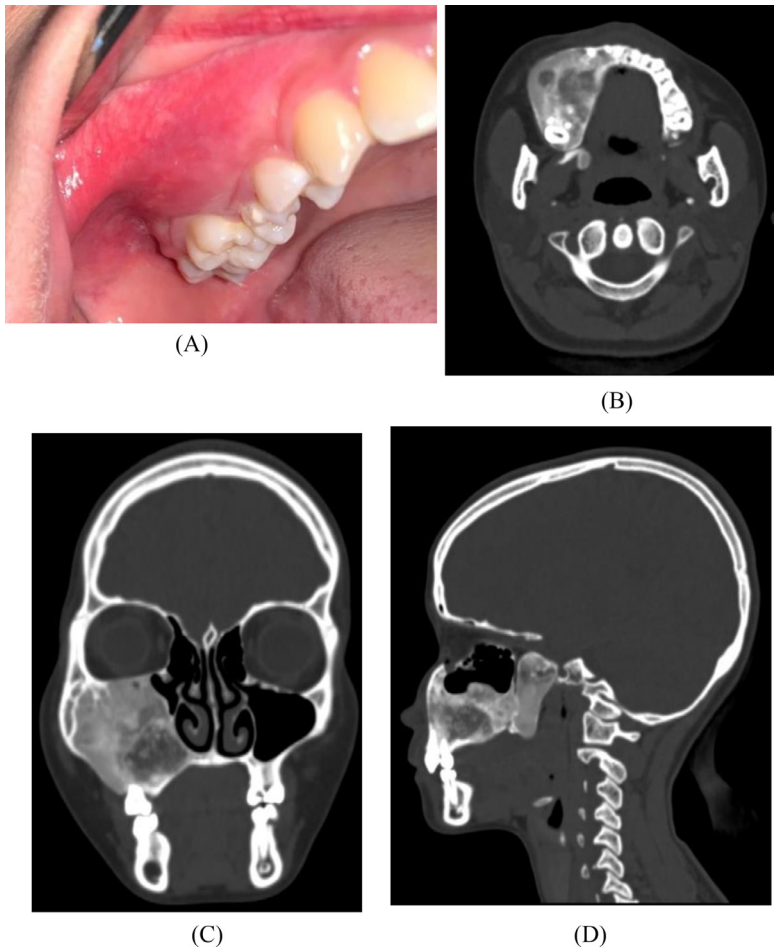

Fig. 2 – Ossifying fibroma. (A) Intraoral inspection shows a slow-growing lesion of the right maxilla. (B) Noncontrast axial CT image demonstrates extensive bone expansion with intact cortex and corticomedullary differentiation of the right maxilla to alveolar process with a clear margin, (C) noncontrast coronal CT image and (D) noncontrast sagittal CT image demonstrates diffuse radiopaque lesion with a mixed bone density characteristic filling the maxillary region extending to the infraorbital and lateral nasal.

The second category consists of ossifying fibroma, cementifying fibroma, fibrous dysplasia, and cemento-ossifying fibroma, which the latter was renamed by WHO into the term "ossifying fibroma" [2]. The pathogenesis is yet to be elucidated, although several authors have stipulated its relation to a disruption in the dental tissue maturation, forming cement and bone tissue [10]. Histopathologic results of the fibro-osseous lesion have many similarities, and exact correlation of the clinical, histopathologic, and radiology findings are requisite for diagnosis.

There is no established staging system for OF, with most literature on the field being case reports or case series. For treatment planning, the categorization of OF may be made based on several aspects: anatomical size [11], radiological [12], and histopathologic features [13]. The anatomical size and its cosmetic considerations may not correlate well with the histology, that is, a small lesion could have an advanced maturity, and vice-versa. Due to the lack of treatment guidelines, it is imperative to assess each patient individually, focusing on the lesion's extension and border. CT scan and 3D-formatted images were appropriate for our patient, as a 2D radiograph could only provide the presumptive diagnosis.

OFs have a neoplastic potential [14]; therefore, the histopathologic examination is essential in determining the treatment plan pre- and postoperatively [15]. However, preoperatively, it is more critical to determine the lesion border to facilitate total excision; the histopathologic examination will further determine if adjuvant chemotherapy is needed, should the lesion have a malignant characteristic. Preoperative biopsy may also help to differentiate other benign fibro-osseous lesions. Diagnosing OF requires a multimodality approach for accurate indication-specific radiological assessment with OPG, CT scan or cone-beam computed tomography (CBCT), and MRI. OPG is considered a primary and adequate diagnostic tool for OF. CT scan or CBCT is used to see the internal structure of the tumor so that it can determine diagnostic staging [12]. It is helpful in pre-surgical planning to assess the precise extension of the tumor. MRI, combined with CT scan, helps to make a correct diagnosis; it also allows assessment of risk structure in the case of intracranial or intraorbital exten-

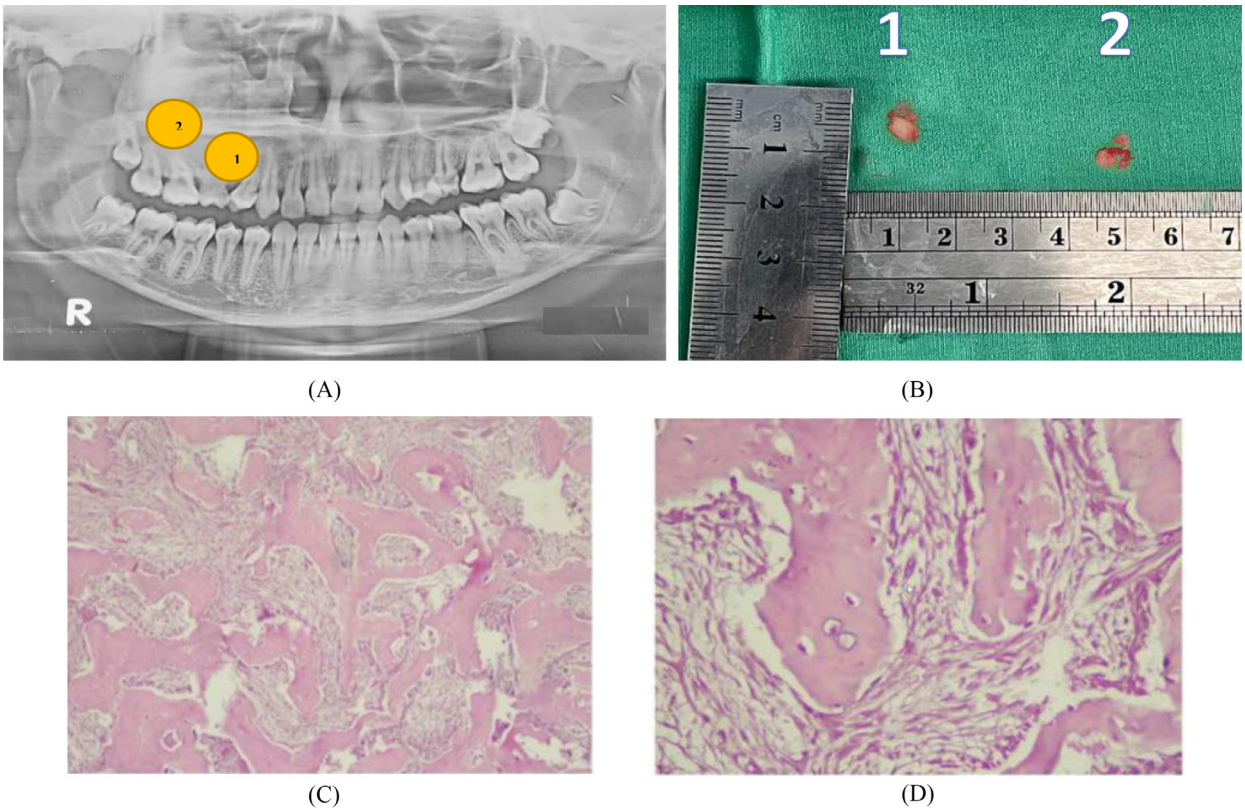

Fig. 3 – Ossifying fibroma (A, B). Incisional biopsy for histopathologic diagnosis. The histopathologic image suggests ossifying fibroma, H&E section (C) 100× and (D) 400× original magnification.

sion and, at last, during follow-up to discriminate scar tissue from mucocele as in the case of the patient treated with midfacial degloving. Since the tumor size was already significant, extending to the skull base, a CT scan with contrast was an appropriate modality in our case; meanwhile, CBCT is used for a smaller mass without extension. Furthermore, there has been no extension to the intracranial or soft tissue infraorbital, making MRI unnecessary.

Clinical presentation

OF's origin has been odontogenic for several years, with periodontal ligament being the most frequent site [1]. However, this postulation has been questionable by the recent presentation of lesions in the frontal, temporal, sphenoid, and ethmoid bones, in which their microscopic features were almost indiscernible. Two possible explanations for this condition are the ability of the lesions to develop from an ectopic periodontal membrane and its mesoderm nature. Meanwhile, the tumor can be produced by similar differentiation of some primitive mesenchymal cells [10]. Extensive mesenchymal induction into bone and cementum fundamental in odontogenesis could be attributable to OF effects on the mandible bone. Therefore, OF development may be a product of any error in the tissue induction process. Factors suggested as triggers include trauma, previous extraction, prior periodontitis, and possible genetic defects, especially mutations of the hyperparathyroidism 2 (HRPT2) gene [3,5].

OF in the early stages was found to be asymptomatic. The tumor then grows gradually to the point that its size causes a lump/swelling of the involved bone, significant functional changes, and facial asymmetry. Clinical features of pain and paraesthesia in Ossifying fibroma are unusual. Mobility and root reabsorption of the involved teeth are common [7]. Our presented case of an 18-year-old female aligned with the "classical" presentation of ossifying fibroma, which is female, has a slow-growing asymptomatic painless lump since 7 years ago; the progressive growth of the lump, affecting the maxilla region, expanded to the right maxillary and sphenoid sinus, sphenoid bone, pterygoid bone, and alveolar process.

Radiographic feature

The radiological appearance has a different pattern based on the amount of tissue mineralization. It is usually a round, oval, relatively smooth, well-defined, and expansile mass. Liu et al. [16] discussed 2 patterns of ossifying fibroma in radiology, including cystic lesions (uni-cystic or multi-cystic) and mixed-density lesions with or without a sclerotic margin, often accompanied by cortical expansion [7,9]. Furthermore, the centrifugal growth pattern presents as the main finding instead of a linear one, being the essential diagnostic feature, leading

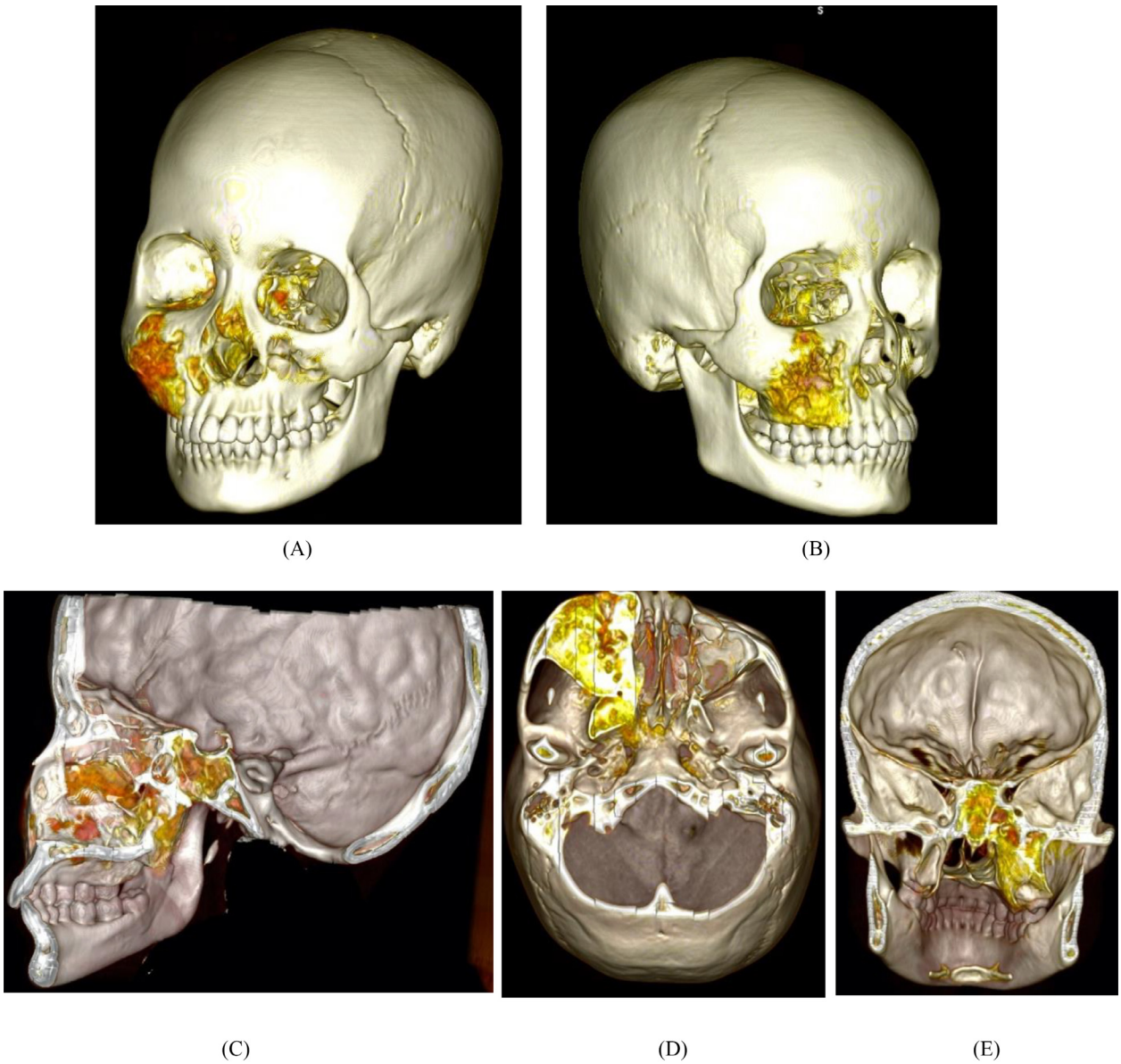

Fig. 4 – (A-E) Ossifying fibroma. CT scan with 3-dimensional formatted images of the maxilla shows mixed density, well-circumscribed bone extending to the right maxillary and sphenoid sinus, right sphenoid bone, and pterygoid bone into alveolar processes.

to the expansion of the tumor in all directions. Radiological findings of ossifying fibroma depend on the lesion's developmental stage.

1. In the initial stages, it appears as a well-defined radiolucent with clear boundaries from surrounding healthy bone with a ground-glass appearance.
2. Later, the lesion tends to enlarge and become a radiolucent-radiopaque mixture with an opacity that appears in the center of the lesion with a lower density compared to the neighboring bone.
3. As the lesions mature, they appear with asymmetric opacities forming concentric bony trabeculae, surrounded by peripheral osteo-condensation often described as an eggshell appearance.

In large expansive lesions, radiological images comprise the thinning of the cortex and weakening of the mass margin. Root divergence resorption emerges due to continuous mass growth [8]. Radiological findings in aggressive lesions are indistinct mass boundaries such as perforations in the cortical bone [4]. In the case reported here, an expansile tumor in the right maxillary region showed a mixed radiolucent-radiopaque density with a circumscribed margin and root resorption, and there are no concentric bony trabeculae with an eggshell appearance; therefore, in our case, this was type 2 according to maturity. This lesion had an equal outward extension in all directions with thinning of the cortical bone and divergence root resorption of teeth 14, 15, 16, 17, and 18. Advanced imaging modalities such as CT scans with bone win-

dow technique are the most important investigations, especially in extensive cases, because they determine the tumor's extent and surrounding tissue deterioration.

Differentiating between OF and other similar lesions of the oral and maxillofacial areas can be challenging, whereas, in our case, the location and density of this lesion make it a differential diagnosis of fibrous dysplasia. Fibrous dysplasia is primarily found in the maxilla, and OF is mainly located in the mandibula, distinguished by these characteristics: (1) Fibrous dysplasia tends to grow longitudinally, whereas OF has compact spherical growth. (2) Fibrous dysplasia has an ill-defined margin with the regular appearance of the involved bone preserved, whereas OF is a well-defined margin with the appearance of a tumor, contour deformity, and disruption of the bone shape [9,17]. (3) OF has a bone density and soft tissue mixture with a thick margin, whereas FD has a ground glass appearance.

Treatment and prognosis

Complete surgical removal is the only curative treatment for OF [6]. The surgical management of ossifying fibroma involves one of these methods: enucleation, curettage, and resection. Clinical and radiological features are needed to determine appropriate surgical management. Smaller and well-circumscribed nodules can be extracted by conservative curettage or enucleation until a healthy bony margin is reached. On the contrary, more radical surgical resection is needed by larger lesions for healthy bone margins and aesthetic recontour. Various authors suggest complete lesion removal as the earliest possible stage [7,18]. Complete eradication of maxillary OF produces more challenge than mandibular OF due to the difference in bony characteristics between the mandible and maxilla and the space available in the form of expansion in the maxillary sinus. Curative treatment may be achieved through supra-marginal excision in most cases; however, a multidisciplinary collaboration between neurosurgeons, plastic surgeons, and otolaryngologists is necessary when the OF involves skull base, orbit, ethmoid and sphenoid sinuses to attain the entire removal of the lump while preserving functional and aesthetic result. In this case, a CT scan is paramount in presurgical arrangement to determine the accurate tumor extension. Appiani et al. [6] recommended presurgical biopsy given the overlapping radiological and clinical features among fibro-osseous lesions, even if the imaging may suggest OF. It is vital to eliminate the possibility of malignant tumors, namely sarcomas, that have broad feature presentations such as intact margins, bone sclerosis, bone erosion, and gross bone destruction, along with different extents of an internal calcification or ossified matrix that may complicate differential diagnosis.

Recurrence rates vary from 6% to 28% among mandibular OF patients. Although the maxillary OF recurrence remains unclear, it may entail a higher probability because of the incredible challenge posed by surgical removal and the larger size. If reoccurrence is identified during follow-up, mandatory conservative treatment is advised. Moreover, radiotherapy is not recommended and even contraindicated given its increased risk of malignant transformation of approximately 0.4%-44% except for specific subtypes of ossifying fibro-myxoid tumors. Nevertheless, no evidence suggests the malignancy risk of OF [8] with a satisfying overall prognosis—after all, there is no incidence of metastatic disease from OF despite its local invasion and recurrence tendency. Recurrence can occur due to inadequate enucleation, and the recurrence period is unpredictable, ranging from 6 months to 7 years after surgery [5]. Therefore, it is recommended that the clinician consider annual follow-up [18] for up to 10 years [5]. Serial radiography with or without contrast will aid in the identification of the recurrence; however, in our patient, regarding the extension to the skull base, maxillary sinus, and infraorbital area, the appropriate examination for follow-up would be the CT Scan with contrast [19].

Conclusion

Our case represents a case of maxillary ossifying fibroma and underlines the possibilities for clinical and radiographic findings spectrums. The diagnosis of ossifying fibroma is oriented toward clinical and radiological results of the lesion, and histological investigation confirms the diagnosis. CT scan played an essential role in assisting total resection, which remains the goal of this lesion; however, surgery involving ethmoid and sphenoid sinuses will require collaboration. Patients who have undergone total or subtotal resection should be followed for recurrence with serial imaging.

Patient consent

The patient has consented to this paper's publication on the condition of requisite anonymity of obtained data.

This paper has also obtained mandatory ethical clearance from Ethical Committee of our hospital.

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