

# FIBROUS DYSPLASIA OF THE MAXILLA: CASE REPORT



## CASE DESCRIPTION

MSSAC, female gender, caucasian, 36 year old patient. Ex-smoker with high blood pressure condition.

Reason for consultation was facial asymmetry.

Both panoramic x-ray and CT scan revealed a radiopaque mass extending from the upper right central incisor to the maxillary tuberosity, with obvious maxillary sinus invasion.

An incisional biopsy was performed using a trephine. The anatomic-pathological examination revealed a fibro osseous benign lesion consistent with fibrous dysplasia (monostotic).

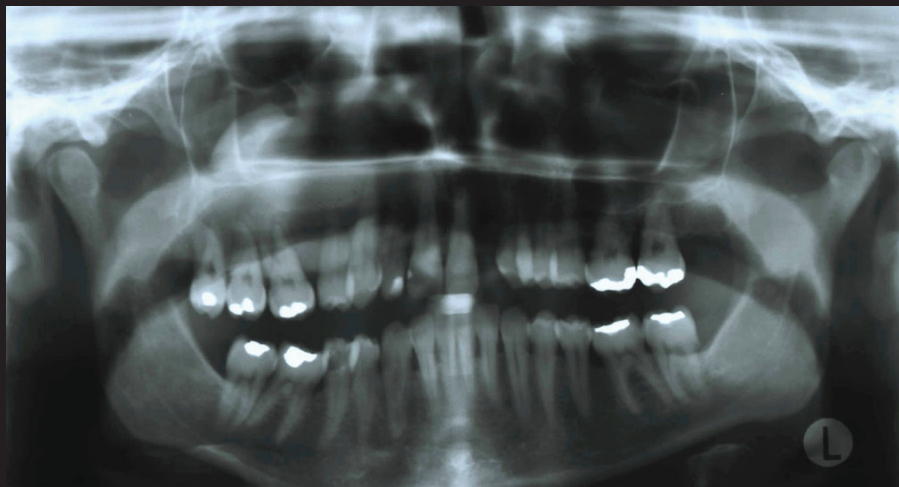
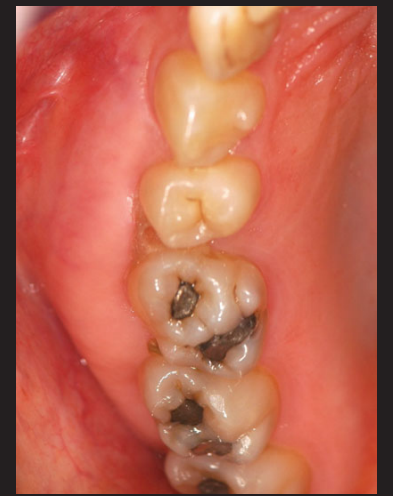
The patient, after being informed of the therapeutic options and because there was no functional or esthetic compromise, decided to monitorize the lesion and refused any surgical approach.



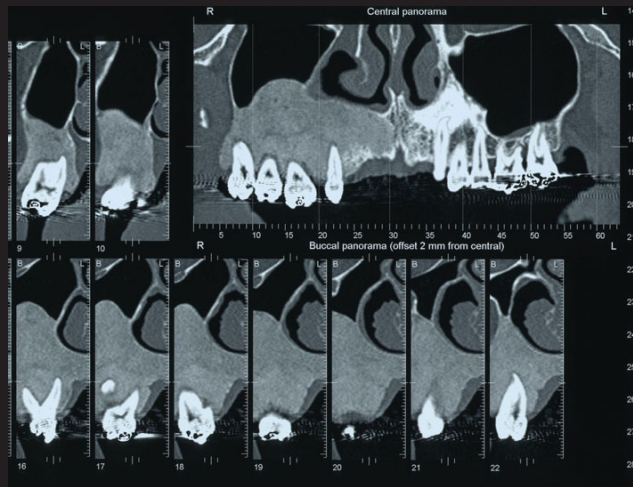
Extraoral photographs



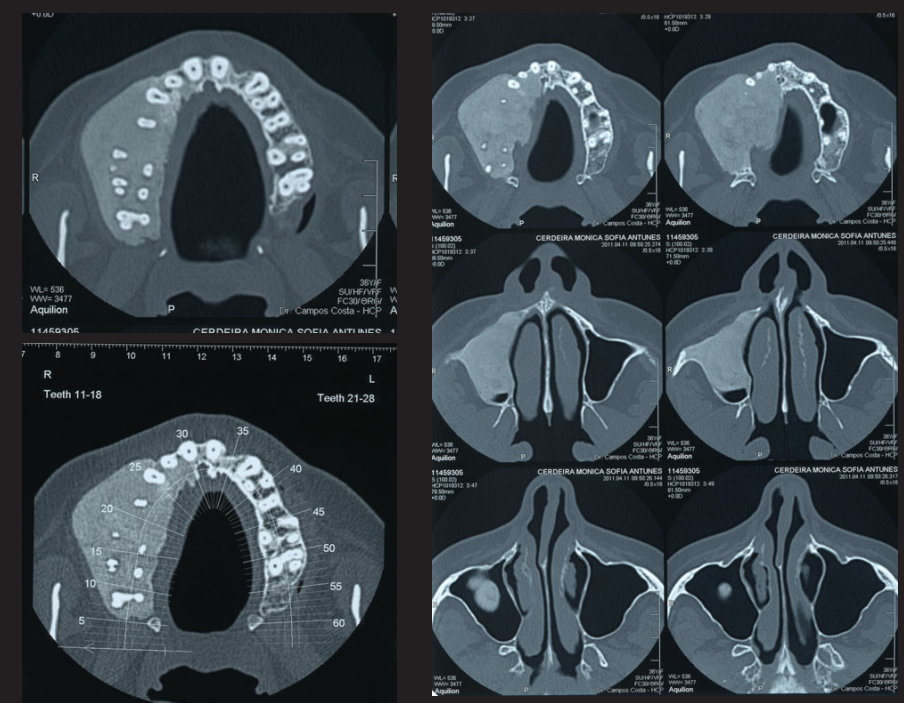
Intraoral photographs



Panoramic X ray



CT scan: coronal view



CT scan: axial view

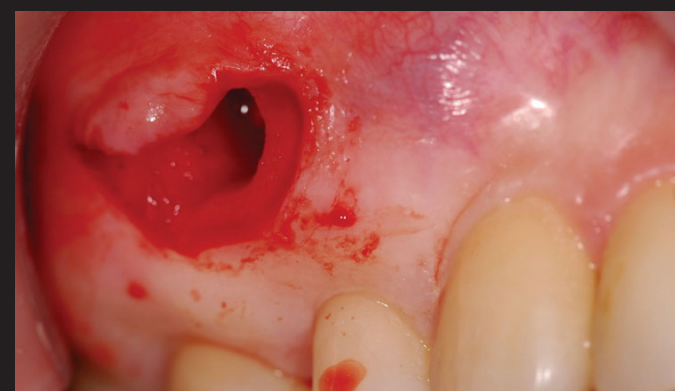
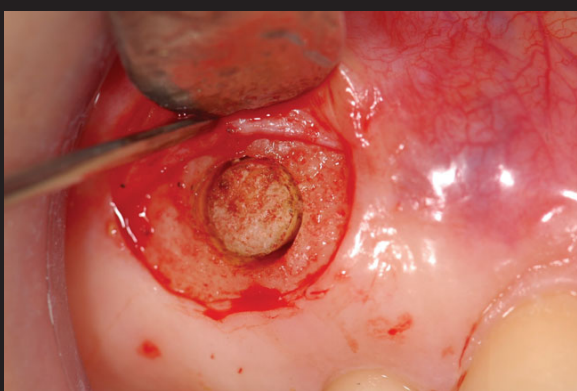
## DISCUSSION

Fibrous dysplasia is a rare bone disease that represents 2.5% of all bone diseases and 7% of all benign bone tumors<sup>1</sup>. It is a sporadic condition that results from a post-zygotic mutation in the *GNAS12* gene (protein linked to the guanine nucleotide,  $\alpha$ -stimulant of polypeptide 1 activity) and is characterized by the replacement of bone with cellular fibrous tissue containing foci of ossification<sup>3,4</sup>. It can involve a single site (monostotic fibrous dysplasia), multiple sites (polyostotic fibrous dysplasia)<sup>5</sup> or occur as part of the McCune- Albright<sup>6</sup>.

In the jaws, fibrous dysplasia is more frequent in the maxilla than in the mandible and can cause severe deformities and blindness<sup>7</sup>. The disease seems to stabilize after bone maturation<sup>3</sup>.

Fibrous dysplasia can be distinguished from ossifying fibroma by the fact that the latter always presents as a circumscribed lesion of varying density radiographically, and contains primarily, if not exclusively, lamellar rather than woven bone lined by osteoblasts<sup>8</sup>. Fibrous dysplasia has often been confused with osseous lesions of hyperparathyroidism indicating similar histopathologic features<sup>9</sup>.

Surgical treatment includes osteoplasty and pediculated grafts<sup>10</sup>.

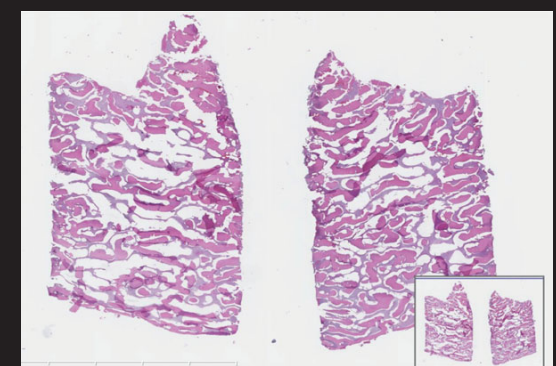


Incisional biopsy with trephine

## CONCLUSION

Fibrous dysplasia is a rare and benign bone disease that can compromise both esthetics and function of the affected individuals, especially when the craniofacial complex bones are involved.

In those situations surgical treatment is recommended in order to restore the normal anatomy of the affected structures. When skeletal growth is stabilized and no functional or aesthetical impairment occurs, the therapeutic approach would be the periodic monitorization of the bone lesions.



Histology

## REFERENCES

- Ricalde P, Magliocca KR, Lee JS. Craniofacial fibrous dysplasia. *Oral Maxillofac Surg Clin North Am* 2012; 24:427.
- Pagotto LE, Maniani PB. Displasia fibrosa monostótica da maxila: revisão da literatura e relato de 2 casos. *Rev. Cir. Buco-Maxilo-Fac.* 2009;9(2):23-32.
- Ricalde P, Horswell BB. Craniofacial fibrous dysplasia of the fronto-orbital region: a case series and literature review. *J Oral Maxillofac Surg* 2001;59(2):157-67; discussion 167-8.
- Marie PJ, de Pollak C, Chanson P, Lomri A. Increased proliferation of osteoblastic cells expressing the activating Gs a mutation in monostotic and polyostotic fibrous dysplasia. *Am J Pathol* 1997;150(3):1059-69.
- Barnes L, Eveson JW, Reichart P, Sidranski D. Pathology and Genetics of Head and Neck Tumors. Lyon, IARC Publications, 2005.
- Albright F, Butler A, Hampton A, Smith P. Syndrome characterized by osteitis fibrosa disseminata, areas of pigmentation, and endocrine dysfunction, with precocious puberty in females: report of 5 cases. *N Engl J Med* 1937;216:727-46.
- Alvarez LC, Cardoso CL, Fleury RN. Monostotic fibrous dysplasia: a 23-year follow-up of a patient with spontaneous bone remodeling. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2009;107(2):228-34.
- Barnes L, Peel RL, Verbin RS, Goodman MA, Appel BN. Diseases of the bones and joints. In: Barnes L, editor. *Surgical pathology of the head and neck*. New York: Marcel Dekker 1985; 883-1044.
- Caudill R, Saltzman D, Gaum S, Granite E. Possible relationship of primary hyperparathyroidism and fibrous dysplasia: report of case. *J Oral Surg* 1977;35(6):483-90.
- Valentini V, Cassoni A, Marianetti TM, Terenzi V, Fadda MT, Iannetti G. Craniofacial fibrous dysplasia: conservative treatment or radical surgery? A retrospective study on 68 patients. *Plast Reconstr Surg.* 2009;123(2):653-60.