Pycnodysostosis is a rare, autosomal recessive genetic condition that causes a decrease in bone remodeling due to a mutation in the cathepsin K gene, resulting in clinical and radiographic manifestations, characteristic of the syndrome. This report aims to describe 2 clinical cases of pycnodysostosis with orofacial involvement. The patients had a short stature, height ranging from 134 cm to 152 cm, stunted extremities, open fontanelles and cranial sutures, osteosclerosis, and medical history of repeated fractures in the left tibia, associated with small impact traumas. In the buccomaxillofacial complex, absence of pneumatization of the facial sinuses, maxillary osteomyelitis after dental extraction, maxillary atresia, increased mandibular angle, and enamel hypoplasia were related. It is concluded that the knowledge of clinical, radiographic, and oral and maxillofacial findings of this syndrome are important for the diagnosis and treatment of patients in the multiprofessional context, thus avoiding complications arising from dental procedures.

20190010

SOFT PALATE FISTULA: A CASE REPORT

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Soft palate fistulas are rare anomalies of doubtful pathogenesis. Many cases appear to be congenital, possibly related to a defect in the development of the second pharyngeal pouch. Some fistulas may result from infection or surgery in the tonsillar region. The lateral soft palate fistulas are usually bilateral but may occur only on 1 side. They are more common in the anterior tonsillar pillar but may also involve the posterior pillar. Classically, the perforations are asymptomatic, ranging from a few millimeters to more than 1 cm. Few cases have been associated with other anomalies. A 17-year-old female patient sought care due to a "hole in the mouth." She reported having the lesion from birth and was asymptomatic. At the clinical examination, a blind bottom perforation was observed, on the left, with a depth of about 6 mm. The patient was referred for genetic counseling, and no local treatment is required.

20190018

HYBRID CENTRAL GIANT CELL LESION AND AMELOBLASTOMA OF THE MANDIBLE

RUBIA TEODORO STUEPP, LUIZ HENRIQUE GODOI MAROLA, FILIPE MODOLO, and, ROGÉRIO GONDAK

Hybrid lesions encompass the occurrence of different entities in 1 lesion. A 67-year-old woman was referred to the oral and maxillofacial surgery service for treatment of mandibular central giant cell lesion (CGCL) previously diagnosed. Intra-oral examination revealed edentulism and a painless swelling extending to the buccal vestibule with hard consistency, without fluctuation and covered with normal mucosae, for an unknown period. Panoramic radiograph revealed a large, multilocular, and well-defined radiolucent lesion extending from the region of teeth #32 to #46, with no evidence of osseous perforation. Initial treatment with intralesional corticosteroids was performed. After 18 months, an increase of the osteolytic lesion extending from the anterior to the posterior left side of the mandible was noted radiographically. After incisional biopsy, the microscopic examination revealed an ameloblastoma associated with CGCL. Marsupialization was performed, and later the enucleation of the residual lesion. The follow-up is still being conducted.

20190019

CLEAR CELL ODONTOGENIC CARCINOMA: A RARE CASE REPORT

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Clear cell odontogenic carcinoma (CCOC) is a rare malignant neoplastic process originating from the odontogenic epithelium presenting high aggressive potential. The patient, a 45-year-old male, was referred to an oral and maxillofacial surgery and traumatology referral service, reporting painful symptoms after a local anterior mandible region fracture. A histopathologic examination revealed epithelial neoplasia fragments characterized by cell proliferation in islands, cords, nests, and, occasionally, cystic spaces. The proliferating cells were pleomorphic, with a markedly clear cytoplasm. An immunohistochemical analysis was then performed, and a strong immunohistochemical reaction to CK14 and CK19 antibodies was detected for the neoplastic epithelial cells. Immunostaining was consistent with a neoplastic process of odontogenic origin. The established diagnosis was, thus, determined as CCOC. The patient was then referred to a cancer treatment reference service to undergo surgical treatment. The patient is currently in regular follow-up, without any clinical-radiographic signs of recurrence.

20190024

CALCIFYING ODONTOGENIC CYST: A CASE SERIES

LAÍS DE BARROS PINTO GRIFONI, MATHEUS HENRIQUE LOPES DOMINGUETE, CASSIO EDVARD SVERZUT, ALEXANDRE ELIAS TRIVELLATO, LUCIANA YAMAMOTO DE ALMEIDA, HEITOR ALBERGONI DA SILVEIRA, and, JORGE ESQUICHE LEÓN

Calcifying odontogenic cyst (COC) is a rare odontogenic lesion derived from the remaining odontogenic epithelium of the jaws. Radiographically, it is a destructive lesion, and the cortical plates of bone are thin and expanded. Here, we report a case series of 3 patients diagnosed with central COC. Microscopic and immunohistochemical analyses were performed. Two cases occurred in women, with mean age of 34.3 years. All cases occurred in the maxilla, without symptomatology, presenting as an expansive tumor-like mass. The differential diagnosis included residual cyst, ameloblastoma, Pindborg tumor, adenomatoid odontogenic tumor, odontogenic fibroma, developing odontoma, and fibro-osseous lesion. Immunohistochemistry supports an odontogenic origin with low proliferative index. COC is an uncommon lesion that should be considered by clinicians in the differential diagnosis of odontogenic cysts or tumors, especially when mineralized deposits are detected on imaginologic exams. Intraosseous COC is mainly treated by enucleation, with rare or low rate of recurrence.

20190026

FIBROUS DYSPLASIA AFFECTING MAXILLARY SINUS AND ZYGOMA: A CASE REPORT

FERNANDA MARCELLO SCOTTI, GILBERTO MELO, LUIZ HENRIQUE GODOI MAROLA, MURILLO CHIARELLI, and, ROGÉRIO GONDAK