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ORAL PRESENTATION

OP - AMELOGENESIS IMPERFECTA AND JALILI SYNDROME: A CASE REPORT. *HERCÍLIO MARTELLI, JÚNIOR, RICARDO D. COLETTA, VERÔNICA OLIVEIRA DIAS, CÉLIA FENANDES MAIA, DANIELLA R. BARBOSA MARTELLI, LUCIANO S. NÁSSER. STATE UNIVERSITY OF MONTES CLAROS, UNIMONTES / STATE UNIVERSITY OF CAMPINAS, FOP-UNICAMP.*

Jalili syndrome (OMIM #217080), caused by mutations in the CNNM4 gene, is a recessively inherited disorder characterized by combination of amelogenesis imperfecta (AI) and cone rod dystrophy. This study reports an 8-year-old girl with a diagnosis of congenital loss of vision (Lebeŕs congenital amaurosis) and yellowish, brittle and painless teeth. Ophthalmological examination confirmed the child's visual impairment, which was caused by cone rod dystrophy of retina. Intra oral examination revealed yellow, stained and misshapen teeth, confirmed the diagnosis of generalized AI. Radiographic examination showed incomplete permanent dentition with the delayed eruption. The presence of congenital cone rod dystrophy in association with generalized AI leaded to the diagnosis of Jalili syndrome. The parents received genetic counseling and comprehensive dental treatment of patient. We emphasize the importance of dentist in the diagnosis of this rare syndrome. Acknowledgments Fundação de Amparo a Pesquisa do Estado de Minas Gerais - FAPEMIG, Belo Horizonte, Brazil and National Council for Scientific and Technological Development-CNPq, Brazil.

OP - CONGENITAL LIPOMATOSIS OF THE FACE: A RARE DISEASE. *MARINA DE OLIVEIRA VERÍSSIMO, OPHIR RIBEIRO, JUNIOR, SERGIO ADAMOLI, CLAUDIA PEREZ TRINDADE FRAGA, DANIEL HACOMAR DOS SANTOS, ROSANA MASTROFRANCISCO, MARCELO MARCUCCI. HOSPITAL HELIÓPOLIS.*

Facial congenital infiltrating lipomatosis is a rare subtype of hemifacial hyperplasia. Abnormal growth of one side of the face occurs in all tissues of the affected area, particularly adipose tissue. Diagnosis is by exclusion, as the etiology of this condition is unknown. However, some factors associated with it include heredity, chromosomal and neural abnormalities, atypical twinning forms, altered intrauterine environment, endocrine and anatomical dysfunction, functional abnormalities of the vascular and lymphatic systems, and central nervous system disorders. Herein, we report a 15-year-old girl who sought treatment in the Hospital Heliópolis of São Paulo for congenital facial swelling that partially regressed over time. Extraoral examination revealed facial asymmetry with normal color, soft consistency, and non-pulsatile swelling in the left mandibular region. Intraoral examination showed a slight erythematous tongue and soft palate on the left side. Magnetic resonance imaging showed thickening and signal heterogeneity in the fat planes of the left perimandibular and perimaxillary regions, with thread-like vascular structures in between. Magnetic resonance angiography revealed an increase in the left submandibular and parotid glands. Based on her medical history, physical examination results, and imaging studies, the patient was diagnosed with congenital lipomatosis of the face.

OP - CALCIFYING CYSTIC ODONTOGENIC TUMOR WITH AN ADENOMATOID COMPONENT: A RARE HYBRID ODONTOGENIC TUMOR. *SIMONE CRISTINA LEAL TOSTA DOS SANTOS, LEONARDO MORAIS GODOY FIGUEIREDO, BRAULIO CARNEIRO, JUNIOR, JEAN NUNES DOS SANTOS, AGUIDA CRISTINA GOMES HENRIQUES. FEDERAL UNIVERSITY OF BAHIA.*

Odontogenic tumors (OTs) represent a heterogeneous group of entities characterized by differences in their biological behaviors and the occurrence of several inductive interactions. Calcifying cystic odontogenic tumors (CCOTs) and adenomatoid odontogenic tumors (AOTs) are well-recognized examples that display a broad spectrum of clinical and histopathological features. CCOTs may occur in association with other OTs, whereas hybrid OTs involving exclusively CCOT and AOT are rare, with only 4 reported cases in the English literature. In this report, we describe the diagnosis and treatment of a patient with hybrid OT. A 60-year-old African American man presented with considerable swelling in the anterior buccal region of the mandible. Panoramic radiography revealed a well-defined, unilocular, radiolucent lesion associated with important root resorption. Excisional biopsy was performed with complete enucleation of the lesion. Microscopic examination revealed a cystic lesion lined with ameloblastomatous epithelium and ghost cell formations. Duct-like structures and extensive areas of dentin-like material were noted. The patient remains recurrence-free 6 months after surgery. Hybrid CCOT/AOT, although rare, should be included in the differential diagnosis of jaw lesions. Despite their unknown pathogenesis, such lesions should be treated conservatively. Keywords: odontogenic tumors, adenomatoid odontogenic tumor, calcifying cystic odontogenic tumor.

OP - SOLITARY FIBROUS TUMOR IN THE FLOOR OF THE MOUTH IN A PATIENT PREVIOUSLY AFFECTED BY A PLEOMORPHIC ADENOMA. *DÉBORA LIMA PEREIRA, ÁGUIDA MARIA MENEZES AGUIAR MIRANDA, CAIO LOVISI, HENRIQUE MARTINS DA SILVEIRA, FÁBIO RAMÔA PIRES. STATE UNIVERSITY OF RIO DE JANEIRO AND ESTÁCIO DE SÁ UNIVERSITY, RIO DE JANEIRO, BRAZIL.*

A solitary fibrous tumor (SFT) is a benign neoplasm derived from pluripotential mesenchymal cells. Although described as a pleural tumor, SFTs affecting extrapleural locations have been reported, but involvement of the oral cavity is rare. A 37-year-old woman was referred for evaluation of a painful swelling in the mouth. Her medical history revealed surgical removal of a pleomorphic adenoma on the left submandibular gland 12 years earlier. Intraoral examination showed a firm, tender to palpation, submucosal swelling on the left side of the floor of the mouth.