

fetuses done by Stout and Collet in 1969 found evidence of two cystic lesions associated with MAAC. These cysts were named median alveolar cyst (MAC). To the best of our knowledge, we are reporting for the first time a bona fide example of MAAC - MAC in a human being.

**Case report:** A healthy 14-year-old Saudi female with an anterior maxillary diastema was referred to the orthodontics clinic for consultation. Clinical examination revealed a double frenum connecting the maxillary lip and alveolar vestibule. A panoramic film and a cone beam CT revealed a radiolucency between the maxillary central incisors extending from the alveolar crest to the incisive foramen area. The labial cortical plate was missing while the palatal was intact. The radiologist interpretation was "enlarged nasopalatine canal". No other physical or dental abnormalities were evident. Upon surgical exploration, no labial maxillary osseous plate was found however, soft tissue was present and excised. Microscopic examination of the excised tissue revealed a cystic process lined by acanthotic nonkeratinizing stratified squamous epithelium with intracellular edema. In addition, sebaceous glands, islands of squamous epithelium with keratin pearl formation and lymphoid infiltrates were seen within the cystic wall. A retrospective review of the imaging studies coupled to the microscopic findings resulted in diagnosis of median alveolar cyst associated with a median maxillary anterior cleft.

**Conclusion:** We report a rare case of MAAC with MAC showing a sebaceous component. It is thought that MAC most likely originates from epithelial invaginations derived from the anterior intermaxillary suture. However, the mechanism involved in the formation of these two conditions remains to be elucidated.

#### IDENTIFICATION OF NOVEL COPY NUMBER ALTERATIONS IN AMELOBLASTOMA AND AMELOBLASTIC CARCINOMA FROM NIGERIA.

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**Background and Objectives:** Ameloblastoma is a benign odontogenic neoplasm, characterized by local invasiveness, facial deformity, tooth displacement, a high rate of recurrence, and malignant transformation. It accounts for 63% of odontogenic tumour in Nigeria. Recently, studies in the genomic landscape of ameloblastoma have identified a number of consistent alterations that may be useful for therapeutic intervention. To date, no whole genome survey of ameloblastoma and ameloblastic carcinoma has been published.

**Methods:** DNA was extracted from RNALater stored tissue using the DNeasy Tissue Kit (QIAGEN), from a cohort of ten ameloblastoma and three ameloblastic carcinoma from UCH, Ibadan, Nigeria. Whole genome analysis was performed using the Oncoscan FFPE Assay Kit (Affymetrix). Data was analysed using Nexus Express for Oncoscan 17.0 and Somatic Mutation Viewer 1.0.1.

**Findings:** Ameloblastoma (n=10) showed a mean genome change of 9.7%, with a mean of 88.7 copy number (CN) aberrations and 7.5% of loss of heterozygosity (LOH), whereas the ameloblastic carcinomas (n=3) had a mean genome change of 6.8% with a mean of 87.3 copy number (CN) aberrations and 3.6% of loss of heterozygosity (LOH). All tumours (benign and malignant) showed CN gain at 8q23.3, affecting the CSMD3 gene. Other commonly affected regions included LOH at 1p34.2-p34.1 and 2q11.2, among others. Ameloblastoma and

ameloblastic carcinomas shared somatic mutations in BRAFV600E, EGFR, KRAS and PTEN genes. One ameloblastoma showed a mutation in TP53 and two (66.7%) ameloblastic carcinomas showed a mutation in the PIK3CA gene, which was not observed in the ameloblastoma cohort.

**Conclusions:** Ameloblastoma and ameloblastic carcinoma do not show extensive genome changes indicative of genomic instability. We have identified novel areas of CN gain and LOH that require further investigation. The mutational profile of these lesions is similar to that reported in the literature. Funding: Pathological Society of Great Britain.

#### THE IMPORTANCE OF IMMUNOHISTOCHEMISTRY AND MOLECULAR STUDIES FOR DIAGNOSING EWING'S SARCOMA OF THE MANDIBLE: A CASE REPORT.. DR. FAISAL ALHEDYAN<sup>A</sup>, DR. FALEH ALSHAHRANI<sup>B</sup>, DR. IBRAHIM O BELLO<sup>C</sup>, DR. RANA ALSHAGROUD<sup>C</sup>. <sup>A</sup> COLLEGE OF DENTISTRY, PRINCE SATTAM BIN ABDULAZIZ UNIVERSITY, ALKHARJ, <sup>B</sup> DEPARTMENT OF ORAL AND MAXILLOFACIAL SURGERY, KING FAHAD MEDICAL CITY, RIYADH, SAUDI ARABIA, <sup>C</sup> COLLEGE OF DENTISTRY, KING SAUD UNIVERSITY, RIYADH

**Introduction:** Ewing's sarcoma (ES) is a malignant small round cell neoplasm primarily affects the bone. It was first described by James Ewing in 1921. ES accounts for 6-10% of all primary malignant bone tumors. It is most commonly found in children between 10-15 years of age. 1% to 2% of cases of ES affect the craniofacial bones. Only a few cases have been reported in the mandible. Here we report a case of EW in the mandible and the use of immuno-histochemistry and molecular studies to confirm its diagnosis.

**Clinical presentation:** A 16 year old female patient was seen at the Department of Oral and Maxillofacial Surgery in King Fahad Medical City. Extra-oral examination revealed diffuse painless swelling on left side of the mandible with reduced mouth opening. Intraorally, an ulcerated large mass was present. CBCT revealed ill-defined radiolucency involving the posterior part of the mandible extending to the ramus. MRI showed a destructive mass in the left mandible with a soft tissue component occupying the left masticator space. PET/CT showed a FDG avid left cervical large mass. An incisional biopsy was taken. Microscopically, the specimen revealed the presence of islands and sheets of monotonous malignant cells infiltrating the bone. The nuclei of the malignant cells were round to oval in shape with fine dispersed chromatin and one or two indistinct nucleoli. The neoplastic cells were positive for CD99 and Fli1 and negative for SATB2. Chromosomal translocation t (11:22) involving the EWS and FLI-1 gene was identified using FISH. Patient was treated with chemotherapy.

**Conclusion:** We reported a case of a malignant tumor with an immunoprofile of Ewing Sarcoma that was confirmed with the identification of chromosomal translocation by molecular study.

#### CONDYLAR HYPERPLASIA: RADIOLOGICAL, HISTOLOGICAL AND IMMUNOHISTOCHEMICAL COMPARATIVE ANALYSIS.

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**Objectives:** the aim of this study was to analyze the radiological parameters and histological features and to evaluate the immunoexpression of bone-related proteins in condylar hyperplasia, comparing to age and gender of the affected patients. Forty specimens derived from the surgical treatment of condylar hyperplasia were selected and clinical data were retrieved from the patient files. Radiological information was obtained from panoramic radiographs comparing the affected to non-affected sides. All cases were histologically reviewed and immunoreactions against TRAP, PTHrp, podoplanin and RANK were performed in all cases by the immunoperoxidase technique. Data were descriptively and statistically analyzed comparing both gender and age of the patients with radiological, histological and immunohistochemical features.

**Findings:** radiological parameters did not correlate with age and gender of the patients; proportion of bone tissue:bone marrow and fibrous:cartilage layers showed no differences when comparing age of the affected patients, but gender of the patients correlated with the latter. There were no differences on TRAP, PTHrp, podoplanin and RANK expression according with gender and age of the affected patients.

**Conclusions:** there were no differences on radiological parameters, proportion of bone tissue:bone marrow and fibrous:cartilage layers and expression of bone-related proteins according with age and gender of patients affected by condylar hyperplasia. Females presented a higher proportion of fibrous:cartilage layers than males.

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#### BILATERAL ORTHOKERATINIZED ODONTOGENIC CYSTS OF THE MANDIBLE. CASE REPORT AND REVIEW OF THE LITERATURE.

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**Introduction:** First described by Wright in 1981, orthokeratinized odontogenic cyst (OOC) represents a developmental condition derived from epithelial dental lamina rests. Radiographically, OOC appears as a unilocular radiolucency associated with an impacted mandibular third molar. Occasional examples of bilateral/multicentric OOCs have been reported in the literature. Here, we present the clinical, radiographic and microscopic features of a patient with bilateral mandibular OOCs.

**Case report:** A healthy 19-year-old male presented with asymptomatic unilocular radiolucencies associated with left and right impacted mandibular 3rd molars. Both molars were extracted and the associated lesions enucleated. Microscopic examination of both specimens showed identical microscopic features consisting of cystic cavities lined by orthokeratinized stratified squamous epithelium with hypergranulosis. A diagnosis of bilateral OOCs was rendered. No recurrence was evident after 4-months.

**Discussion:** Six cases (including ours) of bilateral/multicentric OOCs have been documented in the English and Spanish literature. An analysis of the published demographics of this condition showed that most bilateral/multicentric OOCs have a striking predilection for young adult males (age range: 19-41 years; mean age: 27.3 years) with only one case presenting in a female. Bilateral OOCs are almost exclusively associated with impacted mandibular third molars. However, one report documented

OOCs in all quadrants. The follow-up period for these cases ranged from 4 months-13 years with no recurrence stated.

**Conclusion:** Bilateral OOCs are uncommon and appear to have an excellent prognosis with no recurrence expected. However, more reports with long-term follow-up are needed to draw meaningful conclusions about their biological behavior.

#### COWDEN'S SYNDROME DIAGNOSED BY ORAL LESIONS: CASE REPORT AND REVIEW OF THE LITERATURE. MRS. MAUREEN MARSHALL<sup>A</sup>, MS. DORIS OTERO<sup>A</sup>, MR. SVEN NIKLANDER<sup>B</sup>, MR. RENE MARTINEZ<sup>A</sup>. <sup>A</sup> UNIVERSIDAD ANDRES BELLO, <sup>B</sup> UNIVERSITY OF SHEFFIELD

Cowden's syndrome (CS), also known as multiple hamartoma syndrome, is a rare genodermatosis of autosomal dominant inheritance and variable phenotype. Its origin is a PTEN (phosphatase and tensin homologue) gene mutation, resulting in the development of multiple hamartomatous lesions and an increased risk of malignancy. Clinically, it is characterized by multiple mucocutaneous lesions, including oral and labial papillomatous papules. Oral manifestations in CS are frequent and usually precede the establishment of malignant tumours. Their correct diagnosis may improve early recognition of this entity, leading to an appropriate genetic counselling and close surveillance for the early detection of malignant processes associated with SC.

We report a case of a 58-year-old male patient who was referred to the Oral Pathology Department of Andrés Bello University, Viña del Mar, Chile, with a presumptive diagnosis of "multiple papules" in the oral cavity. Extraoral examination revealed macrocephaly, facial trichilemmomas and acral keratosis. Upon intraoral examination, multiple papillomatous lesions were observed. A biopsy of the oral lesions was taken, which revealed fibro-epithelial hyperplasia. Endoscopy of the upper digestive tract showed acanthosis of the oesophagus and multiple polyps on the antrum of the stomach and duodenum. Thyroid ultrasound showed multinodular goitre. The patient was diagnosed with Cowden's syndrome and has been followed up closely by a multidisciplinary team in order to diagnose any development of malignant tumours.

#### SPREADING OF GLANDULAR MALIGNANCY MIMICKING BONE LESION:

REPORT OF TWO CASES. PROF. ENEIDA VENCIO, MR. DIEGO ARANTES, DR. ALEXANDRE BELLOTTI, PROF. ALINE BATISTA, PROF. REJANE RIBEIRO-ROTTA, PROF. ROBSON GARCIA. FEDERAL UNIVERSITY OF GOIÁS

Peripheral nerves are target for local invasion and spreading in pancreatic, gastric, prostate, and head and neck cancers. Adenoid cystic carcinoma (ACC) accounts less than 10% of salivary gland neoplasms with dual cell population, typically exhibiting three architectural patterns. Distant metastasis and neural involvement are common clinical features.

**Objectives:** To report two rare cases of ACC arising from parotid gland and extending into mandible through mandibular foramen.

**Results:** A 50-year-old woman and 49-year-old man presented with pain and paresthesia in the left face. A swelling was observed and computed tomography detected osteolytic lesion with irregular margins involving complete body and ramus in the left side of the mandible. Clinical diagnosis was established of osteomyelitis and sarcoma. Microscopically, both