mucoepidermoid carcinoma, and no review of advanced imaging characteristics. The purpose of this study was to present the common diagnostic imaging characteristics of intraosseous mucoepidermoid carcinoma seen in plain radiography and computed tomography (CT). Two observers reviewed and recorded the imaging characteristics seen in plain films and 3 complete CT studies of 4 cases of histologically confirmed intraosseous mucoepidermoid carcinoma. In addition, in 1 case the histopathologic features displayed in a whole section through the resected specimen correlated with the radiographic features. The following 5 imaging features were found to be common to all cases: a well defined sclerotic periphery, the presence of amorphous sclerotic bone within the lesion, numerous loculations (majority <8 mm in diameter), lack of septae bordering some of the loculations, and expansion and perforation of the outer cortical plate with extension into surrounding soft tissues. Other characteristics included tooth displacement and root resorption. Some of these characteristics are shared with both desmoplastic ameloblastoma and glandular odontogenic cyst; similarities and differences are presented.


We report herein our experience with 39 basal cell adenocarcinomas (BCACs; 1993-2010) to further define the clinicopathologic features of this rare salivary gland malignancy. The mean age at presentation was 57.6 years, with no gender predilection. The most common sites were parotid gland (82.1%), submandibular gland (5.1%), and upper lip (5.1%); 22.8% of patients had a synchronous or asynchronous salivary gland tumor. One of these patients had Brooke-Spiegler syndrome. Four histologic subtypes were identified: membranous (41.0%), tubulotrabecular (23.1%), solid (20.5%), and cribriform (15.4%).

**THE INCREASED MUC1 EXPRESSION IN RECURRENCE AND MALIGNIZATION OF SALIVARY GLAND PLEOMORPHIC ADENOMA.** A.B. Soares, V.C. de Araujo, A. Altemani. São Leopoldo Mandic Institute and Research Center, State U Campinas, Brazil.

Background. Pleomorphic adenoma (PA) is the most common salivary gland tumor. Although classified as benign, it presents a tendency to recur (RPA) and a risk of malignant transformation. It has been suggested that MUC-1 plays a role in the progression of many tumors, and in the salivary gland it was indicated as a marker to predict RPA. The aim of this study was to evaluate the MUC-1 expression in different phases of the adenoma-to-carcinoma sequence.

**Study design.** Twenty-one cases of PA, 18 cases of RPA, 3 cases of RPA with focal transformation, and 11 cases of carcinoma ex pleomorphic adenoma were analyzed for MUC1 expression by immunohistochemical technique using the antibodies MUC1/DF3.

**Results.** There was a significant difference in MUC1 expression in all of the groups. Comparing nonrecurrent with recurrent tumors, MUC1 reactivity in RPA was stronger than in PA. In all of the different groups of carcinoma, MUC-1 expression was significantly higher in carcinoma than in RPA and PA.

**Conclusion.** This study shows that carcinoma cells overexpress MUC1 and that this molecule is associated with the malignant transformation of this tumor. In addition, this research confirmed that MUC1 is related to the recurrence of PA.


**Background.** Salivary gland tumors comprise a group of tumors with a complex and unclear process of tumorigenesis. Previously we have shown a significant overexpression of Akt and Mdm2 in these tumors. Akt coordinates many proteins within the cell. PTEN and NFKB may play important roles in the understanding of the activation of Akt pathway. PTEN negatively regulates the Akt/P3K pathway, which is important for cell growth, proliferation, and survival. NFKB activation is responsible for direct transcription of over 180 known Nfkβ target genes involved with survival and proliferation.

**Objective.** The aim of this study was to analyze the expression of PTEN and NFKB proteins in salivary gland tumors. Thirty-eight cases of adenoid cystic carcinoma (ACC), 45 of pleomorphic adenoma (PA), and 13 of carcinoma ex pleomorphic adenoma (EXPA) were submitted to immunohistochemistry. Normal salivary gland (NSG) was used as control.

**Results.** Strong cytoplasmic and negative nuclear staining of NFKB in almost all tumors was seen. Eight samples of PA and 1 EXPA showed low rates of nuclear staining. NSG showed variable cytoplasmic expression rates. PTEN stained the cytoplasm of all tumors and NSGs. Also, high rates of nuclear PTEN in all ACC and low rates in 3 EXPA were seen. NSG showed negative nuclear expression. Nuclear PTEN may be associated with other proteins blocking or inactivating tumor suppressors and its own function; therefore, in ACC PTEN may indicate an unexpected activity toward the classic PTEN pathway, and PTEN protein may also be associated with other proteins inactivating tumor suppressors.

**Conclusion.** This study strongly suggests that PTEN may be involved in tumor progression of ACC. On the other hand, cytoplasmic expression of Nfkβ is probably not significant due to the similarity with NSG.

Salivary gland cystadenocarcinomas with both mucinous and papillary components have been rarely described in the literature. Tumors with these features have no uniform nomenclature and present with variable clinical behavior. We report 2 additional cases of mucus-producing papillary cystadenocarcinomas, focusing on their histologic and immunohistochemical features. Case 1 presented as a nodular deeply situated mass in the posterior floor of the mouth of a 59-year-old woman and case 2 as a submucosal nodule in the floor of the mouth of a 54-year-old woman. Both tumors exhibited the same histologic features, including multiple cystic spaces filled with mucus, lined by clear mucus-producing epithelial cells, forming intraluminal papillary projections supported by a thin fibrovascular connective tissue core. Some columnar epithelial cells had atypical morphology, and mitotic figures were rarely encountered. Luminal content was positive for periodic acid–Schiff and mucicarmine staining. The neoplastic epithelial cells were strongly immunopositive for cytokeratin (CK) 7, 8, 18, and 19 and negative for CK 20. The Ki-67 labeling index was 10%, and p53 staining was <5%. Both patients were submitted to wide surgical excision, and there were no signs of recurrence after 1 year in case 1 and 6 months in case 2. Large series of cystadenocarcinomas presenting both mucinous and papillary components are necessary to better understand their immunophenotype, diagnostic features, and biologic behavior.

EXTRASKELETAL MYXOID CHONDROSARCOMA IN A PEDIATRIC PATIENT. M. Romaitach, J. León, O. Almeida, M. Nuyens, R. Carlos. U Campinas, Piracicaba, Brazil; Centro Clínico de Cabeza y Cuello/Hospital Herrera Llerandi, Guatemala City, Guatemala.

Extraskeletal myxoid chondrosarcoma (EMC) is a rare malignant soft-tissue tumor that mainly affects the thighs of men in their sixth decade of life. It has been rarely described in the head and neck area, especially in children and adolescents. We present a case of EMC affecting the infratemporal space of a 13-year-old boy presenting a large painless diffuse mass in his right parotid region lasting 6 months. T2-weighted magnetic resonance imaging exhibited a lesion with high signal and lobular configuration. Histologic evaluation of the specimen obtained by incisional biopsy revealed multiple nodules containing tumoral cells separated by several fibrous septae. The tumoral cells presented vacuolated granular cytoplasm and round nuclei and were supported by an abundant myxoid pale stroma. Periodic acid–Schiff staining with and without prior diastase digestion demonstrated cytoplasmic glycogen in the tumoral cells. Immunohistochemical features of the tumoral cells included positivity for vimentin, neuron-specific enolase, and chromogranin, whereas the tumor was negative for pan-cytokeratin AE1/AE3, epithelial membrane antigen, S100, desmin, muscle actin–specific HHF35, CD57, glucose transport protein 1, and synaptophysin. The Ki-67 labeling index was 42%. The patient was treated by surgical excision and adjuvant radiotherapy but died after 1 year owing to complications of local tumor dissemination.


Background. Myofibroblastic sarcoma (MS) represents a distinct malignant mesenchymal neoplasm composed of myofibroblasts and is different from fibrosarcoma and leiomyosarcoma. MS may arise in soft tissue or bone in adults or children. There is a predilection for the head and neck region. Most MSs are low grade, mimicking of nodular fasciitis, and possibly inflammatory myofibroblastic tumors; a less differentiated high-grade variant exists. High-grade MS is hypercellular, has less collagen production, may exhibit necrosis, and demonstrates hyperchromasia and increased mitotic activity. Marked pleomorphism and multinucleated giant cells that characterize high-grade pleomorphic sarcoma are lacking. The cells of MS express smooth muscle actin and calponin and lack b-caldesmon. MS lacks specific cytogenetic abnormalities as identified in infantile fibrosarcoma t(12;15) and inflammatory myofibroblastic tumor (rearrangement in the ALK gene region). Cytogenetics of MS have demonstrated noncharacteristic chromosomal aberrations with a simpler karyotype than reported with high-grade pleomorphic sarcomas.

Case report. An 86-year-old man presented with a large ulcerated mass of the right posterior palate, with mobile maxillary molars and, upon biopsy, underlying necrotic-appearing bone. Histopathologic findings and immunohistochemical phenotype were interpreted as high-grade MS. Work-up of the patient revealed extensive disease involving multiple sites.

Conclusion. A high-grade MS with an aggressive clinical course is presented. MS is a distinct lesion with defined immunophenotypic features. Low-grade lesions may mimic reactive or benign processes, and high-grade lesions need to be differentiated from other similar-appearing spindle cell sarcomas.


Patients with a history of treated retinoblastoma (RB) have a greatly increased risk of a broad spectrum of secondary malignancies appearing many years later, with a high incidence in the head and neck region. Leiomyosarcomas (LMSs) account for ~20% of these tumors. LMSs in the sinonasal region generally are associated with a locally aggressive course and have a poor prognosis. We report an unusual case of LMS of the nasal sinus area in a 35-year-old African-American man with a history of unilateral RB and radiation therapy. RB may occur in 2 forms. The hereditary form is generally bilateral but can present as unilateral with a positive family history and typically exhibits a germline mutation in the RB1 gene on chromosome 13. The nonhereditary form is usually unilateral but can present as unilateral with a positive family history and typically exhibits a germline mutation in the RB1 gene on chromosome. The nonhereditary form is usually unilateral but can show the same germline mutation in up to 10% of cases. Patients with hereditary RB have a tenfold higher cumulative risk of developing secondary malignancies than those with the nonhereditary form. Most reported cases of sinonasal LMS are in patients with a history of the bilateral hereditary form of treated RB. To the best of our knowledge, this is the second reported case of sinonasal LMS arising in a patient with a history of unilateral RB. The clinical history, radiology, and pathology are presented along with a brief discussion of the literature.

GENITAL EWING SARCOMA OF THE HEAD AND NECK WITH NOVEL EWS-NFATC2. C. Flaitz, J. Hicks. U Texas Dental Branch and Baylor College of Medicine, Houston.

Background. Congenital small round cell tumors (SRCT) are typically leukemias and neuroblastomas. Congenital Ewing sar-