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 mucous-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. Nuyen, R. Carlos, U Campinas, Piracicaba, Brazil, Centro Clínico de Cabeza y Cuello/Hospital Herrera Llerandi, Guatemala City, Guatemala.

Extraskelatal 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 septate. 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 glycosaminoglycans 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 h-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 has 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 molar 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 a 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-
coma (ES) is very rare and may be misdiagnosed as another SRCT.

Case report. A 37 weeks’ gestational age male was delivered vaginally. An 8 × 7 cm blue to purple mass protruded from the left facial/ear region that was not detected by ultrasound at 20 weeks’ gestation. Diagnostic imaging and clinical impression were interpreted as a vascular tumor, and prednisone and lansoprazole therapy was initiated. The tumor rapidly increased in size and ulcerated; therefore, resection was performed on the eighth day.

Pathology. The tumor was composed of sheets of undifferentiated cells with scant cytoplasm, frequent mitoses, minimal stroma, and no differentiation. Flow cytometry revealed CD56 (neural cell adhesion molecule) and CD38 positivity with no evidence of hematolymphoid malignancy. Immunocytochemistry exhibited CD56 and dot-like Golgi staining with CD99. Myogenin (myogenin, desmin), neuroblastic (NB84), epithelial (epithelial membrane antigen, cytokeratin), germ cell (alpha-fetoprotein, CD30, placental alkaline phosphatase), vascular (CD34, CD31), rhadoid (integrase interactor 1) and melanocytic (HMB-45, MelanA) markers were negative. Electron microscopy provided evidence for neuroectodermal (ES) derivation with rare neurosecretory granules, bland nuclei, intercellular junctions and lack of neurites. Cyogenetics identified by fluorescence in situ hybridization EWS rearrangement and novel EWS-NFATc2 translocation (t[20;22]). Despite therapy, the child died of disease 4 months after protocol initiation.

Conclusion. Multimodal diagnostics, including cyogenetic and molecular analyses, are important for definitive diagnosis, proper treatment, and discovering novel translocations in neonaes with SRCT.

ORAL WARTS IN HIV-INFECTED INDIVIDUALS EXHIBIT LACK OF TISSUE AUTOFLUORESCENCE: A CLINICOPATHOLOGIC STUDY. C. Flaitz, M. Nichols, N. Vigneswaran, J. Bouquot, A. Zuluaga. U Texas Dental Branch, Bering-Omega Dental Clinic, Remicalm, Houston.

HIV-infected individuals experience an increased prevalence of oral warts. Multiple and high-risk human papilloma virus (HPV) genotypes have been identified in these lesions, which suggests a potential risk for HPV-associated squamous cell carcinoma. The purpose of this clinical study was to evaluate HPV-induced oral lesions in HIV-infected individuals, using direct autofluorescence visualization, and to correlate the results with histopathologic findings. Consecutive HIV patients from the Bering-Omega Dental Clinic with oral warts and hyperkeratotic lesions were recruited. After informed consent, patient demographics, CD4 count, viral load, medications, and oral cancer risk factors were recorded. Clinical features and location of lesions were evaluated and photographed under white light, violet light–excited autofluorescence (405 nm), and green-amber light reflectance (540-560 nm), using Identafi 3000 (Trimira, Houston, TX). Surgical specimens were submitted for routine microscopic diagnosis. Immunoperoxidase (IMP) studies using HPV Cocktail Broad Spectrum (Biocare Medical, Concord CA) were performed. Twenty-nine patients had HPV lesions and/or squamous cell carcinoma (25 male, 4 female; mean age 47 years; mean CD4 count 339). Four patients were in the control group (3 male, 1 female; mean age 45 years; mean CD4 count 163). In the HPV group, tobacco use was reported in 55% and alcohol use was 38%; the control group reported 100% tobacco and 50% alcohol use. IMP for HPV was positive in wart but not in control subjects. Loss of fluorescence (LOF) was 85% for oral warts, 9% equivocal, and 6% no loss. LOF for the control group was 25% with positive vascular reflectance for lichenoid mucositis. In this pilot study using a multispectral oral examination light, oral warts consistently demonstrated LOF in this high-risk oral cancer group.


Background. Human papilloma virus (HPV) infection has been recently identified as an important etiologic factor in head and neck (HN) pathology, with important treatment and prognostic implications because HPV-positive tumors affect a younger nonsmoking population and have a distinctly better survival after treatment than the HPV-negative cohort.

Study design. The identification and genotyping of high-risk (16, 18, 31, 33, 35, 39, 42-45, 51, 52, 56, 59, and 70) and low-risk (6 and 11) HPV was investigated in 18 HN cancers and precancerous lesions—8 invasive tonsilar squamous cell carcinomas (5 moderately differentiated [MDSCC] and 3 poorly differentiated [PDSCC]), 5 laryngeal tumors (4 squamous papillomas, 1 of which exhibited moderate dysplasia and 1 MDSCC), 2 well differentiated SCC (WDSCC) involving the left arytenoid and fossa of Rosenmuller, respectively, and 1 invasive PDSCC of the soft tissues of the neck of unknown primary—were investigated real-time polymerase chain reaction (PCR). Two invasive WDSCC of the dorsal tongue and the floor of the mouth and 1 lesion of the lateral tongue showed keratinosis with moderate dysplasia were also included.

Results. Of all lesions, 61.1% (11/18) were HPV-16 positive and none demonstrated low-risk HPV subtypes; 5/6 MDSCC were HPV-16 positive and 1 MDSCC was inconclusive; 3/4 PDSCC demonstrated the HPV-16–positive subtype. Neither the oral lesions nor the tumors of the arytenoid and fossa of Rosenmuller showed HPV presence. All laryngeal papillomas were positive for HPV-16.

Conclusions. Despite the small sample size, this study further confirms the detection of high-risk HPV in oropharyngeal carcinoma and laryngeal papilloma. The absence of HPV in oral malignant and precancerous lesions also delineates the limited contribution of HPV to the development of these lesions. These findings should also prompt the clinicians to investigate the presence of HPV in MDSCC and PDSCC, with less emphasis on WDSCC, via real-time PCR.


Primary intraosseous carcinoma (PIOC) is a rare malignancy of the jaws arising from the remnants of residual odontogenic epithelium. PIOC is most commonly seen in the posterior mandible and has a strong male predilection. The incidence of PIOC is extremely low, with <50 cases reported in the literature. We report an additional case of PIOC in the mandible of a 73-year-old patient. The lesion was initially discovered as an incidental