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 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 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 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. Faitz, 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-