Surgical Pathology

311

Id: EP32

Synchronous Undifferentiated and Medullary Carcinomas of the Colon; Recurrence of Undifferentiated Colonic Carcinoma in the Peripancreatic Retroperitoneum

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In the World Health Organization 2010 classification, undifferentiated carcinoma of the colon has been subclassified separately from medullary carcinoma of the colon. It is a rare colorectal malignancy with inconsistent histological features and lack of morphological, immunohistochemical, and molecular evidence of differentiation. Some of these tumors have microsatellite instability (MSI). The tumor has infiltrative borders and generally poor prognosis. Medullary carcinoma is a rare variant of adenocarcinoma and is composed of sheets, nests, and trabeculae with uniform small to medium-sized cells, vesicular nuclei, and prominent nucleoli and abundant eosinophilic cytoplasm with no to rare gland formation, prominent lymphocytic infiltrate, and pushing borders. It is strongly associated with MSI, has a good prognosis, and may be associated with Lynch syndrome. In addition, some authors have proposed a possible link between inflammatory bowel disease and Lynch syndrome. We report a unique case of a 34-year-old man with a 10-year history of ulcerative colitis, who presented with abdominal pain and fever. Colonoscopy and biopsy revealed severe pancolitis and multiple masses. Undifferentiated and medullary carcinomas were present in the subtotal colectomy specimen, showing CK7 immunoreactivity and progressive loss of CK20 and CDX2 from an adjacent adenoma to the underlying medullary carcinoma. A peripancreatic fine needle aspiration was morphologically and immunohistochemically similar to the colonic primary. Mismatch repair analysis revealed loss of MLH1/PMS2, and MSI testing was MSI-high (MSI-H) in all 5 loci tested. BRAF V600E mutational testing was negative. Mutational analysis for MLH1 was negative for a known mutation, and the patient has no known familial history of cancer. This case highlights the importance of recognizing the possibility of CK7-positive, CK20- and CDX2-negative colorectal carcinomas occurring in association with MSI-H ulcerative colitis-related neoplasia.

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Am J Clin Pathol 2015;144:A311