A Rare Case Of Synchronous Colorectal Adenocarcinoma Associated With Transitional Carcinoma Of The Left Distal Ureter

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Citation

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Abstract

Background: Transitional cell carcinoma of the ureter most commonly occurs in the lower third. The papillary form is the most common cell type. Familial adenomatous polyposis most commonly affects the colon and rectum. Malignant transformation is highly probable.

Case Report: The patient was a 60-year old male with constipation and bleeding per rectum. There was a family history of adenocarcinoma colon — his father had died from it and his youngest brother is suffering from it. Imaging revealed left hydronephrosis with a tumor in the distal left ureter. Colonoscopy showed tumors in the rectum and hepatic flexure with isolated polyps in the other parts. Laparoscopic total proctocolectomy with ileal pouch and diverting ileostomy was performed for him.

Discussion: This case is being reported for it's rare and unusual presentation. There are 3 possibilities — both malignancies were part of a syndrome; 2 malignancies occurring simultaneously (carcinoma colon in familial adenomatous polyposis with ureteric transitional cell carcinoma) or transitional cell carcinoma metastasizing to the colon. Based on the histopathology findings, metastasis was ruled out — so the possibility of Cowden's syndrome or familial adenomatous polyposis associated with synchronous ureteric transitional cell carcinoma was thought of.

BACKGROUND

The incidence of ureteric TCC, based on combined autopsy reports, is 1 per 1000-3600. The lower third is usually most commonly affected (70%).[1] Exposure to a large variety of noxious stimuli like chemical carcinogens play a major role in the etiology of TCC. The underlying pathophysiology is thought to involve hyperplastic metaplastic changes secondary to chronic irritation, particularly within the ureter. Hematuria (80%), dysuria and frequency are more commonly reported symptoms in ureteral tumors. Pain is usually dull and colicky and results from obstruction or from direct tumor extension. Approximately 90% of TCCs are curable in patients with superficial, confined tumors. Metastases occur in approximately 11% of cases, colon being a rare site.[2] The conventional and standard technique in the treatment of TCC is a nephroureterectomy with either a standard surgical technique or a laparoscopic approach. Familial adenomatous polyposis (FAP) is an inherited condition (defect in APC gene at 5q21, autosomal dominant trait) primarily affecting the colon and rectum. FAP affects about 1 in 10,000 people. Many polyps, sometimes hundreds, develop on the inner lining of this part of the bowel and eventually become malignant if not treated. These polyps commonly develop just after puberty. Approximately half of all patients will have polyps by age 14; 90% will have detectable polyps by age 25. By age 35 to 40, one or more of these polyps will become cancerous.[3]

CASE REPORT

The patient was a 60-year old male with complaints of right-sided abdominal pain and discomfort for 1 month. He also had constipation and bleeding per rectum. There were no urinary symptoms. Per rectal examination revealed a malignant growth 5cm from the anal verge. Colonoscopy showed a few polyps scattered all over the colon with malignant lesions in the rectum and hepatic flexure. Biopsy from the polyp was non-neoplastic and from the tumor was moderately differentiated adenocarcinoma. CT scan of the abdomen showed a tumor in the lower third of left ureter

with left hydronephrosis. The urologist was consulted and biopsy from the ureteric tumor showed low-grade TCC. A diagnosis of Familial Adenomatous Polyposis with TCC of left ureter was made. Total proctocolectomy with ileal pouch combined with left nephroureterectomy was performed for him. A protective proximal ileostomy was also done. There was a family history of similar disease – patient's father had died of carcinoma rectum. The patient is the eldest of 7 brothers. All of them were screened and the youngest brother was found to have carcinoma of the rectum with isolated polyps in the right colon. He underwent laparoscopic total proctocolectomy with ileal pouch and diverting ileostomy as well.

RESULTS

The patient had an uneventful postoperative recovery. He was discharged on the 10th postoperative day. Chemotherapy was planned after 3 weeks. Total proctocolectomy specimen showed multicenteric mucoid adenocarcinoma of the hepatic flexure and rectum (figure 1-arrows) – moderately differentiated grade and a tubulovillous adenoma with moderate degree of dysplasia. Figure 2 shows the rectal tumor with polyp and figure 3 shows the hepatic flexure tumor. Nephroureterectomy specimen showed high-grade papillary TCC of lower third of the ureter (figure 4).

Figure 1

Figure 1: total proctocolectomy specimen

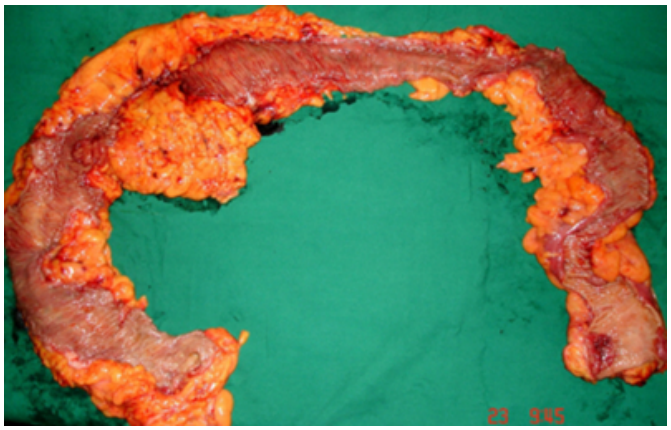


Figure 2

Figure 2: rectal tumor

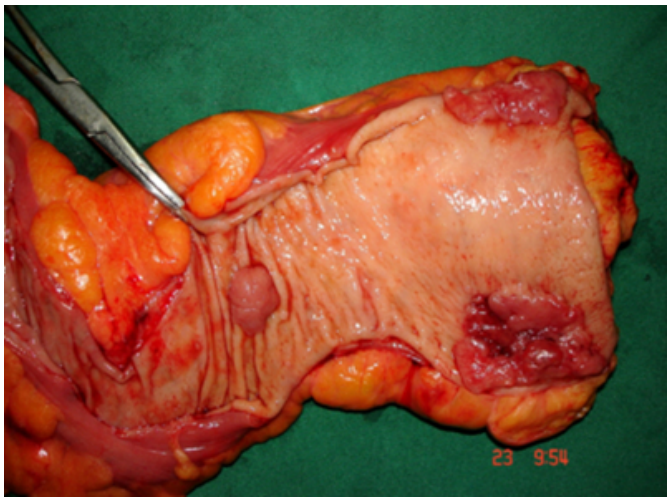


Figure 3

Figure 3: hepatic flexure tumor

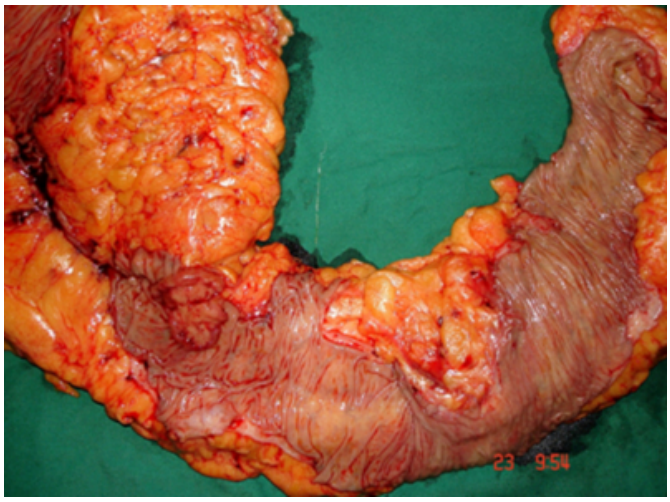
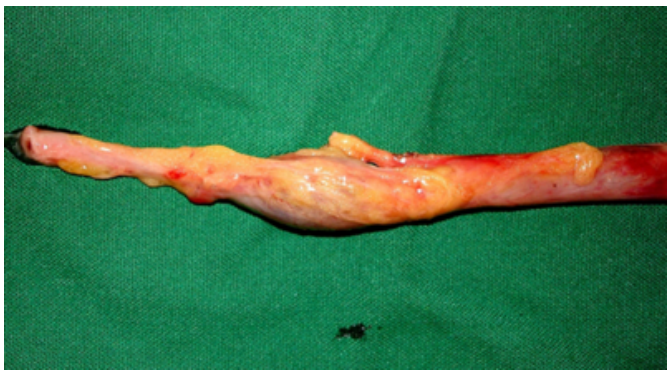


Figure 4

Figure 4: distal ureteric tumor



DISCUSSION

This case is being reported for it's unusual presentation. This type of two primaries arising from different organs is always rare. We attempted to correlate them. There could be three possibilities - TCC metastasizing to the colon, FAP involving the ureter or two completely different primaries. Histology proved that they were indeed different primaries. FAP is not known to be associated with TCC of the ureter or vice versa. Patients with FAP may also develop other malignant and nonmalignant tumors and/or some bone, skin and dental abnormalities. Since the goal is to prevent cancer, the colectomy is done as soon as polyps are discovered. Individuals with FAP are also at increased risk for cancers of the duodenum and biliary tract. However, there are certain conditions like Cowden disease (CD) in which more than one primary tumor has been described; gastrointestinal polyps with malignant potential associated with TCC of the urinary bladder is a rare combination. CD is an autosomal dominant condition with variable expression that results most commonly (80%) from a mutation in the PTEN gene on arm 10q.[3] Internationally, only 200 cases have been published, including separate studies of several generations of affected family members.[4] Although Chen et al reported a few cases of adenocarcinoma of the colon in CD patients, the malignant potential of polyps is low.[5] CD is also associated with some cancers: breast cancer (20-36%); thyroid cancer (7%), colon cancer (2 cases), lung, uterus (endometrial), acute myelogenous leukemia, transitional cell carcinoma of the bladder, cervical carcinoma, non-Hodgkin lymphoma and osteosarcoma. At least 40% of CD patients have a minimum of 1 malignant primary tumor, although with long-term follow-up care, this number may be higher.[6] Many of the cancers are curable if detected early. Close follow-up care of these patients is necessary. According to The International Cowden Syndrome Consortium (criteria for diagnosis), our patient does not fit in. Since genetic studies are not available to us, we can only assume that this is a rare case of FAP with a concomitant TCC of the ureter.

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