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MCKITTRICK-WHEELOCK SYNDROME: A RARE CASE REPORT OF ACUTE RENAL FAILURE

EMIL IOAN MOIS[1,2], FLORIN GRAUR[1,2], ROXANA SECHEL[3], NADIM AL-HAJJAR[1,2]

[1]Department of Surgery, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania
[2]Department of Surgery, Regional Institute of Gastroenterology and Hepatology Prof. O. Fodor, Cluj-Napoca, Romania
[3]Department of Pathology, Regional Institute of Gastroenterology and Hepatology Prof. O. Fodor, Cluj-Napoca, Romania

Abstract

Giant tubular-villous adenoma of the rectum can determine secretory diarrhea, associated with a depleting syndrome of prerenal acute renal failure, hyponatremia, hypokalemia and hypoproteinemia. These symptoms are known as the McKittrick-Wheelock syndrome, and there are about 50 cases reported in literature. We present the case of a 59-year-old woman presented to our emergency department with abdominal pain, prerenal azotemia, and electrolyte disturbances with a background of chronic diarrhea, caused by a giant rectal tumor. Conservative therapy initially improved and normalized renal function, and made surgical resection of the tumor possible.

Keywords: tubular-villous adenoma, hydro-electrolytic disturbances, renal failure

Introduction

McKittrick-Wheelock syndrome is caused by a secretory colorectal tumor and is characterized by fluid and electrolyte depletion. Dehydration, mucous diarrhea, symptoms of hyponatremia (lethargy, headache, weakness, nausea, muscle cramps and seizures), and hypokalemia (fatigue, paresthesia, cramps, ileus, vomiting, hypotension, cardiac arrhythmias and electrocardiographic changes) are the major symptoms of the McKittrick-Wheelock syndrome[1-4]. Approximately 50 cases were reported in the literature[5,6], first described in 1954[7,8]. The cornerstone of the management of McKittrick-Wheelock syndrome is surgical resection of the tumor accompanied by fluid and electrolyte replacement. If those were accomplished, the prognosis is usually good. We report a case of this syndrome and analyze the possible mechanisms that determine acute renal failure.

Case report

A 59-year-old woman presented to the emergency room with abdominal pain, oliguria (less than 500 ml/day) with a background of chronic mucous diarrhea (5 to 7 times/day for the last 5 months) and weight loss (5 kg in the last 3 months). The patient is known with diabetes mellitus under ongoing oral treatment (Siofor 1000 mg X 2/24 h). Physical examination revealed dry skin and mucous membranes. Digital rectal examination revealed a rectal tumor with a soft surface occupying the whole luminal circumference of the rectum, 5 cm from the anal margin (AM).

At the time of presentation she was dehydrated with a blood pressure of 90/60 mmHg and sinus tachycardia – pulse 100 beats/min. Biological parameters: mild nitrate retention with serum creatinine 2.99 mg/dL (normal value (NM) 0.5-0.95 mg/dL), urea 145 mg/dL (NM 15-40 mg/dL), Sodium=137 mEq/L (NM 130-145 mEq/L), Potassium=2.5 mEq/L (NM 3.5-5.4 mEq/L).

The renal function recovered (Na=144 mEq/L, K=5.3 mEq/L, Urea=81 mg/dL, Serum creatinine=0.98 mg/dL) after 1 week of rehydration by i.v. isotonic saline and KCL(30 mgEq/L) replacement. Colonoscopy was performed to investigate persistent diarrhea of unknown cause, which revealed a mass extending from the superior

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Address for correspondence: graurf@yahoo.com

rectum to 5 cm from the anal margin with high suspicion of malignancy (Figure 1). Given the conditions we considered the case as a rectal cancer and we performed an endoscopic ultrasonography to stage the neoplasm. The result was a giant rectal tumor, extending from 5 to 15 cm of AM, limited to the mucosal and submucosal layers, with a suspicion of benign pathology.

We decided to perform a sphincter-preserving surgery. Intraoperatively, we found a 20 cm-long rectal mass; a low anterior rectal resection with total mesorectal excision (TME) (Figure 2) with a mechanical end-to-end colo-rectal anastomosis and ileostomy was done. On the 7th postoperative day, the patient developed a low output anastomotic fistula (about 50 ml on the pelvic drainage) and was managed conservatively.

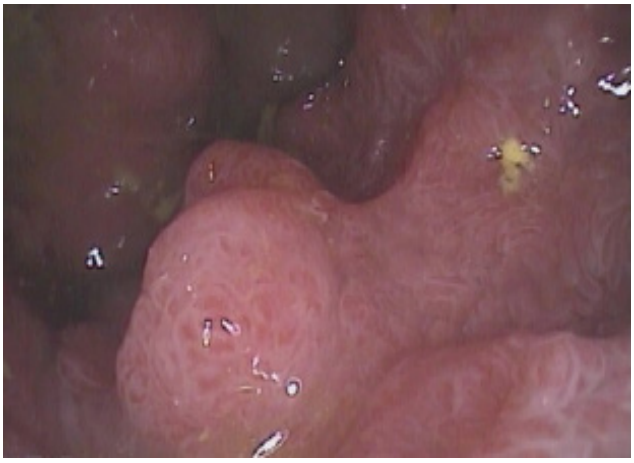

Figure 1. Tumoral mass revealed at colonoscopy.

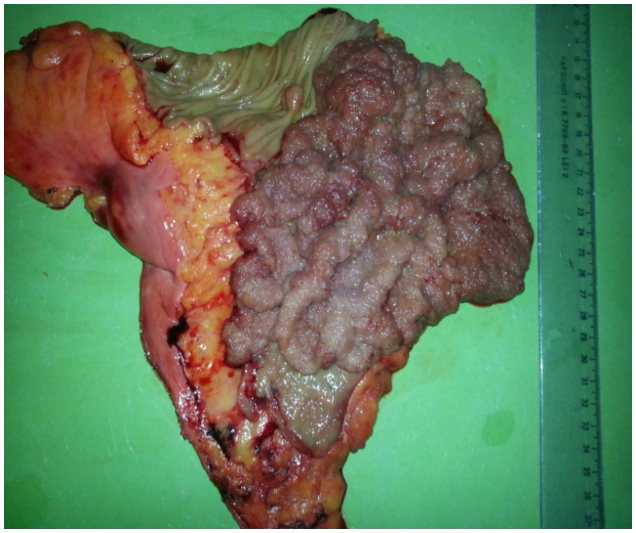

Figure 2. Postoperative specimen.

Histological examination: macroscopically - recto-sigmoidal resection measuring 24 cm long The rectal mucosa is completely covered with soft vegetating lesion, of friable consistency, measuring 14 cm in long axis and 16 cm involving the entire circumference of the rectum. On the cut sections, the proliferation was restricted to the mucosa. Not involving the wall, or adipose tissue. Some diverticular structures were seen. One cm above the tumor, a pediculate polyp was identified, measuring 1.5/1/0.5 cm; microscopically - tubular-villous proliferation which was evenly coating the muscularis mucosa. Aspects of low-grade dysplasia were predominating, with focal high-grade dysplasia, no submucosal layer invasion (Figure 3). Images of mild diffuse inflammatory reaction were found as well, represented by chronic infiltrate in the chorion and submucosa. Surgical margins were free of dysplasia. Twenty-one lymph nodes were examined showing reactive lymphoid hyperplasia. The diverticular structures were uncomplicated and the pediculate polyp showed a synchronous advanced tubular-villous adenoma (low and high-grade focal dysplasia's, Figure 4). Finally, the conclusion is argumentative for a giant rectal tubular-villous adenoma.

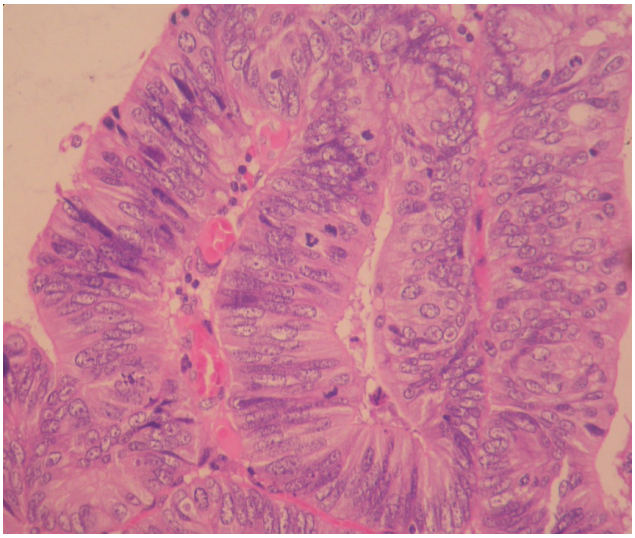

Figure 3. Adenomatous proliferation limited to the mucosa; HE 40X.

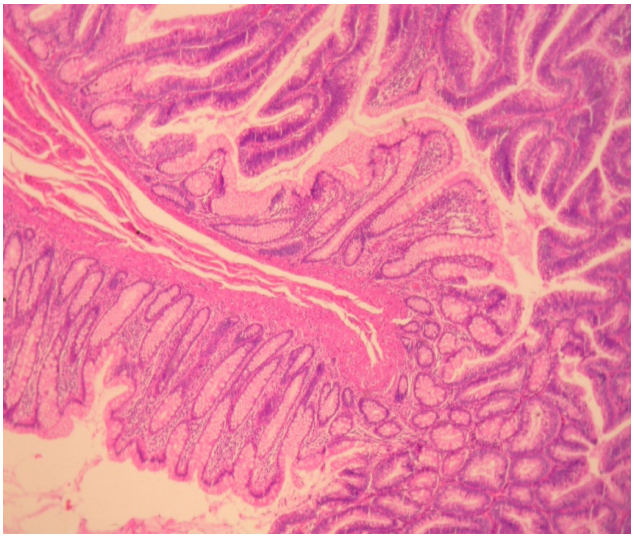

Figure 4. Area with high-grade dysplasia.

The patient was readmitted after 2 weeks with dehydration, renal failure and hydroelectrolyte imbalance and was treated conservatively.

After 6 months of uneventful evolution the patient was readmitted to close the ileostomy and she mentioned that she stopped the oral drugs for diabetes because the laboratory test performed during this period showed normal serum glucose level.

Discussion

Depletion syndrome appears on large villous adenomas of 7 to 18 cm, which represent 3% of villous adenoma [1]. Usually the rectum and sigmoid colon are the places where colonic villous adenomas appear and it is usually a sessile tumor and can be up to 18 cm in diameter. Size, villous configuration and degree of dysplasia should be specified to estimate the risk of malignancy. McKittrick and Wheelock were the first to describe secretory diarrhea caused by villous adenomas in 1954. In general the major symptoms of these villous adenomas are watery, mucinous diarrhea with increased bowel movements, while depletion syndrome is a rare complication. Regarding the mechanism of fluid and electrolyte loss, it seems that the closer to AM and larger the adenoma is, the more unlikely will the bowel be able to absorb the large secretory output of the tumor [9].

In patients with villous adenomas of the rectum studies show that the level of prostaglandin E2 (PGE2) is 3 to 6 times higher than normal. Locally released prostaglandin E2 has been postulated as secretagogue-mediated [10, 11]. PGE 2 synthetase inhibitors can be used to reduce the stool output, with caution in the case of patients with dehydration and renal failure. Fluid and electrolyte replacement along with surgery remains the best option to treat these patients. Our patient underwent a low anterior rectal resection with restoring renal function and electrolytes to normal levels. Other authors also mentioned dehydration and hydroelectrolyte imbalance noted at the first readmission.

Colonic malignant tumors generally develop from benign adenomas. The adenomas have a 2.5 % risk to evolve into colon cancer in 10 years [11], but the risk is increased for villous and larger adenomas [12]. Without treatment the mortality of secretory villous adenomas is 100% [13]. Current therapy consists of tumor resection (endoscopic or surgical) or brachytherapy (endocavitary irradiation), after correction of renal function and hydroelectrolyte imbalance [11]. However brachytherapy and endoscopic resection have poor results and high recurrence rates [9,11].

Conclusion

In conclusion, McKittrick-Wheelock syndrome is a rare and life-threatening disease due to the risk of severe complications caused by renal function impairment and hydroelectrolyte imbalance. The diagnosis could be challenging, because it is a digestive disease with an impact on renal function. The presence of this triad (chronic mucous diarrhea, renal function impairment with hydroelectrolyte imbalance and giant recto-sigmoidal tumor) should raise the suspicion of McKittrick-Wheelock syndrome.

The treatment of this disease is the removal of the tumor by endoscopy or surgery after correction of renal function and hydroelectrolyte imbalance.

The particularities of this case consist of a very large rectal villous adenoma and postoperative normalization of blood glucose level.

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