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McKittrick-Wheelock Syndrome: A Case Report

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Introduction

Mckittrick–Wheelock syndrome was first described in 1954 and is a rare complication of rectal villous adenomas [1]. This condition clinically manifests as large-volume secretary diarrhea, followed by pre-renal acute renal failure with severe electrolyte dysfunction (hyponatremia, hypochloremia, hypokalemia, and metabolic acidosis). The causative lesions frequently lie in the rectal or rectosigmoidal area [2].

Case Report

An 84-year-old Caucasian woman was admitted to the emergency department in October 2014 with complaints of fatigue, dizziness, shortness of breath during minimal physical activity, watery diarrhea, vomiting, nausea, and lack of appetite. It was her sixth hospitalization within six months with similar complaints. She reported feeling well up until May of the same year, at which time she developed fatigue, watery large-volume diarrhea, thirst, brief losses of consciousness, and cramps while gardening. At that time, she was admitted to the regional hospital, where acute kidney failure was diagnosed and a colonoscopy was performed. Rectal mass was found during the endoscopic examination. Taken biopsies showed tubulovillous adenoma. A surgeon consulted the patient, but she refused surgical treatment. After ten days of treatment, she was discharged in good overall condition. Following the initial admission, four additional hospitalizations at the Nephrology Center of Pauls Stradins Clinical University Hospital occurred between July and October 2014. The patient was successfully treated with adequate rehydration and electrolyte substitution therapy, and she denied repeated colonoscopic examination.

The patient had a medical history of myocardial infarction at the age of 65 years, but otherwise reported good health with no other significant medical problems. She had retired from working as the head of a civil service department. She had never smoked, and drank 1–2 alcoholic beverages during social occasions. Her family history was unremarkable, and she had three healthy children.

Upon physical examination during her most recent hospitalization, there were signs of volume depletion; dry mucous membranes and low blood pressure (90/70 mmHg) were observed. Blood tests showed a high hemo- concentration (hemoglobin, 169 g/L; hematocrit,51%). Biochemistry tests revealed renal failure(creatinine, 223 (35-97)μmol/L; urea, 18.6 (2,0-7,1)mmol/L) and severe electrolyte disturbances (sodium, 122 (136-145) mmol/L; potassium, 3.1 (3,6-5,1) mmol/L; chloride, 77 (101-111) mmol/L; calcium, 2.04 (2,2-2,6) mmol/L; phosphorus, 1.7 (0,81-1,5) mmol/L). Urinary analysis showed signs of a urinary tract infection (leucocytes and bacteria in the urinary sediment). Chest and abdominal X-ray examinations were normal. An ultrasound scan revealed signs of chronic renal parenchymal damage (hyperechogenic signals and reduced renal parenchyma). Physical and laboratory changes (hypovolemia with acute kidney injury, uremia, severe hyponatremia, hypokalemia, and hypochloremia) were the same as during previous hospital stays. The patient received antibacterial and intensive rehydration therapies, which resulted in gradual correction of the electrolyte imbalance (Table 1) and improved renal function.

The patient consented to a colonoscopy, which revealed a large (~10–12 cm), rectal, annular villous adenoma, beginning at the anal verge (Figure 1). Histopathologic examination confirmed a tubulovillous adenoma (Figure 2A) with a large population of goblet cells and a fragment containing adenocarcinoma cells. The patient subsequently underwent rectal extirpation, total mesorectal excision and sigmostoma formation with no further complications. The excised 28.5 cm long colon fragment (Figure 3) revealed a broad-based (11.2 cm × 11.9 cm × 2.7 cm) tubulovillous adenoma with a high-grade (GIII) adenocarcinoma (2.2 cm × 1.7 cm × 2.6 cm) invading through the lamina muscularis propria into the subserous layer (Figure 2B). The cancer had spread to four regional lymph nodes; pT3N2M0R0 stage cancer was determined.

The patient was discharged in a good overall condition. Because of her age, chemotherapy was not proposed due to the higher risk of chemotherapy-related side effects. At the 6-month follow-up, the patient was doing well, and her laboratory tests were within normal limits.

Laboratory values (normal range)	Date of hospitalization in October 2014				
	22nd	23rd	24th	26th	27th
Creatinine, μmol/L (35-97)	277	256	182	62	44
Sodium, mmol/L (132–146)	126	123	119	133	142
Chloride, mmol/L (99–109)	71	71	75	103	108
Potassium, mmol/L (3.5–5.0)	3.4	3.4	2.5	3.0	3.6
Magnesium, mmol/L (0.74-1.03)	-	-	-	-	0.79
Urea, mmol/L (2.0-7.1)	-	30.4	-	-	3.9

Table 1: Blood biochemistry values

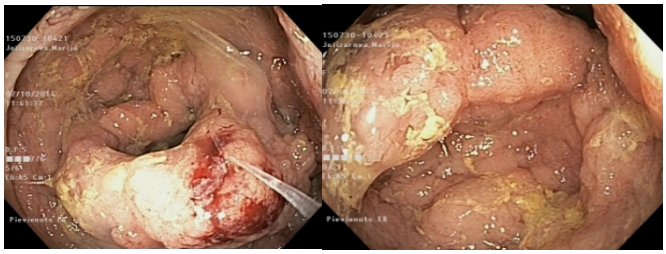

Figure 1: Colonoscopy revealed a rectal circular villous adenoma.

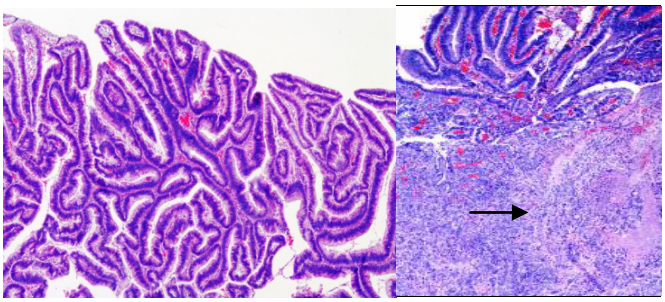

Figure 2: Histopathology of colon examination. Hematoxylin and eosin staining revealed A: Tubulovillous adenoma (× 40 magnification); B: Fragment containing adenocarcinoma cells (arrow; × 40 magnification).

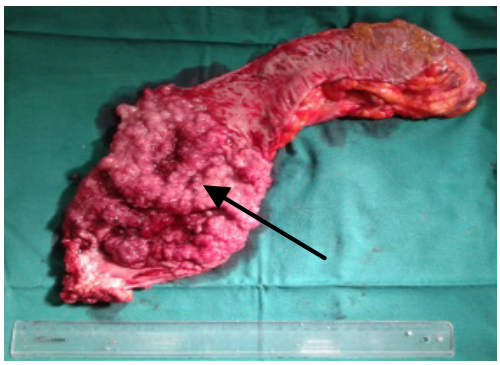

Figure 3: Macroscopic examination of the surgical material. The excised colon fragment (28.5 cm long) contained a broad-based tubulovillous adenoma with a low-grade adenocarcinoma (arrow) invading through the lamina muscularispropria into the subserous layer.

Discussion

Rectal or rectosigmoidal lesions in McKittrick–Wheelock syndrome typically exceed 4 cm in diameter [2]. In the case presented here, the lesion was 10–12 cm long and completely circumferential. Approximately 2% of patients with rectosigmoidal villous adenoma develop hypersecretory complications [3]. Fluid reabsorption was likely inhibited in the patient in the present case due to the limited remaining amount of normal colonic mucosa as a result of the distal villous adenoma [4]. The abnormal secretary function in this syndrome is thought to be mediated by secretagogues. For example, villous adenoma/carcinoma tissue synthesizes approximately 3–6 times more prostaglandin E2 than normal colonic mucosa [5].

Renal failure in McKittrick–Wheelock syndrome is usually reversed with adequate treatment unless complications from the severe electrolyte imbalance and volume depletion occur, which can lead to death. In the patient described in the present case, there was an episode of atrial fibrillation upon admission due to hypokalemia, which could have otherwise been fatal. Thus, early diagnosis is crucial in order to prevent renal complications as well as rectal carcinoma development [6-8]. Colorectal cancer has an annual progression rate of 0.43%. Male sex, older age, rectal site, and villous architecture were associated with an increased colorectal cancer risk in adenoma patients [9].

The only option to completely prevent electrolyte and fluid loss and cancer formation in Mckittrick–Wheelock syndrome patients is surgical resection of the adenoma, as untreated secretary adenoma has a mortality rate of 100% [10]. Laparoscopic surgical resection is also a feasible therapeutic modality for select cases of Mckittrick–Wheelock syndrome [11].

Conclusion

Mckittrick–Wheelock syndrome is rare disease with few reports in the literature. Our presented case demonstrates that the combination of typical clinical signs (watery diarrhea, and electrolyte and fluid loss) of Mckittrick-Wheelock Syndrome can induce acute renal failure, and is an indication for colonoscopy in early stages. Notwithstanding the patient age and wishes for conservative treatment, surgery was necessary to eliminate repeated hospitalizations and to improve the patient's quality of life.

Acknowledgments

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