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McKittrick–Wheelock Syndrome: Case Report

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ABSTRACT

McKittrick–Wheelock syndrome is a rare complication of colorectal villous adenomas characterized by chronic secretory diarrhea and severe electrolyte disturbances. We report a 72-year-old patient with chronic watery diarrhea, severe hypokalemia, hyponatremia, hypotension, and functional acute kidney injury. Colonoscopy revealed a large rectosigmoid tubulovillous adenoma. Laparoscopic resection resulted in complete clinical and biochemical recovery.

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KEYWORDS:
McKittrick–Wheelock syndrome, chronic diarrhea, villous adenoma, hypokalemia, hyponatremia, acute kidney injury.

INTRODUCTION

McKittrick–Wheelock syndrome is one of the rare causes of chronic diarrhea and represents a rare complication of colorectal villous or tubulovillous adenomas. It is characterized by chronic secretory diarrhea responsible for severe hydroelectrolytic disturbances and functional acute kidney injury. Diagnosis is often delayed because of the nonspecific nature of the clinical manifestations, although early management is essential to prevent potentially fatal complications. While correction of metabolic disturbances allows transient stabilization, complete excision of the tumor is the only curative treatment. We report the case of a 72-year-old patient with McKittrick–Wheelock syndrome revealed by chronic secretory diarrhea complicated by severe hypokalemia, hyponatremia, and functional acute kidney injury, successfully treated by surgical resection.

CASE REPORT

We report the case of a 72-year-old patient with a medical history of hypertension, ischemic stroke, superficial thrombophlebitis of the lower limbs, dyslipidemia, osteoarthritis, and depressive syndrome.

He initially presented to the emergency department with recurrent malaise and dizziness, revealing hypotension with hydroelectrolytic disturbances, particularly severe hypokalemia and functional acute kidney injury. These abnormalities were corrected with good clinical improvement, and the patient was discharged home. A few days later, he presented again with the same clinical picture, leading to another hospitalization and an etiological work-up. The patient was subsequently referred to the hospital in his hometown for further management.

On questioning, the patient reported several months of progressively worsening watery diarrhea, without abdominal pain or overt gastrointestinal bleeding, associated with episodes of malaise when standing up, in a context of preserved general condition.

Clinical examination revealed a patient in good general condition with orthostatic hypotension and an unremarkable abdominal examination. Watery diarrhea was objectively documented.

Laboratory tests showed severe hypokalemia at 2.5 mmol/L, controlled with daily supplementation of 4 g of potassium,

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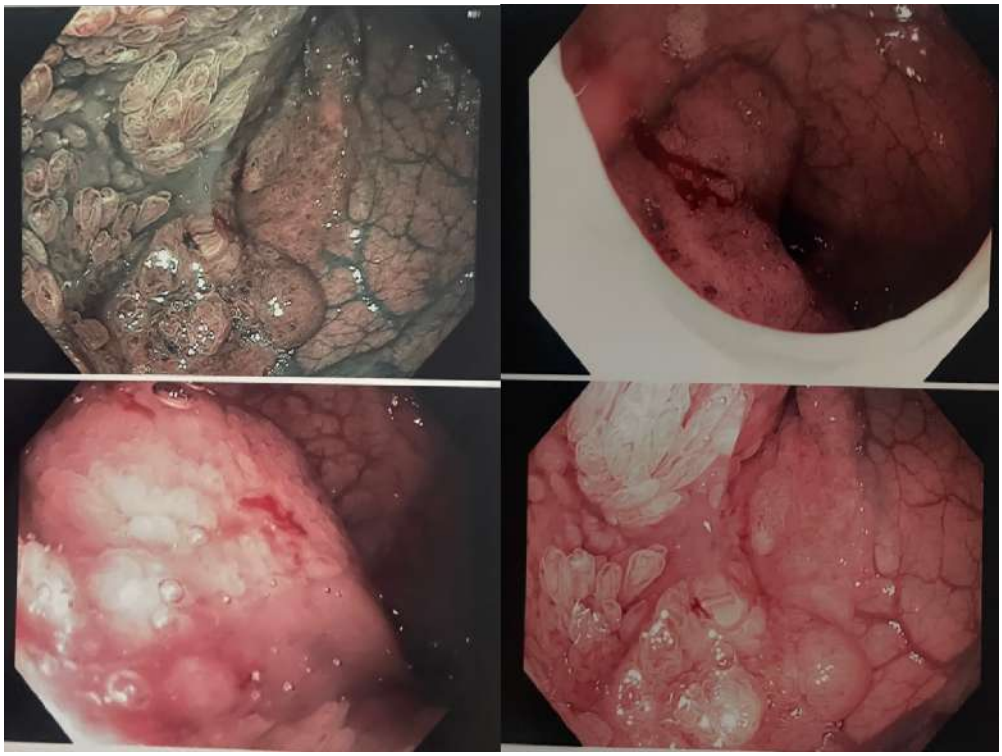
hyponatremia at 127 mmol/L, and acute kidney injury with a creatinine level of 277 mmol/L, which resolved after rehydration.

Abdominopelvic computed tomography showed a tumoral thickening of the rectosigmoid junction measuring 15 cm in length, beginning 10 cm from the anal verge. Colonoscopy subsequently revealed a large, non-stenosing rectosigmoid villous lesion, with its lower pole estimated at 10 cm from the anal verge and extending over a height of 10 cm. The difference in lesion length observed between the two examinations may be explained by differences in the measurement methods used in radiological imaging and endoscopic assessment. Histological examination concluded that this was a tubulovillous adenomatous lesion with low-grade dysplasia and no evidence of invasion.

Based on these findings, a diagnosis of McKittrick–Wheelock syndrome was established. Following multidisciplinary team discussion, surgery was selected. The patient underwent laparoscopic sigmoid resection with end-to-side colorectal anastomosis. The postoperative course was uneventful. Histological examination of the surgical specimen showed findings identical to those observed in the biopsies.

The postoperative course was uneventful, with resolution of the diarrhea and correction of the electrolyte abnormalities.



DISCUSSION

McKittrick–Wheelock syndrome (MWS) is an exceptional complication of colorectal villous or tubulovillous adenomas. Initially described by McKittrick and Wheelock in 1954, it is characterized by chronic secretory diarrhea, severe hydroelectrolytic disturbances, and functional acute kidney injury. Despite its potentially fatal nature, this condition remains largely underrecognized, which explains the frequent diagnostic delay (1).

The largest systematic review published to date, conducted by Orchard et al., identified 257 cases reported worldwide. The authors found a median age of 69 years, a median symptom duration of 24 months, a median serum sodium level of 122 mmol/L, and a median potassium level of 2.7 mmol/L, demonstrating the severity of hydroelectrolytic disturbances before diagnosis (2). Our observation is fully consistent with these data, as the 72-year-old patient had chronic diarrhea evolving for several months, complicated by hyponatremia (127 mmol/L), severe hypokalemia (2.5 mmol/L), and functional acute kidney injury requiring several hospitalizations.

The pathophysiology of the syndrome is now well established. Large villous adenomas have marked secretory activity related to overexpression of prostaglandin E2 (PGE2). PGE2 stimulates adenylate cyclase in colonic epithelial cells, increasing intracellular cyclic AMP concentrations and inducing active secretion of sodium, chloride, potassium, and water into the intestinal lumen. Digestive losses, sometimes exceeding two liters per day, exceed the reabsorptive capacity of the distal colon, particularly when the tumor is located in the rectal or rectosigmoid region. This results in chronic hypovolemia responsible for prerenal kidney injury and sometimes severe

hypotension (3,4). This observation led to the proposed use of cyclooxygenase inhibitors, particularly indomethacin, as a temporary medical treatment. By inhibiting prostaglandin synthesis, indomethacin reduces tumor secretory activity, resulting in decreased stool volume and hydroelectrolytic losses while awaiting surgery (5).

Tumor size and location play a determining role in the development of the syndrome. Most published cases involve lesions larger than 4 cm, located in the rectum or rectosigmoid. The large size provides a greater surface area for secretion, while the distal location of these lesions means that only a minimal area of normal colonic mucosa remains available for hydroelectrolytic reabsorption (3,6).

Diagnosis remains challenging because of the low specificity of the clinical manifestations. Patients generally present with chronic diarrhea, malaise, fatigue, orthostatic hypotension, muscle cramps, or recurrent acute kidney injury (7). These symptoms frequently lead to incorrect diagnoses such as gastroenteritis, hyperaldosteronism, diabetic ketoacidosis, or drug-induced diarrhea. It is often only after repeated episodes of dehydration and identification of a large distal adenoma that the diagnosis is finally considered (2).

In our case, the patient experienced several episodes of severe dehydration with acute kidney injury before the diagnosis was established. This course perfectly illustrates the decompensation phase described by Orchard et al., characterized by progressive worsening of digestive losses leading to marked hypovolemia. The presence of chronic watery diarrhea associated with profound hypokalemia and functional kidney injury should prompt early colonoscopy to search for a distal secretory adenoma.

According to Emrich and Niemeyer, untreated secretory villous adenomas may be associated with a very high mortality rate due to severe electrolyte disturbances and renal failure (6). In the systematic review by Orchard et al., the mortality rate was 5.2% among patients who underwent surgery, compared with 61.5% among those who did not undergo surgical treatment (2).

Management initially relies on intensive correction of hydroelectrolytic disturbances through fluid resuscitation and sodium and potassium supplementation. However, this strategy is only temporary, as abnormalities recur as long as the tumor persists.

Curative treatment consists of complete tumor excision. The choice of technique depends on tumor size, location, and suspicion of malignant degeneration (8). According to the systematic review by Orchard et al., nearly two-thirds of patients (64.8%) underwent surgical resection, including anterior resection, abdominoperineal resection, Hartmann's procedure, or sigmoid colectomy. This approach resulted in resolution of symptoms and durable correction of hydroelectrolytic disturbances, confirming that complete surgical excision of the adenoma is the standard curative treatment for McKittrick–Wheelock syndrome (2).

Minimally invasive techniques, such as transanal endoscopic microsurgery (TEM), transanal minimally invasive surgery (TAMIS), endoscopic mucosal resection (EMR), and endoscopic submucosal dissection (ESD), are increasingly used in the treatment of McKittrick–Wheelock syndrome. However, their use may be limited by the often large size of the adenomas and appears to be associated with a higher risk of recurrence. In the presence of large lesions, resectional surgery, particularly through a laparoscopic approach, remains the preferred strategy, providing excellent functional outcomes and an excellent prognosis after complete tumor excision (9).

CONCLUSION

McKittrick–Wheelock syndrome is a rare but potentially serious condition that should be considered in any patient presenting with chronic watery diarrhea associated with hyponatremia and severe hypokalemia, particularly when accompanied by functional acute kidney injury. Early recognition of this syndrome allows prompt correction of hydroelectrolytic disturbances, which is essential to prevent potentially fatal complications. However, only complete excision of the adenoma provides definitive cure.

Declaration of originality

This case report is original and has not been published previously, nor is it under consideration for publication elsewhere.

Consent

Informed consent was obtained from the patient for publication of this case report and any accompanying images.

Ethical approval

Ethical approval was not required for this single-patient case report, as per institutional guidelines.

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