McKittrick Wheellock syndrome. Giant villous adenoma of the rectum: a case report

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ABSTRACT

McKittrick-Wheelock syndrome is a rare entity caused by giant villous adenomas of the rectum, which is characterized by hypersecretory diarrhea and hydroelectrolytic disturbances that can progress to acute renal failure. A clinical case resolved by surgical treatment is presented.
Keywords: *McKittrick-Wheelock syndrome, villous adenoma, severe diarrhea*

INTRODUCTION

Adenomatous polyps are neoplastic lesions with a 35% risk of cancer when they are ≥30 mm and colonoscopy is the best method for their diagnosis.[1] McKittick Wheelock syndrome is a rare entity first described in 1954 by Leland McKittrick and Frank Wheelock, characterized by hypersecretory diarrhea with severe hydroelectrolytic depletion and acute renal failure, associated with large sessile villous adenomas of the rectum, commonly larger than 4 cm, although they can vary between 3 and 18 cm. The location in the rectum, where water absorption is much lower, generates the symptoms described.
Severe hyponatremia and dehydration are caused by increased secretion of prostaglandins E2 (PGE2) by adenomatous cells. Because these tumors are located near the anus, sodium and water cannot be absorbed, causing hypersecretory diarrhea.[2]
Histopathology frequently finds high-grade dysplasia and foci of adenocarcinoma in 80% of cases. When the diameter is greater than 2 cm, the risk of malignancy is 50%.[3]
For diagnosis, colonoscopy and pelvic magnetic resonance imaging (MRI) with endorectal gel are essential.

CASE

A 75-year-old male patient was referred from his hometown after a prolonged hospitalization for prerenal acute renal failure and exacerbated heart failure, associated with 6-7 daily episodes of mucous secretion over a 4-month period.
His medical history included arterial hypertension, chronic heart failure, and atrial fibrillation.
Physical examination revealed a hypotensive and tachycardic patient, with moderate dehydration and moderate general and nutritional status. Digital rectal examination revealed a mobile vegetative lesion on the anterior aspect, 5 cm from the anal margin.
Laboratory tests revealed hemoglobin 8.29 gr/dl, creatinine 2 mg/dl, BUN 40 mg/dl, hypokalemia, hypoalbuminemia, and negative tumor markers.
Colonoscopy showed a broad-based, polylobulated vegetative lesion, 8 to 12 cm from the anal margin (Fig. 1).
Histopathology reported a villous adenoma with low-grade dysplasia

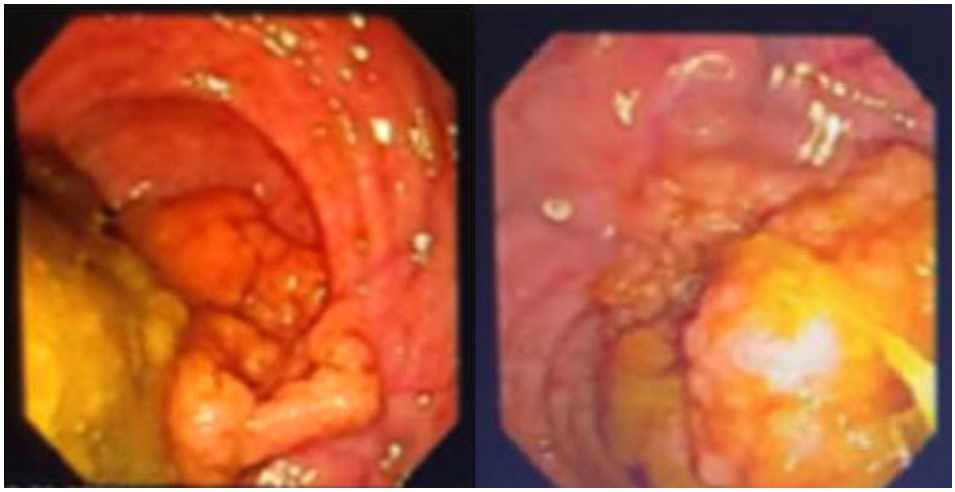

Figure 1. *Colonoscopy showing a vegetative, polylobulated rectal lesion.*

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Received: March, 2023. Accepted: June, 2024

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A pelvic MRI with intravenous contrast and endorectal gel was performed, which showed a 65 mm vegetative mass with muscle involvement on the anterior surface of the middle and lower rectum, 45 mm from the anorectal ring (Fig. 2). Stage: T2N0M0.

A CT scan of the chest, abdomen and pelvis with intravenous contrast was performed, which ruled out metastatic disease. High-dose intravenous N-acetylcysteine preparation was used before and after the study to reduce nephrotoxicity.

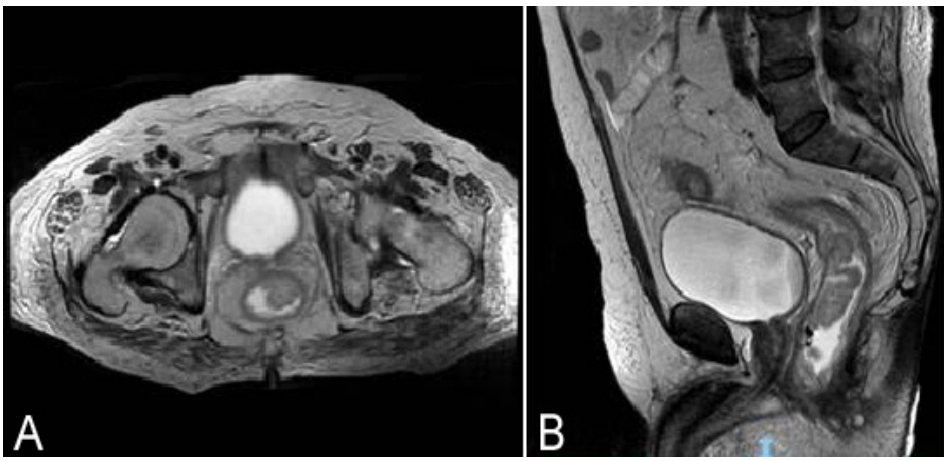

Figure 2. *MRI with intravenous contrast and endorectal gel in T2 sequence. Axial* ***(A)*** *and sagittal* ***(B)*** *images showing an elevated and polylobulated lesion on the anterior surface of the middle and lower rectum. T2N0M0.*

An intravenous line was placed for electrolyte replacement and endorectal indomethacin was administered. The patient was clinically stable. Given the suspicion of a malignant tumor and taking into account the patient's past medical history, it was decided to perform surgery via an abdominal approach. The Hartmann's procedure was performed (Fig. 3) due to the patient's poor general and nutritional status.
The anatomic pathology reported a 20 cm rectosigmoid specimen with a 6 cm polypoid lesion located 1 cm from the distal resection margin and 13 cm from the proximal margin. The 12 lymph nodes removed were negative for malignancy. Histopathology revealed a villous adenoma with foci of high-grade dysplasia and free surgical margins.

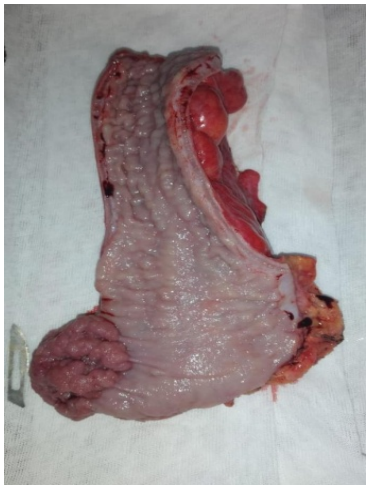

Figure 3. *Surgical specimen with the villous adenoma at its distal end.*

DISCUSSION

Diarrhea caused by a giant villous adenoma is due to the production of PGE2, which increases intracellular cyclic AMP, thereby, activating apical chloride and potassium channels in epithelial cells. This causes massive water and electrolyte losses. Elevated levels of PGE2 can be found in the stool and increased glandular expression of prostaglandin endoperoxide synthase (COX-2) can be found in the tumor histology. When the tumor is removed, PGE2 levels return to normal.
Renal injury, one of the main features of this syndrome, is caused by hypovolemia secondary to loss of intestinal fluid and electrolytes. For the control of diarrhea, the use of endorectal indomethacin, a PGE2 inhibitor, has been shown to decrease adenoma secretion.[2]
Currently, the use of endorectal ultrasound has helped to assess the depth of invasion, allowing the proposal of tumor excision by endoscopic mucosal resection or endoscopic submucosal dissection. Transanal endoscopic microsurgery (TEM) and transanal minimally invasive surgery (TAMIS) have advanced in the treatment of villous adenomas; However, they are reserved for high-volume colorectal pathology centers.[1] These procedures are indicated for benign lesions and early malignant lesions defined as adenocarcinomas confined to the superficial submucosa (T1 Sm1), well or moderately differentiated, small, with no lymphatic, vascular or perineural involvement.[4] In our patient, the lesion was large (more than 6 cm) and there was also suspicion of invasion of the muscularis propria by MRI, so it was decided to perform a rectal resection via an abdominal approach. Taking into account the patient's previous medical history and especially his poor general condition, resection was performed without primary anastomosis

CONCLUSION

The diagnosis of McKittrick-Wheelock syndrome is complex due to the nonspecificity of its symptoms, so the index of suspicion must be high.
Fluid and electrolyte replacement and the application of endorectal indomethacin are useful to stabilize the internal environment and plan a definitive surgical treatment.
The management of these patients must be individualized, taking into account the resection of the lesion according to its characteristics and the clinical condition of the patient, together with the availability and training of the medical personnel involved.

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