

# Laparoscopic Approach in the Surgical Management of McKittrick–Wheelock Syndrome

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## Introduction

McKittrick-Wheelock syndrome is a pathological entity that is represented by the presence of a large rectal tumour with histopathological findings of tubular-villous adenoma that associates important secretion of electrolyte-rich mucin. Thus, this syndrome will inflect multiple diarrheic stools on the affected patients with the secretion of electrolyte-rich mucin that will manifest as an electrolyte imbalance with hypopotassaemia, hyponatremia, and hypochloroemia, as well as with pre-renal chronic kidney injury and severe dehydration syndrome. Initially described in 1954 by Leland S. McKittrick and Frank C. Wheelock, the medical literature comprises only 257 such cases worldwide by the year 2018. We can thus conclude that the debate regarding the diagnosis and therapeutic management of McKittrick-Wheelock syndrome is still in its infancy. Usually, from a clinical standpoint, watery diarrheic stools with mucus discharge and subsequent metabolic issues, are the reason to blame for the generally altered state in which the patients address the physician with nausea, vomiting, fatigability, and even fainting.

The clinical finding characteristic of this disease is the prolonged time of symptomatology progress, usually about 24 months until the first presentation, patients having multiple admissions to the hospital or outpatient clinic in this timeframe with diagnoses ranging from enterocolitis to ketoacidosis or infectious colitis. This determined many authors to define McKittrick-Wheelock syndrome as a rare cause of metabolic coma and to introduce it in the differential diagnosis of prerenal chronic kidney injury.

Nevertheless, this mandated the development of specific medical conservative treatments to normalize altered kidney function and to counteract sodium depletion, a fundamental element in the preparation for surgical resection.

Thus, from a therapeutic standpoint, surgical success leans on the efficacy of the preoperative medical treatment, which may be the only line of treatment for patients that refuse surgical resection. In this case, treatment is going to be an association of indomethacin and octreotide that will reduce fluid loss. We further state that surgical treatment remains the only option that can be curative and radical for this affection and that literature studies show the mortality from villous rectal adenomas specific to the McKittrick Wheelock syndrome to be 100%.

## Material and method

We present the case of a patient diagnosed in 2019 with a rectal tumour, compatible from a clinical, paraclinical, and histopathological standpoint with McKittrick Wheelock syndrome that was operated minimally invasive, through laparoscopy, in the Surgery Clinic of the Emergency County Hospital of Constanta.

## Case report

A 65-year-old patient, with a rectal tumor discovered in 2019, that was not diagnosed or followed up and was neglected therapeutically, with many admissions during the last couple of years in the Nephrology department, where she was investigated and treated for chronic kidney injury through a pre-renal mechanism.

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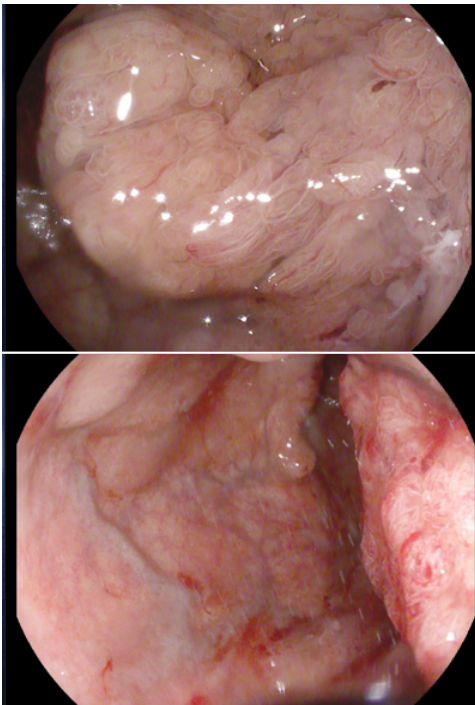
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The patient was admitted in late 2020 through the Emergency Care Unit of Constanta County Hospital, with a generally altered state caused by multiple diarrheic stools at home. The lab work-up revealed a severe dehydration syndrome with nitrous-oxide retention and hyponatremia. After analysis of the biochemical results (creatinine = 3.26 mg/dl, urea = 176 mg/dl, K = 3.6 mmol/l, Na=139 mmol/l, proteins = 6.2 g/dl) the patient was admitted to the Nephrology department with the diagnosis of kidney injury of pre-renal origin. The general state of the patient after starting specific treatment got better, with partial normalization of biomarkers and partial symptom remission, which permitted further investigations.

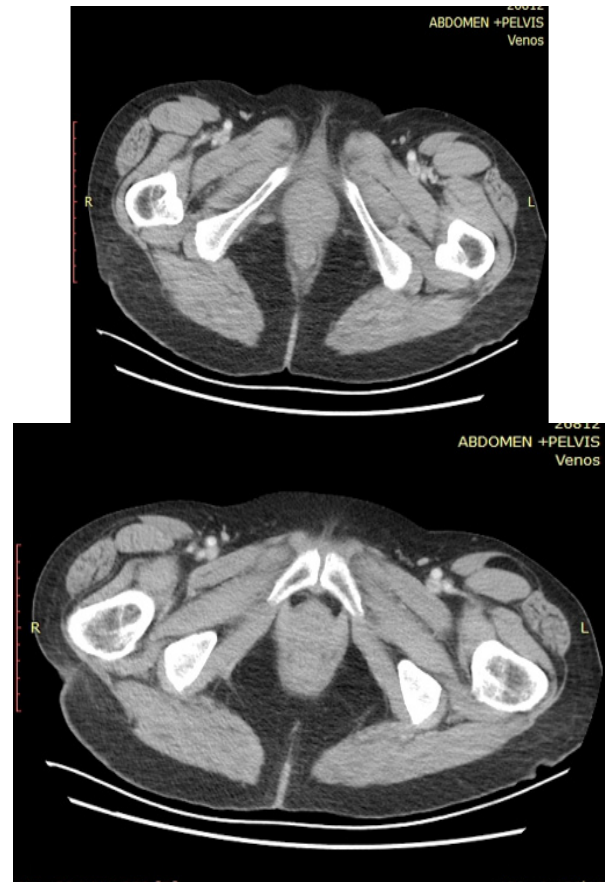
Considering the patient's history, a rectoscopy is ordered that shows internal hemorrhoids in the anal canal and a protrusive tumoral mass at 2 cm from the external anal edge, circumferential but non-obstructive on a length of about 10 cm, covered by friable mucosa that is biopsied. The histopathological result shows a tubule-villous adenoma with high-grade and low-grade intraepithelial dysplasia and metaplasia.



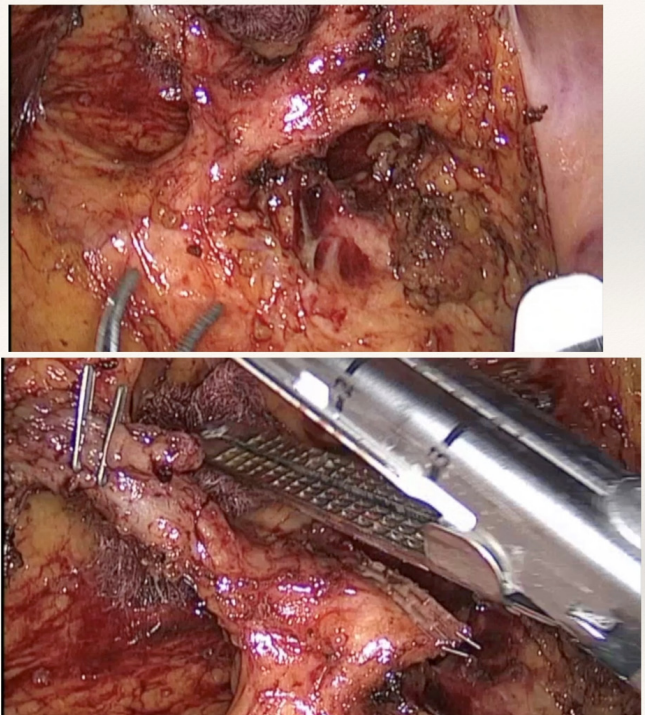
**Figure 1:** Images from the rectoscopic examination showing a protrusive tumoral mass suggestive of a tubulovillous adenoma.

In this context, further investigations are considered, and a native CT scan of the abdomen and pelvis is performed that reveals and confirms a protrusive, iodophilic, circumferential parietal thickening, with imprecise edges, and irregular shape, of about 29 mm that associates the distension of the colon upstream; normal liver with homogenous structure, a suprarenal adenoma, and a left cortical kidney cyst Bosniak, with no. other visible lesions.

The clinical data, biological and imagistic results, together with the rectal biopsy, point to the diagnosis of Mckittrick-Wheelock syndrome, mandating the patient's transfer to the Surgery Department for specialized treatment. After a short preoperative preparation with electrolyte rebalance and mechanical purging of the colon, the patient is scheduled for laparoscopic resection bearing in mind the advantages of this technique regarding

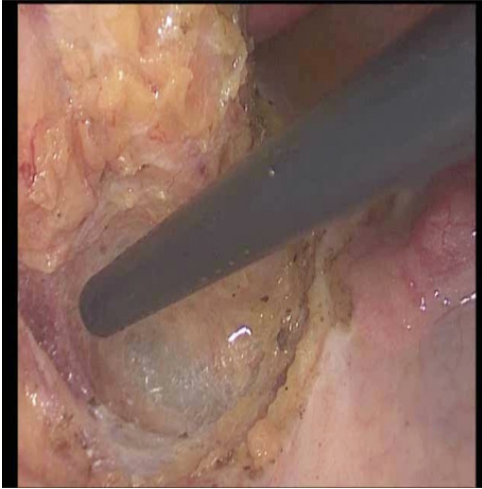


**Figure 2:** Images from the rectoscopic examination showing a protrusive tumoral mass suggestive of a tubulovillous adenoma.

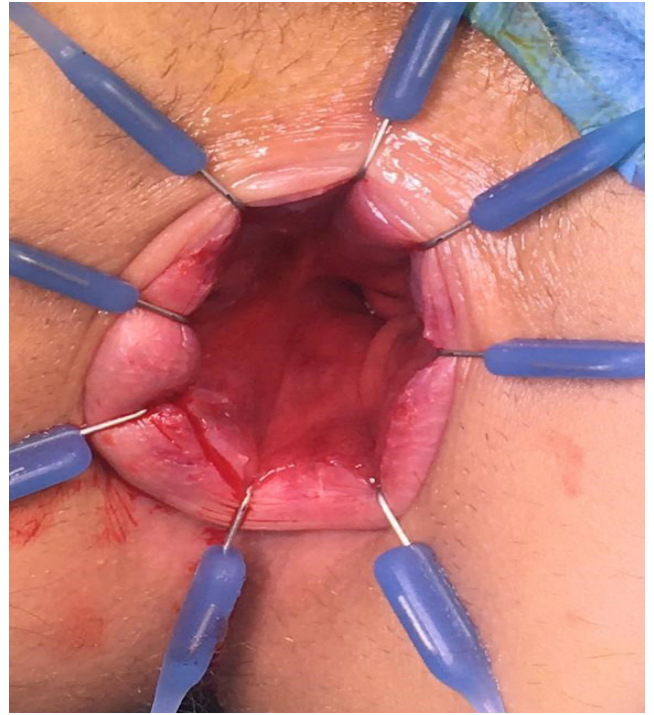


**Figure 3:** Intra-operative laparoscopic view. High vascular dissection up to the origin of the inferior mesenteric artery from the aorta and central lymphadenectomy. Transection of the artery with a Vascular Endo-Gia stapler, after the emergence of the left colic artery (Low-tie). The inferior mesenteric vein was clipped twice.

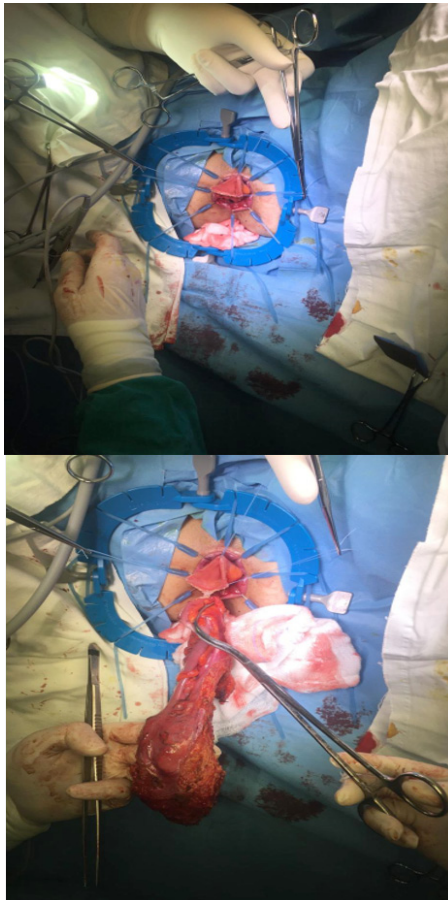
postoperative evolution and the fact that this type of approach is “sphincter-saving” Ultralow laparoscopic rectal resection with complete mesorectal excision and mechanical terminal-terminal colo-anal anastomosis is performed with a protective derivative ileostomy is performed.



**Figure 4:** Laparoscopic intraoperative view of the rectal dissection in the embryological planes that achieves complete mesorectal excision. Dissection in an avascular plane (Holly Plane) all the way to the pelvic floor muscles in between the proper mesorectal fascia and the presacral fascia.



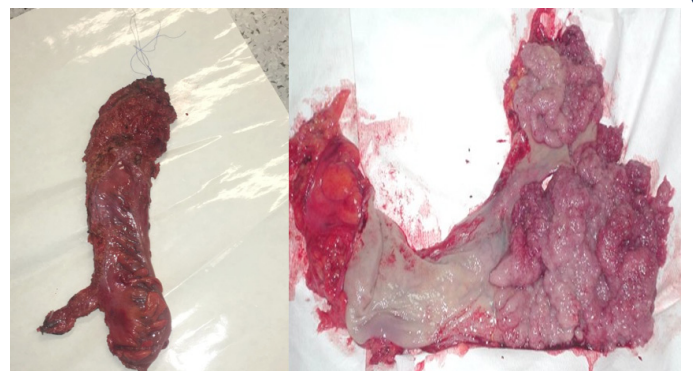
**Figure 6:** Final view of the colo-anal anastomosis.



**Figure 5:** Intraoperative perianal view. Dissection of the mesorectal plane with trans-anal specimen resection followed by hand-sewn colo-anal anastomosis.

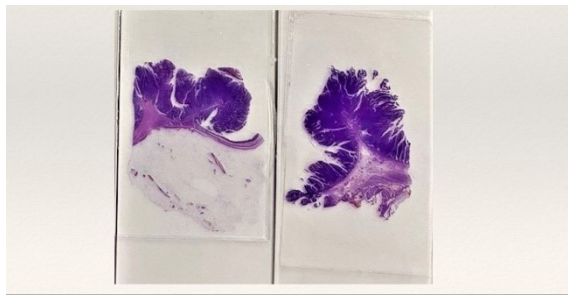
## Results

From a surgical standpoint, the postoperative evolution of the patient was favorable, having no abdominal pain, and minimal peritoneal drainage that was subsequently removed one by one. Intestinal transit restarted through the protective ileostomy that had proper healing. Postoperative wound with good healing. No signs of fever or other general imbalances with discharge on day 7 p.o. Ileostomy reversal was scheduled in 4 weeks' time. Regardless of these favorable results from a pure surgical point of view, we consider the histopathological results on the final specimen to be the definitive appreciation point. After a macroscopic examination of the specimen, it is noted to be 23 cm in length, circumferential, and with a protrusive vegetating tumor, 12/7/2 cm in size, reddish color and friable mucosa.



**Figure 7:** Macroscopic aspect of the specimen.

From a microscopic point of view, the rectal specimen had a tumoral mass compatible with a tubulovillous adenoma with high and low-grade intraepithelial dysplasia/neoplasia, laminae propria with chronic inflammation, and hyperemia. 4 regional lymph nodes non-invaded and surgical margins as well, the closest being the distal margin at about 1.5 mm.



**Figure 8:** Microscopic aspect of the specimen.

### Discussion

McKittrick-Wheelock was first described by McKittrick and Wheelock in 1954. It is a rare disorder with fluid and electrolyte depletion caused by a secretory colorectal tumor. In most cases, a villous adenoma [3]. Most of the patients with this disorder can present with chronic diarrhea and symptoms due to electrolyte imbalances, such as lethargy, muscle cramps, ileus, and vomiting. The incidence and prevalence of MWS are difficult to estimate as some of the cases have also been reported as electrolyte depletion syndromes [4,8,9].

Recent studies have shown that in patients with this condition, rectal secretions have higher concentrations of prostaglandin E2 (PGE2) and intracellular cyclic adenosine monophosphate (cAMP). Additionally, a large surface area of the villous adenomas further causes increased fluid secretion, which exceeds the reabsorption ability of the remaining normal rectal mucosa [8,10,11]. As treatment methods, the surgical intervention aims to remove villous adenoma after correcting fluid and electrolyte imbalances. Also, brachytherapy and endoscopic tumor resection are feasible alternatives [8,11]. Most colon cancers develop from benign adenomas, but the risk is higher when adenomas are villous and large. Secretory villous adenomas have 100% mortality if left without any treatment [12].

### Conclusions

McKittrick-Wheelock syndrome is a rare and life-threatening disease due to the risk of severe complications caused by renal function impairment and hydro electrolyte imbalance. The presence of the clinical triad of renal function impairment with hydro electrolyte imbalance, giant recto-sigmoidal tumor, and chronic mucous diarrhea should raise the suspicion of McKittrick-Wheelock syndrome. The large surface area of the villous adenoma and increased levels of PGE2, which serve as a secretagogue, is responsible for secretory diarrhea and electrolyte disturbances in MWS. The trial of PGE2 synthase inhibitors, such as indomethacin, can be given to the patient while waiting for surgery to improve the symptoms. The treatment of this disease is the removal of the tumor, preferably by minimally invasive surgery by laparoscopic approach, due to the multiple advantages of this technique which will benefit the patient, after the correction of kidney function and hydro electrolyte imbalance.

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