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McKittrick-Wheelock syndrome: a challenging condition

Síndrome de McKittrick-Wheelock: una condición desafiante

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Dear Editor:

The recent McKittrick-Wheelock syndrome (MKWS) case published by Arrazola et al. provides an excellent illustration of this condition's diagnostic challenges [1]; therefore, we would like to share additional recent cases that further highlight the importance of early recognition. They described a 60-year-old male patient who had this rare syndrome that was first described in 1954, presenting with chronic diarrhea, hypokalemia, and headache, and visual impairment, associated with a non-keratinizing squamous cell rectal carcinoma[1]. Two weeks before his admission, he had noticed a painful mass in the left inguinal area, and further abdominal imaging studies revealed a rectal tumor extending to this region. The patient evolved with hypokalemia unresponsive to electrolyte replacement therapy and died before the planned definitive surgical intervention; the authors emphasized the importance of the early suspicion about this syndrome, allowing a surgical treatment[1]. Below are several representative examples of this syndrome.

Case 1. A 79-year-old man presented with syncope, severe electrolyte disturbances and renal failure due to a rectal polyp diagnosed by abdominal images and biopsy[2]. As the piece-meal removal was unsuccessful, he underwent abdominoperineal resection with permanent colostomy, resulting in satisfactory recovery and improved quality of life. The authors highlighted the early diagnosis, accurate preoperative stabilization, and prompt intervention to favor the best results in the management of MKWS patients[2].

Case 2. An 83-year-old woman with a large low rectal polyp had chronic diarrhea, electrolyte disorder, and renal disease, hypoalbuminemia, and anemia[3]. The clinical diagnosis of MKWS was confirmed by laboratory data, colonoscopy, radiological imaging, and histopathological pattern (tubulovillous adenoma and low-grade dysplasia); she underwent a transanal submucosal excision and Delorme's plication with success[3]. The authors emphasized the utilization of this procedure as the best option for the management of those low rectal polyps that are too large to be treated endoscopically.

Case 3. A 67-year-old man presented with weakness and bloody diarrhea with mucus for over a week, and a rectal irregular mass was palpated; he also had hypokalemia, hyponatremia, and hypochloremia, besides very elevated levels of creatinine and urea[4]. After hemodialysis, the electrolyte levels became normal, and the patient was discharged; the manifestations recurred three times during three months, and were similarly managed. The colonoscopy revealed a vegetative mass from the 3rd cm of the rectum to the 13 cm proximal, filling most of the lumen without preventing the progression of the endoscope, there was a vegetative sigmoid lesion of irregular borders occupying 40% of its lumen; the diagnoses were a rectal villous adenoma and an adenocarcinoma of sigmoid colon[4]. He underwent uneventful laparoscopic low anterior resection and loop ileostomy; the authors stressed that synchronous tumor with a villous polyp had never been reported[4].

Case 4. A 73-year-old woman was evaluated with suspicion of colon cancer, because of her frequent loose stools with mucus since 2016, being self-medicated without success[5]. The video colonoscopy showed a rectal tumor of 3 cm, at 12 cm from the anus; it was endoscopically removed and was a villous adenomatous polyp without malignant signs. Over 6 years, the rectal villous adenoma has recurred repeatedly, and at least 3 attempts at endoscopic removal of recurrent neoplasms have been done[5]. In December 2022, she

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presented hypotension, diarrhea, muscle cramps, and episodes of loss of consciousness; the creatinine was over 550 µmol/L, and urea was over 40 mmol/L; so a Grade III prerenal acute kidney injury was diagnosed and treated by nephrologists. The data were consistent with MKWS due to the villous rectal tumor, but the development of metabolic acidosis, usually presented in cases of the syndrome, was not observed[5]. In March 2023, imaging evaluations revealed a tumor (68 mm x 44 mm) narrowing the intestinal lumen and intussusception of the rectosigmoid transition due to large volume; the intervention performed was by direct tumor resection and a preventive ileostomy[5]. It is noteworthy that the patient repeatedly refused tumor removal, but multidisciplinary medical cooperation resulted in her radical surgical treatment with good rehabilitation.

MKWS may be unsuspected, underdiagnosed, misdiagnosed, or underreported; therefore, the following recent additional literature data aim to call more attention to it. The initial manifestations are nonspecific and followed by a latent phase of diarrhea, before the deterioration and decompensation with risk of poor outcomes. Management options depend on the tumor size and site; the small lesions can be treated by endoscopic resection, while the larger ones may require more extensive surgical resections; the most difficult to treat are those sited in the lower rectum or associated with rectal prolapse.

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