CASE REPORT

The McKittrick-Wheelock syndrome: a rare cause of chronic diarrhoea

Jana G Hashash,¹ Jennifer Holder-Murray,² Elie Aoun,³ Dhiraj Yadav¹

¹Division of Gastroenterology, Hepatology, and Nutrition, University of Pittsburgh, Pittsburgh, Pennsylvania, USA

²Department of Surgery, Division of Colon and Rectal Surgery, University of Pittsburgh, Pittsburgh, Pennsylvania, USA

³Division of Gastroenterology, West Penn Allegheny Health System, Pittsburgh, Pennsylvania, USA

Correspondence to
Dr Elie Aoun,
eaoun@wpahs.org

SUMMARY

The McKittrick-Wheelock syndrome, a rare disorder, is caused by fluid and electrolyte hypersecretion from a rectal tumour and patients can develop a depletion syndrome characterised by severe dehydration, hyponatraemia, hypokalaemia and metabolic acidosis. We present a case of a 62-year-old man who presented with chronic diarrhoea that had resulted in numerous previous hospital admissions. On physical exam, the patient showed signs of volume depletion. A soft polypoid mass was appreciated on digital rectal examination. Laboratory tests showed renal failure and significant electrolyte abnormalities. Colonoscopy revealed a large, friable mass in the rectosigmoid region. Biopsies were consistent with tubulovillous adenoma. Subsequently, the patient underwent surgical resection, which on pathology exhibited evidence of high-grade dysplasia, and the patient was diagnosed with McKittrick-Wheelock syndrome. It is essential to identify this condition in a timely manner as it is associated with high morbidity and complications, some of which may be life threatening.

BACKGROUND

The McKittrick-Wheelock syndrome, a rare disorder, is caused by fluid and electrolyte hypersecretion from a rectal tumour. Patients affected by this syndrome can develop a depletion syndrome characterised by severe dehydration, hyponatraemia, hypokalaemia and metabolic acidosis.¹ It is imperative that physicians include this diagnosis in the work up of patients with chronic diarrhoea, because if not treated appropriately, this condition can lead to detrimental outcomes. We hereby present a case of a patient who presented with chronic diarrhoea, signs of volume depletion and electrolyte abnormalities. The timely diagnosis and treatment of the McKittrick-Wheelock in this patient lead to successful outcomes, and at long-term follow-up, the patient was symptom free.

CASE PRESENTATION

A 62-year-old man with a history of monoclonal gammopathy of undetermined significance was admitted for evaluation of chronic diarrhoea of 1.5-year duration. The patient described the frequency of diarrhoea to be approximately 10 times per day; it was profuse, watery, non-bloody, present even at fasting and led to nocturnal awakenings. He denied any abdominal pain, nausea or vomiting, and there was no significant history to suggest an infectious origin. The diarrhoea resulted in six previous hospital admissions for severe dehydration,

electrolyte abnormalities and renal insufficiency. On review of systems, he admitted to an unintentional weight loss of approximately 30 lbs.

INVESTIGATIONS

Significant findings on physical examination included signs of volume depletion, including a blood pressure of 98/54 mm Hg, pulse of 103 bpm and dry mucous membranes. A soft polypoid mass was appreciated on digital rectal examination extending from the levators to beyond the examining finger. Laboratory tests showed renal failure and significant electrolyte abnormalities including: blood urea nitrogen 86 mg/dl (baseline 14 mg/dl; normal 6–20 mg/dl), serum creatine 5.3 mg/dl (baseline 1.0 mg/dl; normal 0.5–1.2 mg/dl), serum sodium 127 mmol/l (normal 135–143 mmol/l), potassium 3.0 mmol/l (normal 3.6–5.0 mmol/l), magnesium 1.6 mg/dl (normal 1.7–2.2 mg/dl), phosphorus 10.1 mg/dl (normal 2.4–4.1 mg/dl) and an anion gap 32 meq/l (normal 8–12 meq/l). Stool studies were negative for infectious organisms and suggested secretory diarrhoea with stool sodium of 110 meq/l, potassium of 43 meq/l, osmolality of 330 mOsm/kg and a calculated stool anion gap of 16 mOsm/kg. Renal failure and electrolyte disturbances improved with intravenous fluid hydration and electrolyte replacement. Colonoscopy revealed a diffuse, non-obstructing circumferential polypoid, friable mass involving the rectosigmoid colon and extending from the proximal anal canal to 30 cm proximally (figure 1). Biopsies were consistent with a tubulovillous adenoma. A contrast-enhanced CT scan of the abdomen and pelvis revealed a lobulated mass in

Figure 1 Diffuse, non-obstructing circumferential polypoid, friable mass involving the rectosigmoid colon and extending from the proximal anal canal to 30 cm proximally.

To cite: Hashash JG, Holder-Murray J, Aoun E, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009208

Figure 2 Lobulated mass in the rectosigmoid colon.

the rectosigmoid colon without associated adenopathy or concerning metastatic lesions (figure 2). Because the cause of diarrhoea was established, hormonal work-up to exclude other causes of secretory diarrhoea were deferred.

TREATMENT

The patient underwent a proctosigmoidectomy via a transanal and transabdominal approach with coloanal anastomosis and diverting loop ileostomy. The final surgical histopathology revealed a giant villous adenoma 17 cm in size with high-grade dysplasia. No neoplasia was identified.

OUTCOME AND FOLLOW-UP

The patient's postoperative course was smooth and uneventful. At the 10-month follow-up postoperatively, the patient had no further episodes of diarrhoea.

DISCUSSION

In 1954, McKittrick and Wheelock first described secretory diarrhoea developing secondary to a villous adenoma; this correlation resulted in the naming of McKittrick-Wheelock syndrome.² Patients affected with this syndrome can develop a depletion syndrome characterised by severe dehydration, hyponatraemia, hypokalaemia and metabolic acidosis.¹ Diarrhoea may be present for years and is initially compensated for by renal adaptation and increased oral intake. As the compensatory mechanisms become exhausted, life-threatening dehydration and electrolyte disturbances develop.³

The majority of reported cases were due to villous adenomas, with some cases secondary to adenocarcinoma, and one case due to an incarcerated rectal prolapse.⁴ About 3% of villous adenomas have secretory activity. Secretory villous adenomas associated with depletion syndrome are large, ranging from 7 to 18 cm in their greatest dimension.⁵ Prostaglandin E₂ (PGE₂) has been implicated as a possible secretagogue in the pathogenesis of this syndrome.⁶ In patients with rectal villous adenoma,

rectal effluents demonstrate PGE₂ levels that are 3–6 times higher than normal.⁷ Treatment is usually surgical, targeting resection of the adenoma, most commonly with a laparoscopic approach, which leads to near resolution of symptoms in the majority of cases.⁸

In the present case, the biopsies revealed tubulovillous adenoma, which was the cause of chronic diarrhoea. The patient underwent a proctosigmoidectomy and at the 10-month follow-up had no further episodes of diarrhoea. This case describes an important condition that all physicians should be aware of, McKittrick-Wheelock syndrome, or depletion syndrome. This case highlights the importance of being aware of McKittrick-Wheelock syndrome and the need to include this diagnosis in the work up of patients with chronic diarrhoea, as a missed diagnosis will lead to recurrent episodes of dehydration and electrolyte imbalances, and eventually, detrimental outcomes.

Learning points

- ▶ The McKittrick-Wheelock syndrome, a rare disorder, is caused by fluid and electrolyte hypersecretion from a rectal tumour.
- ▶ Patients affected by this syndrome can develop a depletion syndrome characterised by severe dehydration, hyponatraemia, hypokalaemia and metabolic acidosis.
- ▶ Although rare, it is important to be aware of the McKittrick-Wheelock syndrome in patients with chronic diarrhoea; left untreated, it can lead to recurrent episodes of dehydration and electrolyte imbalances and eventually, detrimental outcomes.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Jordan GL, Erickson E. Villous papilloma of the rectum with severe fluid and electrolyte depletion. *AMA Arch Surg* 1958;77:248–52.
- 2 McKittrick LS, Wheelock FC Jr. *Carcinoma of the colon*. Springfield, IL, Charles C Thomas, 1954;61–3.
- 3 Winstanley V, Little MA, Wadsworth C, *et al*. The McKittrick-Wheelock syndrome: a case of acute renal failure due to neoplastic cholera. *Ren Fail* 2008;30:469–73.
- 4 Lee YS, Lin HJ, Chen KT. McKittrick-Wheelock syndrome: a rare cause of life-threatening electrolyte disturbances and volume depletion. *J Emerg Med* 2010;43:e171–3.
- 5 Nusko G, Mansmann U, Altendorf-Hofmann A, *et al*. Risk of invasive carcinoma in colorectal adenomas assessed by size and site. *Int J Colorectal Dis* 1997;12:267–71.
- 6 Smelt AH, Meinders AE, Hoekman K, *et al*. Secretory diarrhea in villous adenoma of rectum: effect of treatment with somatostatin and indomethacin. *Prostaglandins* 1992;43:567–72.
- 7 Older J, Older P, Colker J, *et al*. Secretory villous adenomas that cause depletion syndrome. *Arch Intern Med* 1999;159:879–80.
- 8 Targarona EM, Hernandez PM, Balague C, *et al*. McKittrick-Wheelock syndrome treated by laparoscopy: report of 3 cases. *Surg Laparosc Endosc Percutan Tech* 2008;18:536–8.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow