CASE REPORTS

McKittrick-Wheelock Syndrome – a Rare Cause of Acute Renal Failure

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Abstract

Aim. Fluid and electrolyte hypersecretion in the villous adenoma of the rectum is presented in the case of a 74 year old man presenting with a severe fluid imbalance.

Case report. The patient had a 2-year history of mucous diarrhea and, on admission, presented prerenal uremia, hyponatremia and severe hypokalemia. At sigmoidoscopy, a 6/4 cm villous adenoma of the rectum was found. The increased loss of volume, followed by exhaustion of the physiological compensation mechanisms, led to a life-threatening hypokalemia, as well as to acute renal failure. Conservative treatment was followed by a temporary improvement of the renal function. Alternative treatment was: endocavitary irradiation, endoscopic resection and radical tumor surgery. The surgical removal of the adenoma led to complete recovery of the symptoms.

Conclusion: The McKittrick-Wheelock syndrome can be a problem of difficult diagnosis, both for the gastroenterologist and also for the nephrologist. The patient may develop severe complications, which require a sustained treatment.

Key-words

Rectal villous adenoma - acute renal failure - secretory diarrhea - hypopotasemia

Rezumat

Scop. Hipersecreția de fluide și electroliți determinată de un adenom vilos rectal este ilustrată de cazul unui pacient de 74 de ani cu dezechilibru hidroelectrolitic major.

Observație clinică. Pacientul prezenta sindrom diareic cronic de 2 ani și, la internare, avea retenție azotată, hiponatremie și hipopotasemie severă. La sigmoidoscopie s-a decelat un adenom vilos rectal cu dimensiuni de 6/4 cm. Prin creșterea volumului de lichid pierdut și epuizarea mecanismelor compensatorii fiziologice s-a dezvoltat hipopotasemie semnificativă și insuficiență renală acută. Măsurile terapeutice conservatoare au ameliorat temporar funcția renală. Alternativele terapeutice erau: iradierea endocavitară, rezecția endoscopică sau intervenția chirurgicală radicală. Rezecția chirurgicală a adenomului a dus la recuperare completă.

Concluzie. Sindromul Mckittrick-Wheelock poate fi un diagnostic dificil, atât pentru gastroenterolog, cât și pentru nefrolog. Pacientul poate dezvolta complicații severe, care necesită un tratament susținut.

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Introduction

Most patients with colonic adenomatous polyps present unspecific gastrointestinal symptoms or are asymptomatic. The most frequent symptom which can be attributed to colonic polyps and which requires the colonoscopic exploration is rectal bleeding, hidden or clinically manifested, sometimes accompanied by anemia.

Patients with villous adenomas exhibit, in rare cases, secretory diarrhea with considerable loss of fluids and electrolytes, potentially fatal. The tumors causing this syndrome are always located at the rectum or sigmoid level and are usually of a large size (over 3-4 cm diameter).

Case report

We present the case of a 74 year old patient hospitalized in the Gastroenterology Clinic of the Elias Hospital in February 2004, with watery diarrhea (10-15 stools/24 hours), preceeded by colicative pains at the lower abdominal level, and associated with nausea and vomiting. These symptoms

had been present for a week and occurred on the background of a chronic diarrhea syndrome. The personal pathological history showed Biermer anemia and atrophic gastritis, known since 1999, and a villous rectal tumor, diagnosed 3 years ago (2001), and resected endoscopically twice.

At examination, the patient had no fever, was hemodynamically stable, but was dehydrated and had a distended abdomen, sensitive to palpation, especially at the inferior abdominal level. In order to rule out intestinal obstruction, abdominal radiography was performed, which showed important gas distension, especially of the colon, but also of some ileal loops, without pneumoperitoneum or typical hydroaeric images.

Biological tests evidenced leucocytosis and inflammatory syndrome, worsening prerenal uremia associated with increasing hyponatremia and hypopotasemia, the serum potassium with values of 2.9mEq/l (normal values 3.5-5mEq/l). The stool bacteriology was positive for Pseudomonas aeruginosa, sensitive to penemas and cephalosporins.

The diagnosis was of acute enterocolitis with Pseudomonas, complicated with prerenal azotemia dehydration syndrome. A treatment of dyselectrolytemia and antibiotics were initiated.

The rectosigmoidoscopy revealed at the level of the rectum a villous adenoma, sessile, size 4/6 cm (Fig.1), from which multiple biopsies were taken; apart from that, the endoscopic aspect was normal up to 40 cm from the anus. The histopathological examination diagnosed a low secreting villous adenoma structure with moderate and severe areas of epitelium dysplasia (Fig.2). The tumor was also visualized at the ultrasound examination inside the distended rectal lumen as a hyperecogenic, well defined, nonhomogeneous area (Fig.3); the aspect was confirmed by computerized tomography, which did not reveal adjacent adenopathy or signs of local invasion.

The final diagnosis was: secretory villous adenoma of the rectum with severe dysplasia, complicated with chronic diarrhea – the McKittrick-Wheelock syndrome; acute enterocolitis with Pseudomonas aeruginosa; acute renal failure due to dehydration syndrome; dynamic ileus secondary to hypopotasemia.

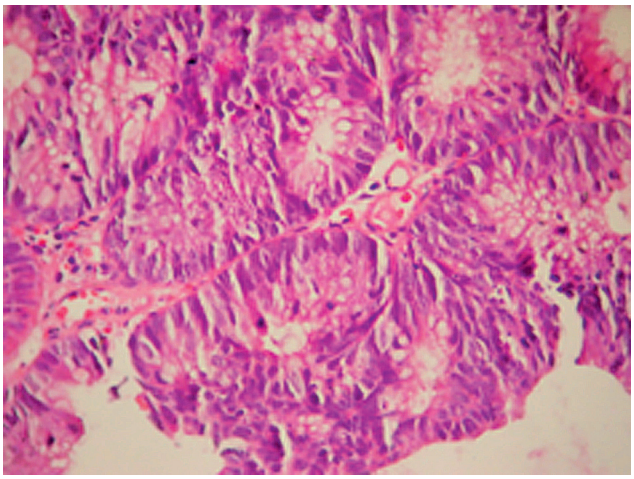

Fig.2 Hystopathological examination: villous adenoma, with mild and severe dysplasia.

The evolution of the patient was unfavourable, because of the persistence of diarrhea and worsening of azotemia and hypopotasemia (2.4 mEq/l), despite substitution therapy and antibiotherapy, and required surgical intervention. Abdominoperineal rectum amputation was performed with terminal colostomy. The curative surgery was temporized almost 2 weeks because of the patient's refusal to have a definitive iliac colostoma.

The postsurgical recovery was difficult, with lingering infectious complications, tangled with toxicoseptic shock and persistence of renal failure. Hypopotasemia at the beginning did not respond to treatment, probably because of the secondary hyperaldosteronism (it improved after administration of spironolactone) and its clinical expression were arrhythmias (atrial tachycardia 2/1). The evolution was favourable, with slow improvement of the clinical and biological state, eventually resulting in full recovery.

Fig.1 Rectosigmoidoscopy: villous rectal tumor.

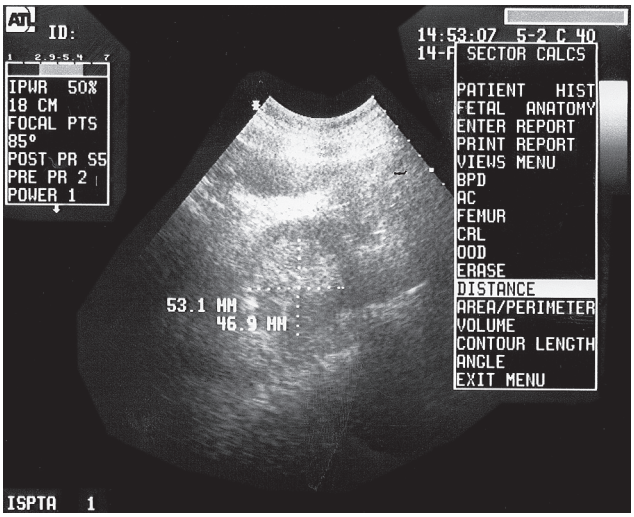

Fig.3 Abdominal ultrasound: hyperecogenic structure, well delimited, inside the rectal lumen.

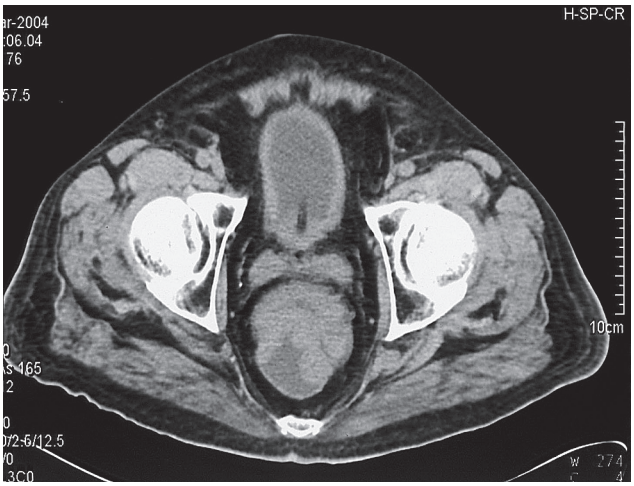

Fig.4 Abdominopelvic CT: villous tumor inside the digestive lumen; no adenopathies or signs of local invasion.

Fig.5 Surgical specimen.

Discussion

Colonic polyps can be divided into two major categories: neoplastic, represented by adenomas and carcinomas, and non-neoplastic (1). By definition, all colorectal adenomas are dysplastic. Dysplasia may be mild, moderate or severe (1-3). Some polyps display the whole spectrum of dysplasia, but the adenoma is always classified according to the most advanced stage. In our patient, the villous adematous polyp exhibited severe dysplasia, which, together with in situ carcinoma, belongs to the category of high-grade dysplasia, with highest risk to evolve towards malignancy. In necroptic series (4), only 13-16% of adenomas are larger than 2 cm diameter and in most cases these are found in distal colonic segments.

Adenomatous polyps have clinical relevance because of their malignant potential. The main characteristics related to the potential of neoplastic evolution are their size, hystological type and degree of dysplasia (1). At the hystopathological examination of specimens from surgical or colonoscopic polypectomies (3-5), it can be observed that the lesions with the highest risk are represented by the large adenomas with villous pattern and a high degree of dysplasia, i.e. "adenomas with advanced pathology" (1). This was the case of our patient.

It is generally accepted that most colonic neoplasms derive from benign adenomas. It was estimated that the doubling rate of the adenoma, even in those growing fast, is of minimum 4-6 months (2). Moreover, for a period of 10 years, there is a risk of 2.5% for an adenoma to evolve to colon cancer (3), but the risk is higher if the tumor is large and villous. It is estimated that the adenoma with severe dysplasia becomes malignant over 3-4 years (1,2). Our patient was diagnosed 3 years before as having a villous tumor, and presently the hystopathological examination exhibited severe dysplasia, without areas of adenocarcinoma. The computed tomography did not identify signs of locoregional invasion.

This case meets the diagnostic criteria of the McKittrick-Wheelock syndrome (6-8), namely secretory diarrhea with severe electrolyte and fluid depletion syndrome ("cholera-like"), associated with large rectal villous adenoma (> 3-4 cm diameter). Biological investigations revealed hyponatremia, hypopotasemia, hypochloremia, metabolic acidosis and dehydratation, indicating acute renal failure (9). From the physiopathological point of view, the villous adenoma is accompanied by excessive water and sodium secretion and an important potassium secretion (8), generated by prostaglandin E_2. Recent clinical studies showed the decrease of prostaglandin E_2 levels after administration of indomethacin and somatostatin (8,10).

Untreated secretant villous adenoma shows a mortality of 100% (6). Therapy consists of hydroelectrolyte balance correction, endocavitary irradiation, endoscopic resection or surgical removal.

Hydroelectrolyte rebalancing is a compulsory step before curative therapy. In our case, hypopotasemia was refractory to the substitution therapy and renal failure was persistent, even after surgery. The endocavitary irradiation was not the best solution, taking into account the biological status which imposed a radical attitude. A study on irradiation results showed a lower response to radiotherapy and a recurrence rate of 32% for rectal adenomas, as compared with adenocarcinomas (11). In our patient, an endoscopic resection was made, which implies a possible recurrence of the tumor and of the symptomatology due to secretion of prostaglandin E_2.

Particularities of our patient were the latency period in the evolution of the McKittrick-Wheelock syndrome and the clinical presentation as pseudoobstruction due to dynamic ileus and not to tumoral obstruction. The hospitalization was precipitated by the association of Pseudomonas enterocolitis, on the background of a chronic diarrhea syndrome, which determined major hydroelectrolytic imbalance and potassium loss. Postoperative maintenance of hypopotasemia was probably due to secondary hyperaldosteronism, as antialdosteronic treatment was followed by a favorable response. Subsequently, the evolution was slowly favorable with the improvement of

symptomatology and complete recovery of the renal function.

In conclusion, the McKittrick-Wheelock syndrome can be a difficult diagnosis, both for the gastroenterologist and also for the nephrologist. The syndrome may lead to severe complications, which require sustained treatment.

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