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Letters to the Editor

McKittrick-Wheelock syndrome - Prolapsed giant villous adenoma of the rectum

Key words: Villous adenoma. Rectal prolapse.

Dear Editor,

Degenerate hypersecretory villous adenoma of the rectum is an uncommon, life-threatening condition. Its diagnosis is clinically challenging since cardinal symptoms are neurological and metabolic in nature because of significant fluid and electrolyte disorders induced by severe diarrhea. Even though the condition was first described back in 1954 its pathophysiology remains unknown. Management is surgical and includes lesion excision once renal function and fluid-electrolyte parameters are brought back to normal.

Case report

A 76-year-old female patient was admitted to the surgery department because of anemia, ascites, overall weakness, neurological stupor, and a prolapsed, irreducible rectal mass. The patient had suffered from mucous diarrhea, rectal bleeding, and rectal prolapse for years. Physical examination revealed a prolapsed, incarcerated rectal growth (Fig. 1) that was 10-cm long and secreted a considerable amount of mucus. Laboratory tests showed anemia with 6 g/dL hemoglobin, 18 % hematocrit, as well as acute renal failure (BUN, 158 mg/dL, creatinine, 1.94 mg/dL), hyponatremia and hypokalemia (sodium, 126 mEq/L; potassium, 2.6 mEq/L). An urgent computerized tomography (CT) scan of the abdomen was performed, which revealed a large prolapsed rectal mass, ascites, and multiple liver metastases, some of them with necrosis. Because of severe pain from the prolapsing tumor, and abundant mucorrhea and rectorrhagia, the patient was surgically managed following the correction of renal failure, and underwent a palliative transanal resection. She died 24 hours after surgery from cardiorespiratory complications.

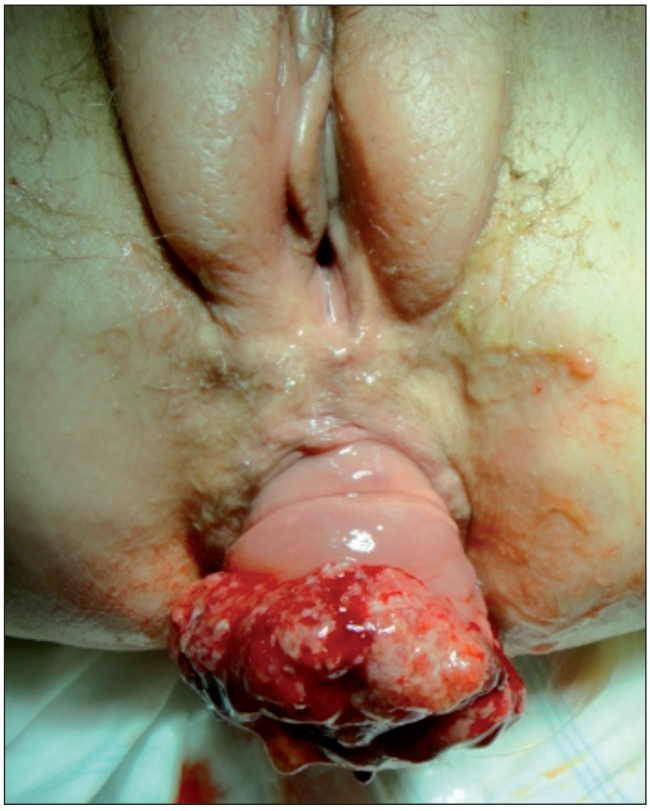

Fig. 1. Appearance of the huge, friable, prolapsed adenoma with abundant mucus secretion.

Discussion

Hypersecretory villous adenoma of the rectum, initially described by McKittrick and Wheelock (1) in 1954, presents with a picture characterized by the following triad: a) severe fluid-electrolyte imbalance (pre-renal acute renal failure, metabolic acidosis, neurological signs and symptoms secondary to ionic imbalance); b) copious hypersecretory mucorrhea; and c) presence of a giant villous adenoma of the rectum or rectosigmoid. This is a rare condition that accounts for 3 % of villous adenomas and most commonly occurs in the seventh decade of life (2). Globular-looking cells loaded with mucine stand out in the histological examination, and malignant transformation may occur in 20-30 % of cases (3). According to size and location they may manifest with subocclusive symptoms (higher tumors) or tenesmus (tumors near the anus) (4), but we found no presentation as a prolapsed lesion in the literature.

From a pathophysiological standpoint these hypersecretory adenomas show high levels of cyclic AMP and adenylyl-cyclase. Prostaglandin E2 (PGE2) is the mediator involved in the development of hypersecretory diarrhea, as PGE2 levels are up to three times higher than those seen in non-hypersecretory adenomas (5).

Initial management must include aggressive fluid-electrolyte replacement, a key measure to recover from severe kidney and metabolic disorders. Significant volume loss must be corrected. The use of 400 mg/day indomethacin may be temporarily indicated given its inhibitory action on PGE2 (4) to reduce the adenoma's secretory component. However, the use of indomethacin should be carefully considered because of the risk of exacerbating renal failure (6).

Its use as a therapeutic agent must be envisaged as a "bridge" therapy towards surgical resection, which is the lesion's definitive treatment (7). Endoscopic resection is indicated for smaller adenomas, bigger tumors must be surgically resected. The technique of choice is dependent upon distance to the anal margin, size, percentage of circumference involved, and rectal wall invasion depth.

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