Rectal malakoplakia

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ABSTRACT

Malakoplakia is a rare chronic granulomatous disease with potential to affect multiple organs. Gastrointestinal involvement is considered one of the most common locations outside of the urinary tract. Herein, we present the case of a man who presented to our clinic with painless hematochezia. Physical examination findings were unremarkable; however, colonoscopy revealed a frond-like/villous, fungating, and polypoid nonobstructing circumferential mass in the rectum. Histopathological examination of the lesion revealed findings consistent with malakoplakia.

KEYWORDS Colon; colonoscopy; macrophages; malakoplakia; Michaelis-Gutman bodies

Malakoplakia is a rare chronic granulomatous condition that may manifest in multiple organs. Presentation can vary depending on the organ involved, presenting a diagnostic challenge. The gastrointestinal tract is the second most common site of involvement after the urinary tract. The most common sites of colonic involvement are the rectum, sigmoid, and right colon, in descending order of frequency. The proposed predisposing factors include localized infection, immunosuppression, neoplasia, and systemic illness.¹ Herein, we present the case of a man who presented with complaints of painless hematochezia and was diagnosed with malakoplakia through rectal mass biopsy during colonoscopy.

CASE DESCRIPTION

A 33-year-old man presented to our clinic with multiple episodes of painless hematochezia following initiation of warfarin for recently diagnosed lower-extremity deep venous thrombosis. He had a complex neurosurgical history, which included resection of a right temporal glioblastoma multiforme tumor followed by chemotherapy and radiation. He also underwent subsequent craniotomy with ventriculo-peritoneal shunt placement and was placed on oral dexamethasone for recurrent cerebral edema related to a residual brain tumor. Despite discontinuation of warfarin, the patient continued to have multiple episodes of bright red blood per rectum. Digital rectal exam revealed bright red blood on the

gloved exam finger. Follow-up colonoscopy was significant for a frond-like/villous, fungating, and polypoid nonobstructing circumferential mass in the rectum (*Figure 1a*). Biopsies were suggestive of inflammatory changes and malakoplakia. After prolonged discussion, a decision was made to continue dexamethasone in order to minimize cerebral edema related to the residual brain tumor. Family members opted to forego a proposed bowel resection. He was started on a prolonged course of ciprofloxacin; however, he developed severe sepsis as a result of a urinary tract infection attributed to extended-spectrum beta-lactamase *Escherichia coli* infection and died shortly thereafter.

DISCUSSION

Malakoplakia was first described by Michaelis and Gutmann in 1902 after it was observed in the bladder. It was named by Van Hanseman in 1903. The term *malakoplakia* stems from the Greek *malakos* (soft) and *plakos* (plaque) and reflects its usual appearance as a friable, yellow mucosal lesion on endoscopy. Coliform bacteria in macrophage cytoplasm (von Hanseman histiocytes) (*Figure 1b,c*) and laminated intracytoplasmic inclusion bodies (Michaelis-Gutmann bodies) (*Figure 1c*) are considered diagnostic features. It is hypothesized that a defect of macrophage phagolysosomal response to bacterial infection is the cause of malakoplakia. Glucocorticoids and azathioprine have been shown to contribute to macrophage abnormality, and their

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Figure 1. (a) Colonoscopy showing a frond-like/villous, fungating, and polypoid nonobstructing circumferential mass in the rectum. (b) CD68 stain highlighting the sheets of macrophages. (c) Hematoxylin and eosin stain showing numerous histiocytes and Michaelis-Gutmann bodies.

discontinuation has been shown to reverse macrophage abnormalities and clinical symptoms. The presence of a bacterial infection has been described in most reported cases, regardless of the affected organ. The two most commonly isolated organisms include *Klebsiella pneumoniae* and *Escherichia coli*, but other organisms including gram-negative, gram-positive, and acid-fast bacilli have also been isolated.¹ Fungi, including *Paracoccidioides brasiliensis*, and viruses have also been implicated in isolated cases.²

Malakoplakia can present with a variety of symptoms, but the most common complaints include abdominal pain, fever, diarrhea, and rectal bleeding.³ None of these symptoms are specific; therefore, diagnosis is often incidental, as was the case in our patient. The most susceptible include the immunocompromised (posttransplant patients, patients with AIDS or diabetes mellitus) and those with conditions requiring immunosuppressive therapy, as was the case with our patient, who was on a prolonged course of steroids for cerebral edema.⁴

Given the rarity of the condition, there are no standard treatment guidelines. However, the mainstay of treatment involves antibiotic therapy and surgical resection. Preferred antibiotics are those with high macrophage concentration, as seen with quinolones and trimethoprim-sulfamethoxazole. Both antibiotics appear to kill the bacteria associated with malakoplakia and can penetrate the defective host macrophages. Prior cases describe successful treatment of colonic malakoplakia with a prolonged course of sulfamethizole and trimethoprim.⁵ A few case reports also describe ascorbic acid as effective.^{6,7} The duration of treatment can vary, depending on the extent of the infection. In this case, the patient was prescribed ciprofloxacin, which has been shown to have excellent penetration into macrophages.⁸ He was compliant with the medication but developed severe sepsis as a result of a urinary tract infection attributed to extended-spectrum beta-lactamase *Escherichia coli* infection.

Rectal malakoplakia continues to be rare, with only a limited number of cases describing gastrointestinal

involvement. Proper histologic examination is imperative for a correct diagnosis. This is a rare case in which malakoplakia was diagnosed preoperatively on biopsy samples. Awareness of colonic and rectal malakoplakia is important in patients taking steroids, as illustrated in this case. Given the potential for significant morbidity if left untreated, malakoplakia should be included in the differential diagnosis in immunosuppressed patients with abnormal lesions on colonoscopy.

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1. Han SH, Joo M, Chang S, Kim HS. Malakoplakia affecting the umbilical cord. *J Pathol Transl Med*. 2015;49(2):177–179. doi:10.4132/jptm.2015.02.04.
2. Pillay K, Chetty R. Malakoplakia in association with colorectal carcinoma: a series of four cases. *Pathology*. 2002;34(4):332–335. doi:10.1080/003130202760120481.
3. Weinrach DM, Wang KL, Cisler JJ, Diaz LK. Pathologic quiz case: a 54-year-old liver transplant recipient with diffuse thickening of the sigmoid colon. Malakoplakia of the colon associated with liver transplant. *Arch Pathol Lab Med*. 2004;128(10):e133–e134.
4. Damjanov I, Katz SM. Malakoplakia. *Pathol Annu*. 1981;16(Pt 2):103–126.
5. Christensen M, Knuhtsen S, Knudsen E. Malokoplaki i colon behandlet med sulfamethizol og trimetoprim [Colonic malakoplakia treated with sulfamethizole and trimethoprim]. *Ugeskr Laeger*. 2011;173(7):509–510.
6. Fudaba H, Ooba H, Abe T, et al. An adult case of cerebral malakoplakia successfully cured by treatment with antibiotics, bethanechol and ascorbic acid. *J Neurol Sci*. 2014;342(1-2):192–196. doi:10.1016/j.jns.2014.05.006.
7. Vattimo A, Lupinacci RA, Kerzner A, da Silva JH. Malacoplasia do intestino grosso, bexiga e retroperitônio: relato de um caso [Malakoplakia of the large intestine, bladder and retroperitoneum: a case report]. *AMB Rev Assoc Med Bras*. 1990;36(3-4):153–156.
8. van Furth R, van't Wout JW, Zwartendijk J, Wertheimer PA. Ciprofloxacin for treatment of malakoplakia. *Lancet*. 1992;339(8786):148–149. doi:10.1016/0140-6736(92)90212-L.