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Malakoplakia of the large intestine: an incidental extremely rare finding

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SUMMARY: **Malakoplakia of the large intestine: an incidental extremely rare findingy.**

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Malakoplakia is a rare inflammatory disease, most commonly found in the urinary tract. It appears be related to a functional deficiency of macrophages, resulting in an inability to destroy digested bacteria and it is associated with various conditions that cause immunodeficiency. A rare case of malakoplakia of the colon in a healthy 68-year old male is presented. The patient underwent emergency surgery with colon resection and an end stoma with closure of the distal bowel (Hartmann's procedure), due to incarcerated ventral hernia and sigmoid-colon rupture. He underwent reversal of the Hartmann's procedure four months after the initial operation. The histological examination from the anastomotic rings revealed Michaelis–Gutmann bodies that are pathognomonic of malakoplakia. He received per os ciprofloxacin, bethanecol and ascorbic acid for 12 months. Follow-up endoscopy did not exhibit any signs of the disease. A case of a healthy patient presenting with malakoplakia without any underlying disease that causes immunodeficiency is extremely rare. Treatment of malakoplakia involves the eradication of microorganisms. Cholinergic agonists, such as bethanechol and ascorbic acid, as well as antimicrobial treatment with trimpethoprim/sulphamethoxazol and rifampicin are most commonly being used. Long-term antimicrobial treatment has been reported (6 months to 3 years).

KEY WORDS: Malakoplakia - Gastrointestinal tract - Colon - Michaelis-Gutmann bodies.

Introduction

Malakoplakia is a term of Greek origin, deriving from "malakos" and "plaka" meaning soft and plaque respectively. It is a rare granulomatous disease first documented in 1902 (1-3). Malakoplakia is most commonly observed in the urinary tract, while the gastrointestinal tract is involved in less than 10% of the reported cases (4, 5).

The pathogenesis of the disease is even nowadays not entirely clear. Malakoplakia appears to be related to functional deficiency of macrophages, resulting in inability to destroy digested bacteria. Furthermore, it is associated with many conditions that cause immunodeficiency, such as organ transplantation, chronic corticosteroid use, chemotherapy, sarcoidosis, malnutrition, ulcerative colitis and HIV (6, 7).

A rare case of a healthy 68-year male, undergoing reversal of Hartmann's procedure, diagnosed from the histological examination with malakoplakia is presented.

Case presentation

A 68-year old healthy male presented to the emergency department of the "Sismanoglion" General Hospital of Athens, Greece due to acute abdominal pain. He was diagnosed with incarcerated ventral hernia and rupture of the sigmoid colon. He underwent emergency surgery with colon resection and an end stoma with closure of the distal bowel (Hart-

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mann's procedure). The patient received intravenous antimicrobial treatment with cefuroxime and metronidazole. He had a satisfactory recovery and was discharged on the 9th postoperative day.

After 4 months the patient complained of light abdominal pain. It was decided to proceed to an elective stoma take-down. His preoperative laboratory findings were within normal limits. At that point, he underwent reversal of the Hartmann's procedure. The histological examination of the anastomotic rings revealed collections of granular histiocytes within the muscularis propria (Figure 1). Several histiocytes contained intracytoplasmic round bodies (Michaelis–Gutmann bodies) of varying size (Figure 2). Based on these findings the patient was diagnosed with malakoplakia.

The patient received per os ciprofloxacin 500 mg/12 hours, as well as bethanecol 25 mg/8 hours and ascorbic acid 60 mg/24 hours for a total of 12 months. After 1.5 month of treatment the symp-

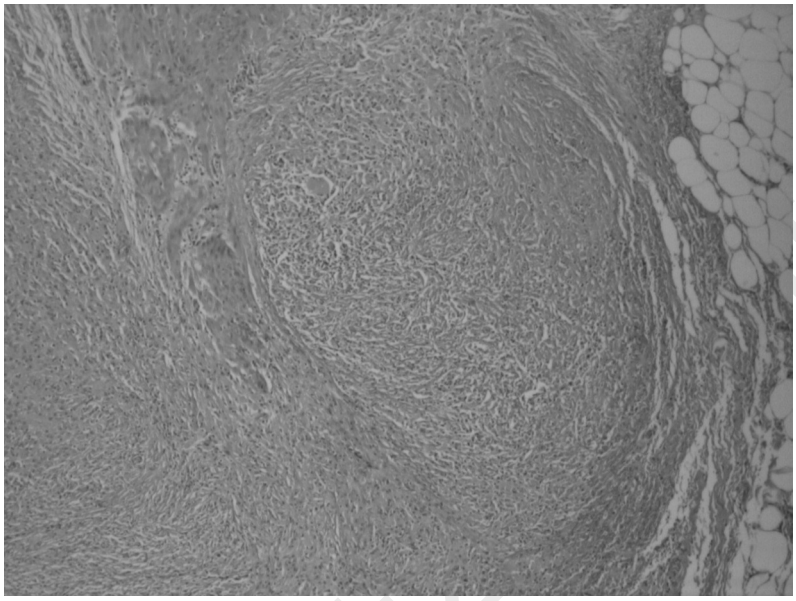

Figure 1 - HEx10: Collections of histiocytes within muscularis propria.

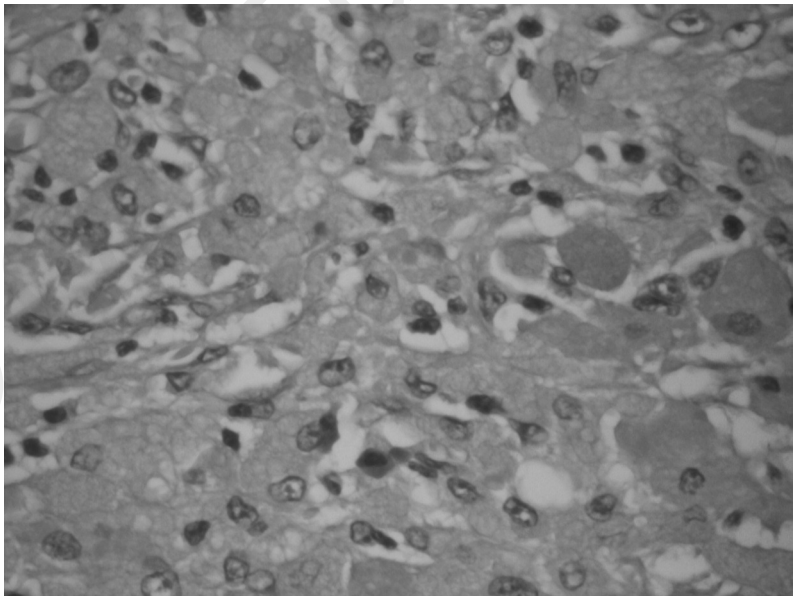

Figure 2 - HEx40: Michaelis-Gutmann bodies of varying size.

toms of abdominal pain disappeared. During this treatment he showed no drug-related side effects.

A whole year after Hartmann's reversal, the patient had a colonoscopy. Histological examination did not reveal signs of malakoplakia. Furthermore, chronic conditions, such as Human Immunodeficiency Virus infection, sarcoidosis, ulcerative colitis, Crohn's disease or Whipple disease that cause immunosuppression and have been related with malakoplakia were ruled out.

The outpatient follow up, 2.5 years until today, has confirmed that the patient remains healthy, without any signs or symptoms of an underlying disease.

Discussion

Malakoplakia is a chronic granulomatous disease, characterized by the accumulation of distinctive macrophages containing the pathognomonic intracytoplasmic structures, the Michaelis-Gutmann bodies (8). From 1902 until today about 450 cases have been described, with less than 10% of those referring to the gastrointestinal tract. A case of a healthy patient without any underlying disease that causes immunodeficiency, like the present one, is extremely rare (3, 6). To our knowledge in Greece there have only been 2 cases of large-intestine malakoplakia reported (9, 10).

Most cases of gastrointestinal tract malakoplakia involve the colon and rectum (11). Clinically it can present with diarrhea, abdominal pain, intestinal obstruction, rectal bleeding, nausea and vomiting, constipation, fever or it can be asymptomatic. Therefore, there are not any characteristic signs and symptoms (12, 13).

The present patient complained, after the Hartmann's procedure, of light abdominal pain. Diagnosis is possible only with histological examination and the presence of the Michaelis-Gutmann bodies (1-3). Therefore, it is extremely difficult to include this disease within the differential diagnosis.

Endoscopically, malakoplakia presents in the early stages as soft yellow to tan mucosal plaques or in the late stages as raised, grey to tan lesions in various sizes with peripheral hyperemia and a central depressed area (14). The reported patient had a follow-up colonoscopy, 12 months after stoma reversal, showing no such sings. Additionally, the obtained biopsies did not reveal the Michaelis-Gutmann bodies that are pathognomonic for malakoplakia.

The pathogenesis of the disease is not entirely clear. However, it has been linked to many chronic conditions that lead to immunodeficiency (15). Additionally, chronic infections with *Escherichia coli*, *Proteus mirabilis*, *Mycobacterium tuberculosis*, and *Staphylococcus aureus* have also been associated with the disease (8). It is of note that the described patient had an unremarkable medical history, without any chronic condition that causes immunosuppression.

Generally, malakoplakia should be differentiated from ulcerative colitis, Crohn's disease, sarcoidosis, Whipple disease, Chediak-Higashi syndrome, and malignancies such as colon cancer and lymphoma (2, 6, 8, 16-18).

Treatment of malakoplakia involves the eradication of the microorganisms. Cholinergic agonists, such as bethanechol and ascorbic acid, have been used to increase the intracellular cyclic Guanosine MonoPhospate (cGMP) to cyclic Adenosine MonoPhosphate (cAMP) ratio to improve lysosomal function (8).

Additionally, antimicrobial treatment with trimpethoprim/sulphamethoxazol, rifampicin and most commonly ciprofloxacin have been used. Long-term antimicrobial treatment has been reported (6 months to 3 years) (8).

Our patient, after receiving 1.5 month per os ciprofloxacin, had no signs or symptoms of disease. He continued the treatment for a total of 12 months.

A diagnosis of malakoplakia is extremely difficult. If such a diagnosis is made, from the histological findings, the physician must also focus on finding the underlying condition that is usually associated with malakoplakia, since this is the most crucial factor affecting mortality.

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Disclosure of interest

The Authors report no conflict of interest.

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