International Surgery Journal
Silva AA et al. Int Surg J. 2024 Apr;11(4):655-657
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20240757

Case Report

Rectal malakoplakia simulating a locally advanced rectal cancer: a case report

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Received: 07 February 2024
Accepted: 06 March 2024

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ABSTRACT

Malakoplakia is a rare and chronic granulomatous disease, resulting from the accumulation of large granular macrophages containing basophilic inclusion bodies in the cytoplasm named Michaelis-Gutmann bodies. The gastrointestinal tract is the second most common site after the urinary tract, though malakoplakia has the potential to manifest in multiple organs. Rectal malakoplakia is difficult to diagnose due to its unspecific clinical and radiological presentation, usually mimicking a malignancy. Most report cases are associated with immunosuppressive diseases or chronic prolonged illness. We present the case of a 64-year-old male with a history of anal pain. A pelvic magnetic resonance imaging showed a rectal mass with 38 mm invading the mesorectum. Colonoscopy was performed confirming a mass-like lesion and biopsies were taken. Histopathological examination revealed features consistent with malakoplakia. The patient underwent long-term oral antibiotic treatment and during follow-up there was a regression of the lesion and resolution of symptoms.

Keywords: Malakoplakia, Rectal malakoplakia, Granulomatous disease, Michaelis-Gutmann bodies, Von Hansemann cells

INTRODUCTION

Malakoplakia or Von Hansemann's disease is an extremely uncommon chronic infectious granulomatous lesion.[1] It was first reported by Michaelis and Gutmann in 1902 and in 1903 named by Von Hansemann.[1-4] The term malakoplakia stems from the Greek *Malakos* (soft) and *Plakos* (plaque) and reflects its usual appearance as a friable and yellow mucosal lesion on endoscopy.[1,2] It is hypothesized that a defect of macrophage phagolysosomal response to bacterial infection is the cause of malakoplakia.[2]

Malakoplakia is most commonly found in the urinary tract but has been reported in the gastrointestinal tract, lung, brain, lymph node, adrenal, tonsil, conjunctiva, skin, bone, abdominal wall, pancreas, retroperitoneum and female genital tract.[3] The most common sites of colonic involvement are the rectum, sigmoid and right colon.[2] Generally, malakoplakia occurs in immunocompromised patients, though some malakoplakia cases in immunocompetent patients has been reported.[3,5] On the other hand, malakoplakia is very rare in young people without underlying disease.[3]

The clinical presentation of rectal malakoplakia is variable and non-specific and imaging findings are very similar with malignancy which turns the definitive diagnosis only possible with the histopathological examination.[1,5] In spite of malakoplakia is a benign disease, in some cases may be accompanied with a malignant tumor.[5]

CASE REPORT

A 64-year-old male patient presented with a history of anal pain. He had multiple diseases such as type 2 diabetes treated with insulin, dyslipidemia, hyperuricemia and obesity. The patient denied rectal bleeding, constipation, diarrhea, vomiting, abdominal pain or weight loss.

Digital rectal examination revealed a palpable mass in the low rectum located at 1 cm from anal verge. The pelvic magnetic resonance imaging (MRI) showed a tumor with 38 mm invading the mesorectum and two suspected lymph nodes in the perirectal fat (Figures 1 and 2). The hypothesis of a locally advanced rectal cancer was raised.

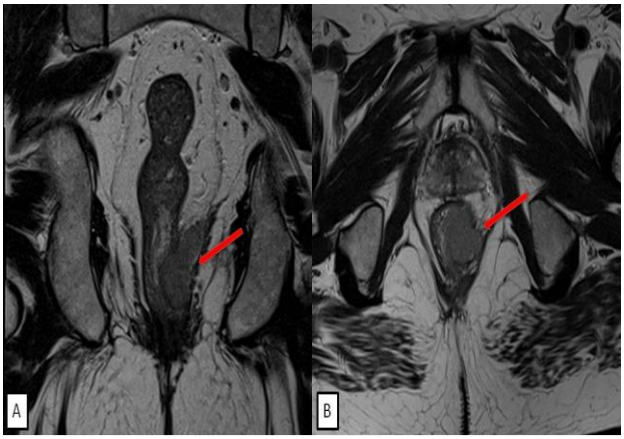

Figure 1 (A and B): Left-sided rectal mass (red arrow) invading the mesorectum on MRI; coronal plane; and transverse plane.

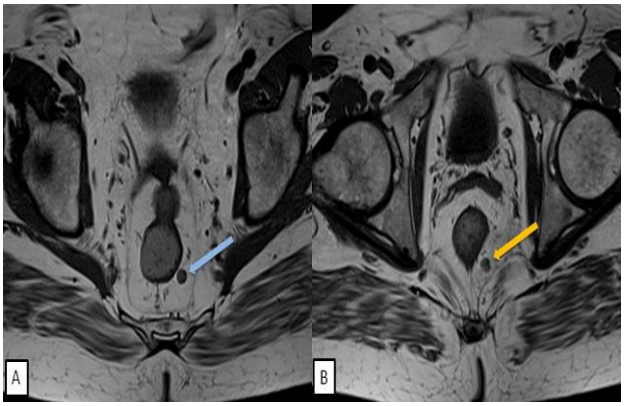

Figure 2 (A and B): MRI showing two hypointense round lymph nodes in the mesorectum (blue and yellow arrows).

Colonoscopy revealed a friable polypoid tumor with 20 mm in diameter and biopsies were taken. Histopathological examination indicated intense histiocyte infiltration with round laminated basophilic inclusion bodies in the cytoplasm (Michaelis-Gutmann bodies), without evidence of malignancy. Immunohistochemistry analysis tested positive for CD68 and CD31, negative for cytokeratin (AE1/AE3) and histiocytes contained intracytoplasmic round bodies reacted positively with periodic acid-Schiff (PAS) staining with diastase resistant. Concomitantly intra-cytoplasmatic calcium and iron was positive. According to histopathological findings the diagnosis of malakoplakia was established. The patient underwent long-term oral antibiotic treatment with trimethoprim-sulfamethoxazole for 120 days and during follow-up there was a regression of the lesion and resolution of symptoms.

DISCUSSION

Malakoplakia is a rare acquired granulomatous condition, more prone to affect females with a prevalence female-to-male ratio of 4:1. Its incidence peaks in middle-aged individuals.[1] The etiology and pathogenesis of malakoplakia is still not fully understood, but multiple possible mechanisms have been suggested.[3] It is hypothesized that a defect of macrophage phagolysosomal response to bacterial infection is the cause of malakoplakia.[1-5] The involvement of microorganisms is supported in patients with malakoplakia who have chronic infections with various organisms such as *Escherichia coli*, *Proteus mirabilis*, *Staphylococcus aureus* and *Mycobacterium tuberculosis*. Nevertheless, other factors contribute to the development of malakoplakia. One of them is the impaired capacity to phagocytosis by macrophages or monocytes due to a defect in lysosomal processing of microorganisms resulting in an incomplete digest of microorganisms leading to its accumulation in lysosomes with mineralization of calcium and iron salts on residual microorganism glycolipids.[3] On the other hand, malakoplakia is frequently associated with systemic diseases such systemic lupus erythematosus, tuberculosis, diabetes mellitus, sarcoidosis and neoplasms. Thus, nearly 40% of patients have primary or acquired immunosuppressed states.[4] In this case report, the patient had a long-standing type 2 diabetes treated with insulin but poorly controlled which probably predisposed him to an acquired immunodepression state.

The gastrointestinal tract is the second most common site of involvement by malakoplakia being rectum and colon more frequently reported.[2,3] The clinical presentation of rectal malakoplakia is unspecific, ranging from asymptomatic to diarrhea, abdominal pain or discomfort, rectal bleeding, constipation, anorexia, and intestinal obstruction.[1,2] None of these symptoms were revealed by the patient of our case report. The main symptom of this patient was anal pain which is rarely described in rectal malakoplakia.

The diagnosis of rectal malakoplakia is generally made by histopathological examination. It is defined by characteristics features of accumulated histiocytes (Von Hansemann cells) with abundant granular basophilic inclusions that are PAS positive and diastase resistant.

Michaelis-Gutmann bodies are also characteristic, corresponding to calcified detritus of incompletely digested bacteria mineralized by iron and calcium deposits within phagolysosomes (positive to Von Kossa calcium or iron staining).[1] Endoscopically, rectal malakoplakia commonly presents as three gross forms: unifocal lesions, widespread mucosal multinodular involvement and large mass lesions.[4] Usually, the appearance of these lesions' mimic malignancy on endoscopy.[4,5]

Medical treatment of malakoplakia is currently the main treatment and consists of appropriate antibiotic therapy and mitigation of immunosuppressed state.[1] There are also some case reports that association of these treatments with surgery was described.[4-6] It is also advocate by some authors that in cases showing limited involvement of the colon, surgical resection is usually curative and therefore indicated.[4] Otherwise, surgery may also be indicated when pharmacologic treatment is ineffective.[1] However, given the rarity of this condition there are no standard treatment guidelines.[2] Some principles of medical treatment indicate antibiotics with high macrophages concentration, as seen with quinolones and trimethoprim-sulfamethoxazole.[1,2] There are case reports describing the use of cholinergic agonists such as bethanechol to increase intracellular cyclic guanosine monophosphate/ cyclic adenosine monophosphate ratio in macrophages, thus improving lysosomal bactericidal function.[1] Another principle of the medical treatment that is not stablished is its duration, however long-term treatment is advised although its efficacy still remains debated.[2,3] The medical treatment of malakoplakia has usually good results as it is self-limiting and a benign disease.[1] Nevertheless, it is important to be aware that rectal malakoplakia can coexist with malignancy, and in these cases the treatment must follow the current guidelines treatment of rectal malignancy, which could consist in surgical intervention.[1,6] Our patient underwent long-term oral antibiotic treatment with trimethoprim-sulfamethoxazole for 120 days and the follow-up with colonoscopies and imaging revealed a regression of the lesion and clinically the anal pain resolved.

CONCLUSION

Rectal malakoplakia is a rare, benign and granulomatous disease that has the capacity to mimics a malignant rectal lesion. The unspecific clinical presentation and radiological findings leads to suspect of a malignancy rectal disease until the histopathological examination allows the definitive diagnosis of malakoplakia. However, in some cases malakoplakia may be associated with rectal malignancy and in these cases the treatment differs from those in which there is only malakoplakia. Currently, the first-line treatment of malakoplakia consists in long-term antibiotics. Though, more studies are necessary to develop accurate guideline treatments.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Silva AA, Pires GC, Simões LV, Lourenço BR, Rocha CC, Meireles LJ. Rectal malakoplakia simulating a locally advanced rectal cancer: a case report. Int Surg J 2024;11:655-7.