

**Case report**

# **What Lies Beneath – Malakoplakia. A Rare Submucosal Lesion of the Colon**

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## **ABSTRACT**

### **Aims:**

To add to the literature and discuss the clinical significance of the rare finding of colonic malakoplakia

### **Presentation of Case:**

We present a case of a rare submucosal malakoplakia of the ascending colon discovered on colonoscopy.

### **Discussion:**

Malakoplakia is a rare multiorgan granulomatous disease that can be found within the gastrointestinal tract. Its appearance can vary including polyps, ulcerations, plaques, flat lesions, erosions or even large mass lesions. Given its varied endoscopic appearance there exists a broad differential diagnosis including malignancy or pre-malignant neoplasm. Malakoplakia within the gastrointestinal tract has been observed a co-existence with colorectal adenocarcinoma however no direct causal association has been identified.

### **Conclusion:**

Given the possible malignant differentials of colonic lesions, a histologic assessment is required for the diagnosis of malakoplakia. Malakoplakia is generally thought to be a benign condition with no specific follow up required in the gastrointestinal tract.

*Keywords: Malakoplakia, Submucosal, Colon, Gastrointestinal*

## 1. INTRODUCTION

A variety of submucosal lesions can be found in the colon and are often an incidental finding. Most are benign including lipomas, leiomyomas, and lymphoid aggregates. Malignant causes also exist including neuroendocrine tumours and metastatic lesions. Soft lesions generally represent lipomas, confirmed by a positive "pillow sign". However, accurate diagnosis of a firm submucosal lesion requires histologic assessment. We present a case of a rare benign firm submucosal lesion discovered on colonoscopy.

## 2. PRESENTATION OF CASE

A 53-year-old female underwent gastroscopy and colonoscopy for investigation of iron deficiency anaemia. There were no gastrointestinal symptoms or history of weight loss. There was no history of immune suppression, autoimmune disease or chronic infection. Gastroscopy was unremarkable. At colonoscopy, one 3mm firm submucosal nodule was found in the proximal ascending colon (Figure 1 Panel A/B). This was resected en-bloc using a cold polypectomy technique. No other abnormalities were found.

Histology demonstrated a well-defined lesion in the mucosa and submucosa composed of abundant histiocytes admixed with small numbers of lymphocytes, plasma cells, neutrophils and rare eosinophils. Many of the histiocytes contain characteristic Michaelis-Gutmann bodies which are Periodic acid-Schiff positive (Figure 1 Panel C/D).

## 3. DISCUSSION

Malakoplakia is a rare granulomatous disease that can involve many organ systems. The exact pathogenesis is poorly understood but is thought to be related to impaired macrophage ability to phagocytose bacteria leading to formation of pathognomonic Michaelis-Gutmann bodies [1]. This dysfunction is hypothesised to be related to abnormalities in the lysosomal system within macrophages. Organisms isolated from malakoplakia lesions include *Escherichia coli*, *Klebsiella pneumoniae*, *Mycobacterium tuberculosis*, *Proteus*, *Rhodococcus equi*, *Staphylococcus aureus* and *Pseudomonas aeruginosa* [2]. There has been an association of malakoplakia with conditions of immune dysregulation [2].

Malakoplakia is most commonly seen in the genitourinary tract followed by the gastrointestinal tract. Less commonly it is also reported to involve the pancreas, lymph nodes, central nervous system, middle ear, tongue, tonsils, conjunctiva, thyroid, bone, skin, prostate, breast, adrenal and respiratory tract [2, 3]. Gastrointestinal malakoplakia is the second most common organ system involved though still an uncommon entity with a limited number of cases reported in the literature, likely due to under recognition and under reporting. The diagnosis can be challenging due to its nonspecific clinical presentation and rarity. Patients can present with symptoms of abdominal pain, diarrhoea and haematochezia, however

in many cases the lesion is asymptomatic and discovered incidentally like in our case [3, 4]. In spite of what its name suggests (derived from the Greek “malakos” soft and “plakos” plaque), the endoscopic appearance can vary including polyps, ulcerations, plaques, flat lesions, erosions or even large mass lesions [3, 4]. Malakoplakia has been identified from the stomach to the anus though most commonly observed in the sigmoid colon and rectum [3]. Given its varied endoscopic appearance there exists a broad differential diagnosis including malignancy or pre-malignant neoplasm.

Malakoplakia in the genitourinary tract has been reported in co-existence with urothelial carcinoma, MALT lymphoma, and prostate cancer [1,4]. Within the gastrointestinal tract, malakoplakia has been observed a co-existence with colorectal adenocarcinoma however no direct causal association has been identified [5]. It has been hypothesised that coincidence of adenocarcinoma with malakoplakia may be a result of the distortion of the local microbiota by the malignancy [6]. Malakoplakia is generally thought to be a benign condition with no specific follow up required in the gastrointestinal tract, though given its co-existence with colorectal cancer, would warrant a careful search for colorectal cancer.

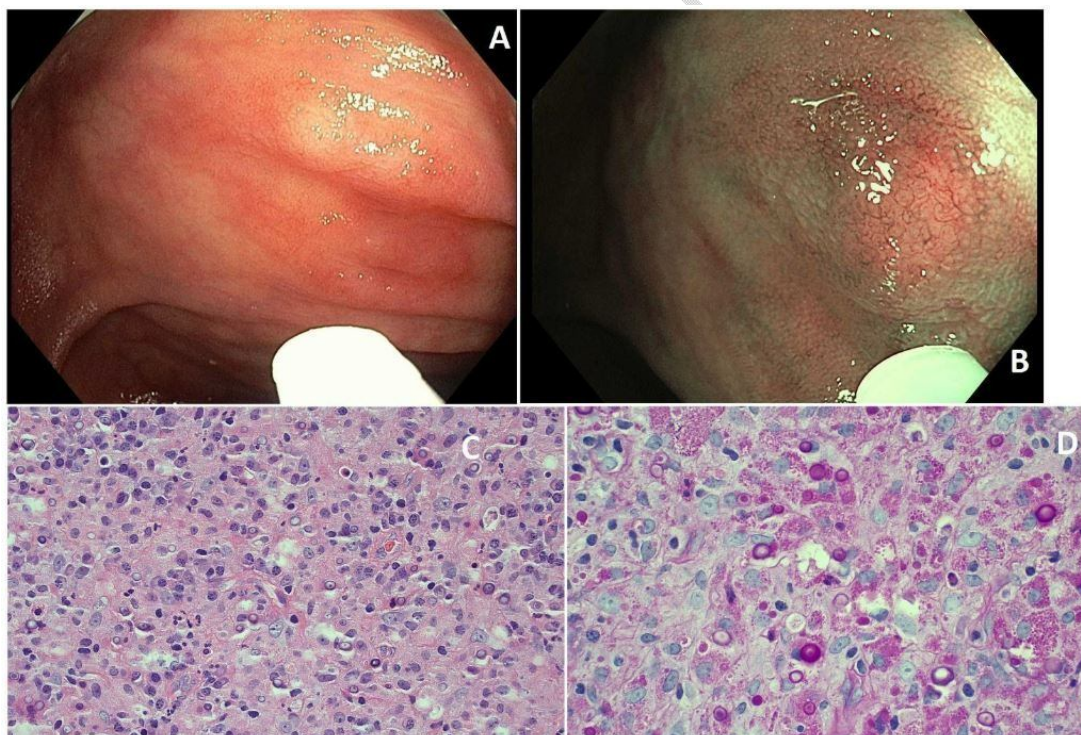


Fig.1. Panel A/B: White light and NBI image of submucosal nodule found in the ascending colon Panel C/D: Periodic acid-Schiff (x40) stain with abundant eosinophilic granular cytoplasm and numerous intracellular Michaelis-Gutmann bodies

#### **4. CONCLUSION**

Given the possible malignant differentials of colonic lesions, a histologic assessment is required for the diagnosis of malakoplakia. Malakoplakia is generally thought to be a benign condition with no specific follow up required in the gastrointestinal tract, though given its co-existence with colorectal cancer, a careful search for colorectal cancer would be warranted.

#### **CONSENT (WHEREEVER APPLICABLE)**

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

#### **ETHICAL APPROVAL (WHEREEVER APPLICABLE)**

Ethics approval was gained from the Metro South Human Research Ethics Committee.

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