

# **Rectal Malakoplakia presenting as a mass and fistulous tract in a renal transplant patient**

Muhammad Yousif, Zaigham Abbas, Muhammad Mubarak

Department of Gastroenterology and Histopathology, Sindh Institute of Urology and Transplantation, Karachi.

### **Abstract**

Malakoplakia (MK) is a rare, chronic inflammatory disorder with characteristic morphologic features. It most commonly involves urogenital organs but can affect any organ system in the body. Gastrointestinal tract is the second common site of involvement. It commonly occurs in non-transplant patients but transplant patients are also vulnerable to it. We present a case report of a forty year old male renal transplant patient, who received a kidney from his brother with 1 haplotype and 4 antigen match. He was on regular post transplant follow up with stable graft

function. Fifteen months post transplant he presented with the complaint of painful defecation, a swelling in the perianal region and inability to sit down properly. Biopsy examination showed malakoplakia with characteristic Michaelis-Gutmann bodies. Culture of the tissue grew E Coli. Immunosuppression therapy was curtailed and patient was started on ciprofloxacin 500mg OD for 6 months. The lesions regressed completely after six months of the above therapy and the patient became completely symptoms free.

### **Introduction**

Malakoplakia (MK) is a rare, chronic inflammatory

disorder with characteristic morphologic features. It most commonly involves urogenital organs but can affect any organ system in the body. Gastrointestinal tract is the second common site of involvement. To date, forty one cases of colorectal malakoplakia have been reported in English language literature, mostly in association with transplantation or malignant tumours. We present a case of MK in a renal transplant patient involving the rectum and perianal region.

### Case Report

A forty year old male patient underwent live related renal transplantation for end stage renal disease secondary to adult polycystic kidney disease (APCKD) on 27-07-2000. Donor was brother with 1 haplotype and 4 antigen match. He made uneventful recovery from transplant surgery and was on regular post transplant follow up with stable graft function. Maintenance immunosuppression consisted of cyclosporine, azathioprine and prednisolone in standard doses. Fifteen months post transplant he presented to GI OPD with the complaints of painful defecation, a swelling in the perianal region and inability to sit down properly. Perianal examination revealed a small nodular lesion about 2 cm in diameter around the anal margin, yellowish brown in colour with central ulceration, everted edges and pussy discharge. It was non tender and no blood oozing was seen. There was also a perianal fistula at 5'0 clock position. On digital rectal examination, external sphincter tone was normal, irregular thickening of mucosa was felt. Proctoscopy showed irregular thickening of the mucosa with a polypoid lesion at 5'0 clock position with everted margins and communication with fistulous tract. Rectum was very tender. Sigmoidoscopy was performed which showed opening of fistulous tract at 5'0 clock and about 1 cm nodular lesion, yellowish in colour at 5 cm in the rectum. Another 2cm fungating mass was found adjacent to it (Figure 1). A 1cm yellowish plaque lesion was also seen



Figure 1. Photograph showing a fungating mass in the rectum.



Figure 2. Photograph showing yellowish plaque like lesions just above the dentate line.

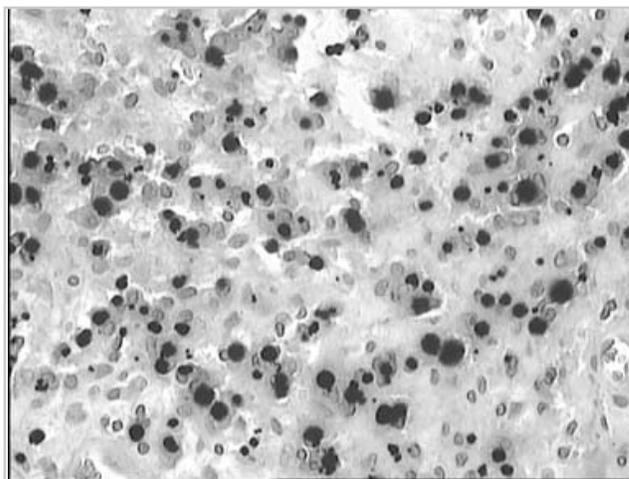


Figure 3. Photomicrograph showing intense black staining of the Michaelis-Gutmann bodies for calcium. ( Von Kossa stain. X 400)

2cm above the dentate line (Figure 2). Biopsy of these lesions was taken and sent for histopathology and culture. A clinical diagnosis of fistula with carcinoma rectum was made. CT scan of abdomen and pelvis showed enhancing soft tissue lesion in the anus invading anal canal and extending into fat planes. Biopsy examination showed malakoplakia with characteristic Michaelis-Gutmann bodies (Figure 3) and no evidence of malignancy. Culture of the tissue grew *E. Coli*. Immunosuppression therapy was curtailed and patient was started on ciprofloxacin 500mg OD for 6 months. The lesions regressed completely after six months of the above therapy and the patient became completely symptom free.

### Discussion

Malakoplakia is a rare, chronic granulomatous inflammatory disorder first described in 1902 by Michaelis and Gutmann,<sup>1</sup> and named by Von Hansemann in 1903<sup>2</sup> (from greek "malakos" meaning soft and "plakos" meaning

plaque). Lesions of malakoplakia are yellowish, soft plaques or nodules, usually elevated and sometimes with a central depression.<sup>2-4</sup> Diagnosis is usually made on histological examination of biopsy material. Diagnosis by fine needle aspiration has been reported.<sup>6</sup> Microscopically, malakoplakia is characterized by the presence of foamy histiocytes with distinctive basophilic inclusions, which are known as Michaelis-Gutmann bodies.<sup>5-7</sup> Malakoplakia most commonly involves the genitourinary tract, but may affect any organ/site of the body.<sup>3-9</sup> GIT malakoplakia is rare but presents in patients who are immunocompromised and have defects in macrophage function. Our patient, being a renal transplant recipient was on immunosuppression. Colorectal malakoplakia was first described by Turner and Lattes in 1965.<sup>5</sup> To date, forty one cases of gastrointestinal MK have been documented in English language literature. The most common sites of colonic involvement are the rectum, sigmoid and right colon in descending order of frequency.<sup>7</sup>

Lesions of malakoplakia are often misdiagnosed early in the course of disease as neoplastic lesions, as in our case and are correctly diagnosed only after biopsy. Cultures of the lesions can yield bacteria, most commonly *E. Coli* as seen in our case. Other enteric bacteria may be found in some cases.<sup>6,7</sup>

Pathogenesis of malakoplakia has been extensively investigated and it is believed to result from the inadequate killing of bacteria by macrophages or monocytes that exhibit defective phagolysosomal activity. Partially digested bacteria accumulate in monocytes or macrophages and lead to deposition of Ca and Iron on residual bacterial glycolipid. The presence of the resulting basophilic inclusion structure, the Michaelis Gutmann body is considered pathognomonic for malakoplakia.<sup>3-9</sup> Studies have suggested that a decreased intracellular cGMP level may interfere with adequate microtubular function and lysosomal activity, leading to incomplete elimination of bacteria from macrophages.<sup>7</sup> Malakoplakia can occur in all age groups and occurrence of the disease is associated more with the presence of an immunosup-

pressed state affecting macrophage function than with age.<sup>9</sup>

Lesions usually present as soft yellowish plaques or nodules, but can present as polypoid masses, abscess, a malignancy, a lymphoma or a benign skin tag. Lesions are often chronic.<sup>1-9</sup>

Risk factors for the development of malakoplakia include prolonged therapy with systemic corticosteroids, organ transplantation, diabetes mellitus, lymphoma and rheumatoid arthritis.<sup>5</sup>

Therapy of malakoplakia includes both medical and surgical aspects.<sup>10</sup> Medical treatment consists of antibiotics that concentrate in macrophages e.g. Quinolones, TMP-SMX. Bethanechol, choline agonist has been used in combination with antibiotics and surgery. It increases cGMP levels.<sup>10</sup> Our patient was given ciprofloxacin 500 mg OD for 6 months along with a reduction of immunosuppression. Lesions almost disappeared after 4 months of treatment.

## References

1. Michaelis L, Gutmann C. Ueber Einschlusse in Blasantumoren, *Z Clin Med* 1902; 47:208-15.
2. von Hansemann D. Uber malakoplakia der harnblase. *Virchows Arch A Pathol Anat Histopathol* 1903; 173:302-8.
3. McClure J. Malakoplakia of gastrointestinal tract. *Postgrad Med J* 1981; 57:95-103.
4. Sanusi ID, Tio FO. Gastrointestinal malakoplakia. Report of a case and a review of the literature. *Am J Gastroenterol* 1974;62:356-66.
5. Turner JY, Lattes R. Malakoplakia of colon and retroperitoneum. *Am J Clin Pathol* 1965; 44:20-31.
6. Cipolletta L, Bianco MA, Fumo F, Orabana P, Piccinino F. Malakoplakia of the colon. *Gastrointest Endosc* 1995; 41: 225-8.
7. De la Garza T, Nunenz-Rasilla V, Alegre-Palafox R, Albores-Saavedra J. Malakoplakia of the colon: report of case, with a review of eight previous case reports. *Dis Colon Rectum* 1973;16:216-23.
8. Gonzalez- Angulo A, Corral E, Garcia- Torres R, Quijano M. Malakoplakia of the colon. *Gastroenterology* 1965; 48:383-7.
9. Ranchod M, Kahn LB. Malakoplakia of the gastrointestinal tract. *Arch Pathol* 1972; 94:90-7.
10. van der Voort HJ, ten Velden JA, Wassenaar RP, Silberbusch J. Malakoplakia. Two case reports and a comparison of treatment modalities based on literature review. *Arch Intern Med* 1996;156: 577-83.