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Malakoplakia: Rare Disorder with No Significant Racial Prevalence

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Abstract

Introduction: Malakoplakia is a rare acquired granulomatous inflammatory condition. Microscopically it is characterized by sheets of histiocytes with abundant eosinophilic granular cytoplasm (known as von Hansemann cells) with intracytoplasmic rounded concentric basophilic inclusions (Michelis-Guttmann bodies). We reviewed all cases of malakoplakia diagnosed at our institution in the past 15 years, including clinical, macroscopic, and microscopic features as well as treatment and prognosis. **Methods:** Gross and microscopic features of cases diagnosed as malakoplakia from 2005 to 2019 at our institution were reviewed. Clinical history, including sites of involvement, clinical presentation, imaging, past medical history, treatment, follow-up and prognosis, as well as demographic characteristics were obtained. **Results:** Eighteen surgical pathology cases of malakoplakia were identified. Most cases occurred in female (14/18, 77.8%) and all in adults (median age 48.5 years; range 31 - 87). Half of the patients were Caucasian, 44% African American and 6% were reported as "other" race. One third (6/18) of patients presented with a mass-like lesion on imaging or endoscopic studies. As to site of lesion, 61.1% (11/18) involved GU tract, 27.8% (5/18) gastrointestinal (GI) tract, 5.6% (1/18) liver, and 5.6% (1/18) vagina. 38.9% of patients (7/18) had antibiotics treatment upon diagnosis. **Conclusion:** Malakoplakia is a rare disorder occurring in a wide age range, more commonly in females, with no significant racial prevalence. While the GU and GI tracts are the most common sites, other anatomic locations can be involved. Accurate diagnosis and appropriate treatment are important to avoid recurrence.

Keywords

Malakoplakia, Von Hansemann Cells, Michelis-Guttmann Bodies

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1. Introduction

Malakoplakia, a term derived from the Greek adjectives "malakos" (soft) and "plaka" (plaque), is a rare acquired chronic granulomatous inflammatory condition. It was first described by Michaelis and Gutmann in 1902 as soft yellowish plaques found on mucosa of urinary bladder [1]. Microscopically it is characterized by sheets of histiocytes with abundant eosinophilic granular cytoplasm (known as von Hansemann cells) with scattered rounded concentric intracytoplasmic basophilic inclusions termed Michelis-Guttmann bodies [2]. Although the pathogenesis of malakoplakia is poorly understood, it is thought to involve gram negative bacteria and result from the insufficient killing of bacteria by macrophages associated infection, immunosuppression, or autoimmune diseases [3]. Most cases of malakoplakia occur in the genitourinary (GU) tract [4]. But involvement of structures outside the GU tract has been reported with increasing frequency, including gastrointestinal (GI) tract, pancreas, testis, prostate, skin, breast, lungs, thyroid gland, oropharynx, retroperitoneum, lymph nodes skeleton and brain [5] [6]. In this study, we reviewed all cases of malakoplakia diagnosed at our institution in the past 15 years, including clinical, macroscopic, and microscopic features as well as treatment and prognosis.

2. Methods

The surgical pathology cases diagnosed at the University of Alabama at Birmingham (UAB), from 2005 to 2019 were retrieved, after approval by institutional review board. Gross and microscopic features of all cases diagnosed as malakoplakia were reviewed. Clinical history, including sites of involvement, clinical presentation, imaging, past medical history, treatment, follow-up and prognosis, as well as demographic characteristics (age, gender, and ethnicity) were obtained and analyzed.

3. Results

A total of eighteen surgical pathology cases with a diagnosis of malakoplakia from 2005 to 2019 were identified (**Table 1**, **Figure 1**).

Most cases occurred in female (14/18, 77.8%) and all in adults (median age 48.5 years; range 31 - 87 years). Half of the patients were Caucasian, 44.4% African American and 5.6% were reported as "other race" (**Table 2**, **Table 3**).

22.2% of the cases (4/18) presented in the setting of organ transplantations as 2 kidney transplants, 1 liver transplant and 1 heart transplant. And 22.2% (4/18) cases were with malignancy (2 colon cancers, 1 chronic lymphocytic leukemia, 1 B-cell lymphoma). Other clinical settings included systemic lupus erythematous with lupus nephritis (1 case, 5.6%), diabetes mellitus (2 cases, 11.1%), recurrent infection (4 cases, 22.2%); no significant medical history was noted for the remaining patients.

Two of the cases were recurred with history of malakoplakia.

Table 1. Cases of Malakoplakia diagnosed at UAB from 2015-2019.

Case#	Age/Sex/Race	Site of lesion	Clinical presentation	Past medical history (PMH)	Treatment/Outcome
1	49/M/C	Abdomen	Intraabdominal abscess	Cirrhosis, Liver transplant	Antibiotics/resolved
2	53/M/C	Right colon	Cecum mass	Colon cancer	Supportive/resolved
3	49/M/C	Colon	Sigmoid colon mass	Sigmoid colon cancer	Supportive/ not documented
4	60/F/AA	Kidney, allograft	Urine tract infection	Kidney transplant	Antibiotics/resolved
5	59/F/AA	Left lateral bladder	Hematuria	Diabetes Mellitus	Supportive/resolved
6	44/F/AA	Unspecified GI biopsy	Rectal bleeding	SLE, lupus nephritis, kidney transplant	Antibiotics/resolved
7	44/F/C	Bladder	Follow up for malakoplakia bladder	kidney donor candidate, no PMH	Plaque resected/ resolved
8	44/F/C	Bladder	Hematuria, E Coli UTI	kidney donor candidate, no PMH	Plaque resected/ recurred
9	87/F/C	Bladder	Bladder "mass"	Recurrent UTI	Antibiotics/ not resolved
10	31/F/AA	Bladder	Gross hematuria	Recurrent pyelonephritis	Nephrostomy tube/ resolved
11	48/F/AA	Kidney	Recurrent UTI	Recurrent UTI	nephrostomy/resolved
12	48/F/AA	Bladder	Recurrent UTI	Recurrent UTI	Antibiotics/ not documented
13	45/F/AA	Left kidney	E Coli bacteria in urine	Heart transplant	Antibiotics/resolved
14	58/F/C	Vagina	Pelvic and perinephric mass	Pelvic mass	Antibiotics/resolved
15	38/F/AA	Kidney	Acute kidney failure	Renal dysfunction	Supportive/resolved
16	57/F/AA	Rectum	Screening colonoscopy	Diabetes Mellitus	Supportive/ not documented
17	73/M/"Other"	Rectum	5-year follow up for polyps	CLL	Supportive/ not documented
18	44/F/C	Unspecified GU biopsy	Hematuria	No PMH	Supportive/ not documented

M: Male, F: Female, C: Caucasian, AA: African American, SLE: Systemic Lupus Erythematosus, CLL: Chronic lymphocytic lymphoma, UTI: Urinary tract infection.

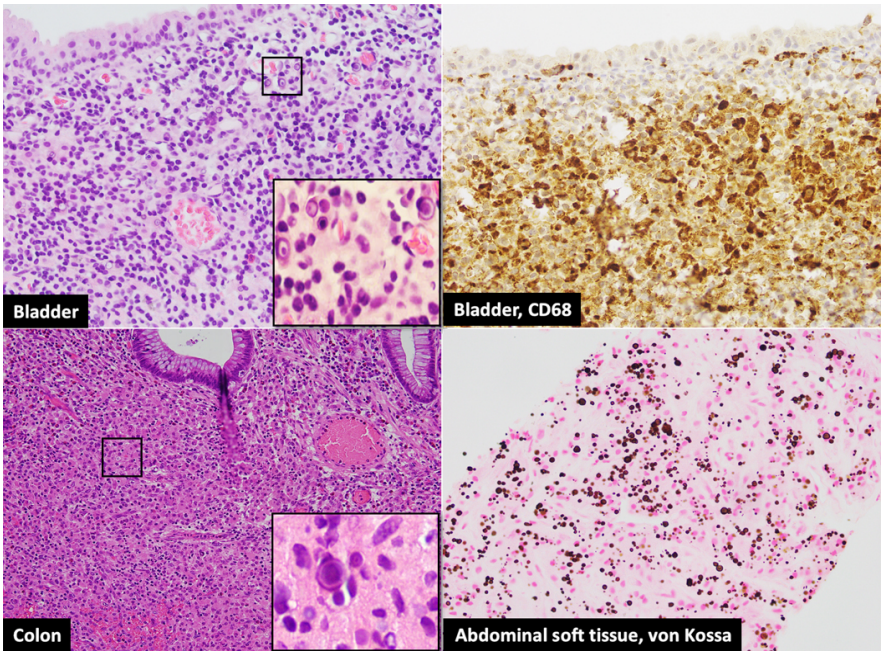

Figure 1. Upper: Malakoplakia in a bladder biopsy (400×). (Inlet: Von Hansemann cells with Michaelis-Gutmann bodies; B: CD68 stain highlighting macrophages). Lower left: Malakoplakia in a colectomy specimen of a patient with colonic cancer (200×). (inlet: Michaelis-Gutmann body). Lower right: von Kossa stain highlighting Michaelis-Gutmann bodies in abdominal soft tissue (400×).

Table 2. Sex distribution in patients with malakoplakia.

Sex	No. of cases	%
Male	4	77.8
Female	14	22.2

Table 3. Race distribution in patients with malakoplakia.

Race	No. of cases	%
Caucasian	9	50
American African	8	44.4
Other	1	5.6

Four patients had positive results of microbiological cultures (Escherichia coli) of either blood or urine.

One third (6/18) of patients presented with a mass-like lesion on imaging or endoscopic studies.

As to the sites of the lesions, 61.1% (11/18) involved GU tract (6 in bladder, 4 in kidney, 1 in unspecified GU tract), 27.8% (5/18) gastrointestinal (GI) tract (2 in colon, 2 in rectum, 1 in unspecified GI biopsy), 5.6% (1/18) in abdominal soft tissue, and 5.6% (1/18) in vagina (Table 4).

Table 4. Sites of involvement.

Site of involvement	No. of cases	%
Bladder	6	33.3
Kidney	4	22.2
Unspecified GU tract	1	5.6
GU total	11	61.1
Colon	2	11.1
Rectum	2	11.1
Unspecified GI biopsy	1	5.6
GI total	5	27.8
Abdominal soft tissue	1	5.6
Vagina	1	5.6

44.4% of patients (8/18) had antibiotics treatment upon diagnosis. Two cases (11.1%) underwent resection of the mass/plaque-like lesions, while the other cases were treated supportively including nephrostomy tube or stent placement, and fluid maintenance. In 11 patients (61.1%) with follow-up, malakoplakia resolved after treatment.

4. Discussion

4.1. Epidemiology

Since 1902 hundreds of cases of malakoplakia have been reported. Review of literature of prior reports showed it was most commonly observed in GU tract (approximately 75%) with a female to male ratio 4:1. The average age at presentation varies from 6 weeks to 90 years with average of 50 years [7]. No racial predilection has been reported.

Our single-institution study on the surgical pathology cases from the past fifteen years showed malakoplakia occurred in a wide age range (31 to 87 years) with average of 48.5 years. We did not find a case in pediatric population at our institution, although prior studies reported bimodal age incidence with a small peak occurring in childhood and a main peak in the fifth decade of life [7].

Our study showed female predilection (female to male ratio 3.5:1) in malakoplakia. And no significant racial prevalence is identified (50% Caucasian vs. 44.4% African American).

4.2. Medical Histories

The pathogenesis of malakoplakia is unknown, but most theories include the role of microorganisms, an abnormal immune response and abnormal lysosome function with macrophages [8] [9] [10]. It often occurs in patients with some

immunosuppression status. In our study, half of patients are on immunosuppressant therapy (status post transplantation, chemotherapy for solid malignancy, leukemia or lymphoma, or for systemic lupus erythematous). One third of patient have history of diabetes mellitus or recurrent urinary tract infection. Only 22.2% had cultured-proved microorganism infection (Escherichia coli). The remaining cases had no significant medical history.

4.3. Clinical Presentation

No specific symptoms are associated with malakoplakia. One third of patients presented with a mass-like lesion via imaging or endoscopic studies. Malakoplakia in the other cases were found incidentally during the either the routine biopsy for candidate transplant donor, after transplantation, or resection of tumor.

4.4. Sites of Malakoplakia

Prior study reported malakoplakia was most commonly observed in GU tract (approximately 75%), and GI tract was the second most common site (10%), with rectum, sigmoid and descending colon to be the most common sites of involvement, and colorectal adenocarcinoma as the most common associated condition [5] [7] [8] [11] [12]. Our study also show GU tract is the most common site (61.1%); GI tract is the second most common site of involvement (27.8%) including 2 cases in colon, 2 cases in rectum, 1 case in unspecified GI biopsy, 40% of the GI cases with colonic adenocarcinoma, 40% with tubular adenoma. As reported previously, malakoplakia can occur at other anatomic locations. We had 1 case in the liver status post liver transplantation, and 1 case in vagina with history of large B cell lymphoma status post chemotherapy.

4.5. Treatment and Prognoses

Two of the cases in our study were recurred with history of malakoplakia.

Antibiotics, cholinergic agonist, and discontinue of immunosuppressive agents have been believed the main approaches in prior reports [7]. In our study, patients obtained antibiotics treatment (44.4%, including daptomycin, mikamycin, vancomycin, cephalexin, levofloxacin, azithromycin, ciprofloxacin), resection of the mass/plaque-like lesions (11.1%), while the other cases were treated supportively including nephrostomy tube or stent placement, and fluid maintenance. In 11 patients (61.1%) with follow-up, malakoplakia resolved after treatment.

5. Conclusion

Malakoplakia is a rare disorder occurring in a wide age range, more commonly in females, with no significant racial prevalence. Most patients with malakoplakia have some type of immunosuppression, although it can occur in patients with no significant medical histories. While the GU and GI tracts are the most common sites, other anatomic locations can be involved. Accurate diagnosis and

appropriate treatment of malakoplakia are important because it can recur, and the prognosis is good with appropriate treatment.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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