A Rare Etiology of Back Pain: A Case Report of Retroperitoneal Malakoplakia

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ABSTRACT

Retroperitoneal malacoplakia is an inflammatory granulomatous disease rarely reported in the literature. A 59-year-old woman with diabetes mellitus came into our clinic complaining of back pain that had lasted about one month. Computed tomography scan of the abdomen revealed a loculated low-attenuation septated mass located between the inferior liver and right kidney. Because she had a high fever, antibiotics were administered empirically. During perisurgical intervention, she was found to have one retroperitoneal mass. Pathologic findings of the tumor revealed malacoplakia with distinctive Michaelis-Gutmann bodies. At three years of follow-up, the patient was found to have had no recurrence of malacoplakia. Herein we review her and review of published studies on patients treated for malacoplakia.
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Key words：Malakoplakia, Back pain, Michaelis-Gutmann body

Introduction

Malacoplakia is a rare inflammatory granulomatous disease which occurs most frequently in the urinary tract.[1] To the best of our knowledge, there are only a very few documented cases of retroperitoneal malacoplakia in the literature. Malakoplakia is diagnosed based on its unique hitsopathological features only. Antibiotics,

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bethanechol, ascorbic acid and surgical resection all constitute treatment options for this disease entity.[2] Herein, we describe a case involving a 59-year-old woman with retroperitoneal malakoplakia treated with antibiotics and surgical intervention. We also reviewed relevant literature.

Case Report

A 59-year-old female patient presented to our clinic complaining of intermittent back pain for one month. Her past medical history included hypertension and newly diagnosed diabetes mellitus for which she was receiving antihypertensive agents and oral hypoglycemia agents. Her current complaint manifested as right upper-quadrant abdominal pain, intermittent fever (up to 38°C), anorexia and significant body-weight loss that had progressed for two weeks. Physical examination showed chronic ill-looking appearance, anicteric sclera, tenderness over the right upper quadrant of her abdomen, and a right flank pain.

Laboratory investigations revealed a hemoglobin concentration of 14.0 (normal 13.5-17.5) g/dL, leukocyte count of 1 0.3 (normal 4.3-10.8) x 109/L, and platelet count of 334 (normal 130-400) x 109/L. Lactate dehydrogenase was 571 (normal 105-333) IU/L. Other biochemistry tests including renal function and liver function were within normal limits. Blood culture disclosed E. coli, five days subsequent to culture. Abdominal ultrasonography revealed a moderately fatty liver and one lobulated heterogenous hypoechoic lesion located between the inferior liver border and the right kidney. Abdominal computed tomography revealed a loculated low-attenuation septated mass located between the inferior liver and the right kidney (Fig. 1), leading us to highly suspect retroperitoneal abscess. Initially, spiking fever persisted despite administration of ceftriaxone. Later a pigtail catheter was performed, and a pus-like substance was drained. During the course of treatment, however, the pigtail slipped out accidentally one day later. Surgery was arranged during her hospitalization. Perisurgically, we located one retroperitoneal, soft, yellowish and plaque-like tumor 8 x 8 x 4 cm^3 between the patient's liver, gallbladder, duodenum, colon and kidney. The tumor's level of adhesion over various adjacent organs was explored. Culture of the turbid discharge revealed E. coli, as was found in the blood culture. Pathologic features of the mass included a number of pinkish, granular and/or vacuolated benign histiocytes between plasma cells, lymphocytes, and a focal aggregation of neutrophils. The presence of distinctive Michaelis-Gutmann bodies (small dark round to targetoid calcospherites) was also observed (Fig. 2 and 3). These finding led to a diagnosis of malacoplakia. The patient's fever subsided the day following the surgical removal of the tumor. Ceftriaxone therapy was discontinued two days after the disappearance of fever, and no complications were noted post-surgery. She showed good clinical outcome and was discharged. Over three years of follow-up, no recurrence of malakoplakia has been observed.

Discussion

In this report, we present a rare case of malakoplakia, an unusual granulomatous disease. Most commonly

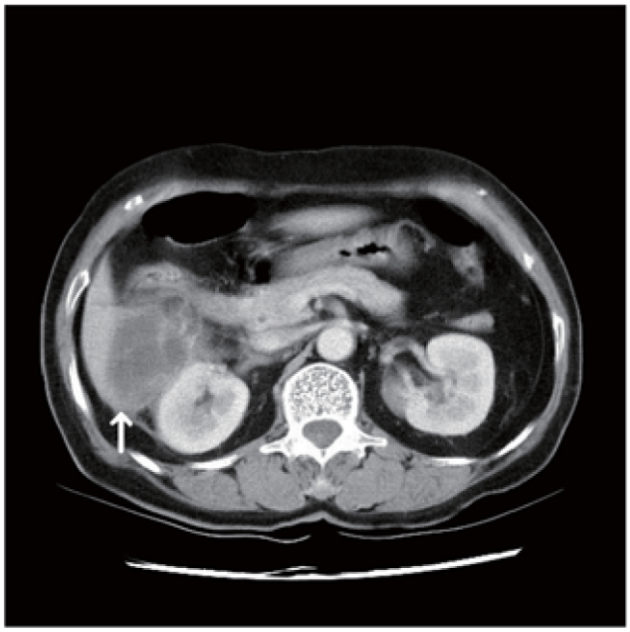

Figure 1. One septated, heterogeneous hypodense lesion may be seen to be located between the inferior segment of the patient's right liver and her right kidney (white arrow).

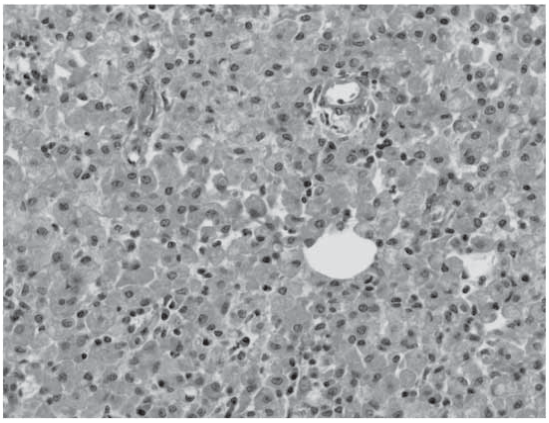

Figure 2. At low power, dense epitheliod histiocytes may be seen to infiltrate into the tissue of this retroperitoneal mass.

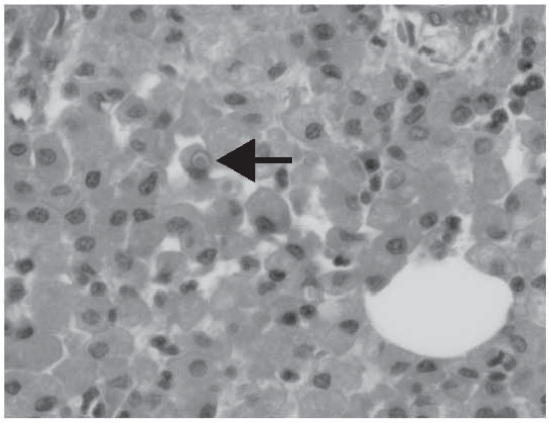

Figure 3. At high power, round calcospherite inclusions within the eosinophilic cytoplasm (Michaelis-Gutmann bodies) may be seen to be present (black arrow).

the urinary tract is involved (about 60% of all cases reported in the literature).[3] The bladder appears to be the most-commonly involved organ (70% of reported cases).[4] We found no clear etiology, but chronic systemic infection involving microorganisms combined with an immunocompromised response is widely accepted as an underlying mechanism for malakoplakia.[3] Malakoplakia is reported to be caused by one of several microorganisms, including Escherichia coli (up to 60% of all cases), Mycobacterium tuberculosis, Proteus var spp., Klebsiella pneumoniae, Staphylococcus aureus and Rhodococcus equi , such as is found in AIDS patients.[3,5,6] Malakoplakia is characterized by the presence of Michaelis-Gutmann bodies, which are intracellular or extracellular, round to oval bull's-eye or targetoid eosinophilic structures which consist primarily of calcium phosphate and an iron salt.[7] A diagnosis of malacoplakia can, therefore, be confirmed by a periodic acid Schiff's stain or by special stains for calcium and iron phosphate.[1,3,7]

We reviewed a total of 84 cases of malakoplakia, most of which well documented in the literature with most of the studies, including the relevant diagnostic methods undertaken, for the period 1995-2004. Malakoplakia commonly appears in the urinary tract system, though 40 of the cases we reviewed had malakoplakia in locations other than urinary tract system Table 1.[8] Of the 84 cases we reviewed, 28 cases were diagnosed by means of surgical resection; and 54 cases were diagnosed following biopsy histopathology. Two cases were diagnosed in AIDS patients following autopsy. Clearly, the majority of such patients were diagnosed by way of biopsy. The occurrence of a certain level of immunosuppression has been found in most cases with malakoplakia. Among the 84 patients we reviewed, 13 of patients had AIDS, seven had underlying malignancies and five had undergone organ transplantations. If immunosuppressive agents were being administered, the consumption of these agents should be stopped, if possible.[9] Other malakoplakia-attributable causes include diabetes, alcoholism and steroid use. We have found and reviewed 11 cases from the international literature, and found less than one such case per year reported worldwide.[10-20] Studies on the pathogenesis of malakoplakia have been conducted by Abdou and his colleagues, who found that malakoplakia resulted from the partially digested bacteria by macrophages or monocytes that display impaired phagolysosomal activity. The incomplete killing of microorganisms in monocytes or macrophages lead to a deposition of calcium and iron on glycolipid of residual bacteria. Thus, the appearance of basophilic inclusion structure, the Michaelis-Gutmann body, is characteristic of malakoplakia (Fig. 4).

Amongst the studies we reviewed, surgical resection of the lesion(s) and the administration of antibiotics appeared to have been the most-commonly used treatment regimen. Antibiotics that can penetrate the cell membrane and which are able to be taken up by macrophages are the drugs of choice for this disease.[9] Trimethoprim-sulfamthoxazole, fluoroquinolones and macrolide are agents that have been reported to be appropriate choices for the treatment of intracellular bacteria, one or more of which should be considered for the treatment of malakoplakia-afflicted patients (especially fluoroquinolones).[2,9,20] Additionally, short-term treatment with ceftriaxone followed by long-term treatment with sulfamethoxazole/trimethoprim has been reported

Table 1. Summary of published studies reporting a possible association with malakoplakia

Reported Cases of Malakoplakia in the Literature*					
No.	Author	Age	Sex	Location	Medical History
1	Lowitt et al	51 y/o	M	Perianal, inguinal, scrotum	Kidney Tx
2	Lowitt et al	67 y/o	M	Right temple	Kidney Tx
3	Moore et al	69 y/o	F	Right axilla	RA, breast cancer
4	Rao et al	40 y/o	F	Inguinal, broad ligament	N/A
5	Leclerc et al	64 y/o	M	Perianal	RA
6	Addison et al	35 y/o	M	Left eyelid	Kidney Tx
7	Almagro et al	64 y/o	M	Perianal	Lymphoma
8	Arul et al	75 y/o	F	Vulva	RA
9	Baez-Giangreco et al	50 y/o	F	Abdominal wound	N/A
10	Barnard et al	31 y/o	M	Right axilla	HIV
11	Biggar et al	32 y/o	M	Abdomen	Kidney Tx
12	Biggar et al	44 y/o	M	Perianal and left lung	Kidney Tx
13	Biggar et al	42 y/o	M	Right axilla	SLE
14	Bodokh et al	70 y/o	M	Buttock	Chronic hepatitis C
15	Carloz et al	75 y/o	M	Right hand and wrist	N/A
16	Chaudhry et al	41 y/o	M	Peritoneal	DM
17	Colby et al	74 y/o	M	Perianal	MPD
18	Davis et al	55 y/o	M	Gluteal cleft	HIV
19	Douglas-Jones et al	67 y/o	M	Left neck	N/A
20	Feldmann et al	81 y/o	F	Frontal mass	DM
21	Font et al	56 y/o	M	Internal canthus of eye	Sarcoidosis
22	Herrero et al	44 y/o	F	Buttock	Kidney Tx
23	Kumar et al	60 y/o	F	Nasolabial sulcus	N/A
24	Lou et al	2 mon	M	Colorectal and perianal	Immunodeficiency
25	Mehregan et al	68 y/o	M	Left inguinal region	N/A
26	Mehregan	66 y/o	M	Right axilla	RA, DM
27	Neiland et al	53 y/o	F	Perineum	Kidney Tx
28	Palazzo et al	42 y/o	M	Inguinal region	Lymphoma
29	Toubes-Klingler et al	41 y/o	M	Frontal scalp, right lung	HIV, Hepatitis B
30	Pang et al	64 y/o	F	Left neck mass	Thyroidectomy
31	Porrazzi et al	60 y/o	M	Gluteal fold	DM
32	Price et al	62 y/o	M	Chest	N/A
33	Reiner et al	65 y/o	M	Ureterocutaneous fistula	N/A
34	Remond et al	51 y/o	M	Perianal	Heart Tx
35	Sarkell et al	69 y/o	M	Left arm and flank	Escherichia coli sepsis
36	Scullin et al	55 y/o	F	Abdominal wall	N/A
37	Sencer et al	22 y/o	F	Arm	N/A
38	Sian et al	52 y/o	F	Inferior abdomen	Kidney Tx
39	Singh et al	30 y/o	M	Perianal	Dermatomyositis
40	Wittenberg et al	51 y/o	M	Left thigh	HIV, DM

Tx indicates transplantation; RA, rheumatoid arthritis; N/A, not available; HIV, human immunodeficiency virus; SLE, systemic lupus erythematosus; DM, diabetes mellitus; and MPD, myeloproliferative disorder.

*Adapted from [8] Kohl SK, Hans CP. Cutaneous malakoplakia. Arch Pathol Lab Med. 2008;132:113-7.

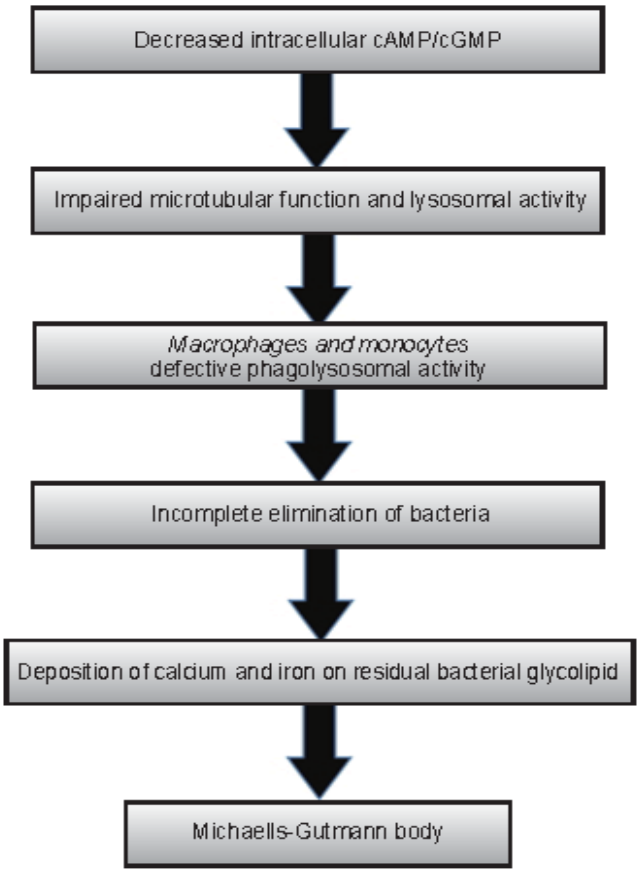

Figure 4. The pathogenesis of malakoplakia

to result in successful resolution.[21] The use of the cholinergic agonist bethanechol provides an advantage with regard to promoting phagocytic activity and has been considered by some investigators to be an appropriate supplemental medication to antibiotics.[22] In conclusion, the appropriate treatment goals for the malakoplakia-afflicted individual is early diagnosis, avoidance of complications, and timely intervention including antibiotic therapy and surgical intervention.

Conflicts of Interest Statement

The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in the manuscript.

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罕見背痛原因：後腹膜腔軟化症-個案報告

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摘　要

後腹膜腔的軟化斑是相關文獻報導中罕見的發炎性肉芽腫病變。一名59歲患有糖尿病的女性主述有長達一個月的背痛，腹部電腦斷層掃描顯示位於肝臟下方和右腎之間的位置有腫塊，因此使用經驗性抗生素來治療高燒狀況，同時以外科手術摘除後腹膜腔的腫塊。病理學呈現出具有軟化斑獨有特徵的Michaelis-Gutmann bodies，經過3年的追蹤治療，並沒有任何軟化斑復發的跡象。我們也回顧近年與軟化斑病人已發表有關研究文獻的綜述加以分析與比較。

關鍵詞：軟化斑、背痛、Michaelis-Gutmann小體

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