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## Case Report

## Malakoplakia of the liver in a patient with acquired immunodeficiency syndrome

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## ABSTRACT

Malakoplakia, a rare chronic granulomatous disease that often associates with immunosuppression or immunodeficiency, has been reported to affect many organs, mainly in the genitourinary tract. Herein, we report a case of malakoplakia of the liver in a 44-year-old man with acquired immunodeficiency syndrome who was poorly compliant with highly active antiretroviral therapy and subsequently died of multiple systemic infections. Malakoplakia of the liver was observed incidentally on autopsy.

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## 1. Introduction

Malakoplakia is a chronic granulomatous disease caused by an abnormal inflammatory response to infection that is usually described in immunosuppressed or immunodeficient patients. It is most commonly seen in the urinary bladder, although it has been reported in other locations, including the testis, prostate, epididymis, gastrointestinal tract, retroperitoneum, bone, adrenal glands, brain, and liver [1]. We present a case of liver malakoplakia discovered on autopsy in a patient with acquired immunodeficiency syndrome (AIDS).

## 2. Case report

A 44-year-old homosexual man was a sex worker at a local karaoke club and had unprotected sexual intercourse with more than three partners. He presented to Tzu Chi General Hospital in December 1999 with fever, dysuria, and facial rash. Human immunodeficiency virus status was confirmed by enzyme-linked immunosorbent assay and Western blot. From January 2000 to September 2002, he was poorly compliant with antiviral treatment and experienced general malaise, dizziness, anorexia, intermittent nausea, and diarrhea while his human immunodeficiency virus RNA titer was persistently high. He was subsequently

admitted in March and June 2004 for chronic watery diarrhea, and a stool acid-fast stain was positive for cryptosporidium. Syphilis was confirmed by serologic test for syphilis/rapid plasma reagin (+) and *Treponema pallidum* haemagglutination (+). Genital herpes and warts were noted. The patient was discharged after symptoms were controlled with antibiotics and antifungal prophylactics.

He again presented to the hospital in September 2007 because of a left gum mass. Biopsy showed T-cell lymphoma and he underwent palliative radiotherapy with 21.6 Gy. His condition soon deteriorated with poor appetite, nausea and vomiting, weight loss (4 kg in 4 months), painful perianal and penile ulcerations, generalized erythematous papules, and persistent watery nonbloody diarrhea. Because of the poor prognosis, he agreed to hospice care. The patient died in January 2008 and an autopsy was performed with prior consent of the patient and his family.

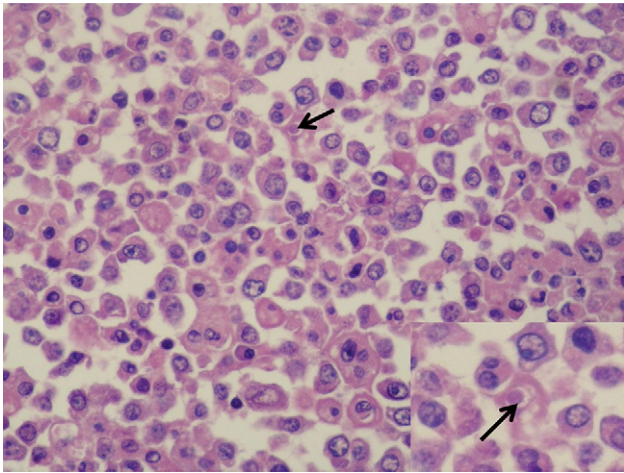
On autopsy, the liver weighted 1500 g and appeared smooth and firm. Diffuse fatty infiltration was noted. A 1 cm × 0.5 cm × 0.5 cm whitish firm nodule was noted in the liver parenchyma (Fig. 1). Microscopically, the main mass was well circumscribed and composed of lymphocytes, plasma cells, and large eosinophilic macrophages containing round, laminated Michaelis-Gutmann bodies (Fig. 2). A Gram stain showed gram-positive cocci in the histiocytes. Other systemic findings include myelofibrosis and hemophagocytosis in the bone marrow. The lungs showed bronchopneumonia involving all lobes, which was the cause of death. No definite malakoplakia or *Pneumocystis carinii* pneumonia was found. In addition, *Cryptosporidium* cysts were found in the small intestinal mucosa. No evidence of systemic infection was noted on autopsy.

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**Fig. 1.** Grossly, this is a well-defined, whitish, soft nodule measuring 1 cm × 0.5 cm × 0.5 cm.



**Fig. 2.** Histopathology shows infiltration with lymphoplasmic cells admixed with foamy histiocytes (×400) and characteristic Michaelis-Gutmann body formation (arrow, right lower panel) (hematoxylin and eosin stain).

### 3. Discussion

Two cases of liver malakoplakia have been reported previously. Robertson et al [1] reported one in a 54-year-old woman with systemic lupus erythematosus. After noting liver enlargement, a liver scan was performed and a liver abscess or tumor was impressed. However, a liver biopsy showed Michaelis-Gutmann bodies, which stained positive on a periodic acid-Schiff stain and Von Kossa stain for calcium. Steroids were discontinued and antibiotics were instituted. Unfortunately, the patient died of septic shock 18 months after diagnosis. Hartman et al [2] reported the

other in a 19-year-old Hispanic man with a history of suspected mucopolysaccharide disorder that resulted in a neurodegenerative condition. He presented with abdominal distention because of ascites. After excluding most causes, a needle core biopsy of the liver was performed revealing Michaelis-Gutmann bodies. They stained positive for periodic acid-Schiff, Von Kossa calcium, and colloidal iron stains, and were positive for CD68 on immunohistochemistry. After death, permission for autopsy was denied.

The pathogenesis of malakoplakia, although not yet fully elucidated, may be similar in all organs because of the similar morphologic features of the disease regardless of the site of involvement. Three possible mechanisms may be involved, specific microorganisms (*Rhodococcus equi* is common in patients with AIDS), an abnormal or altered immune response, and defective lysosomal function causing an abnormal macrophage response [3]. Michaelis-Gutmann bodies are formed when organic and inorganic materials (calcium and iron) are deposited on residual bacterial glycolipids, which phagocytes fail to digest [4].

Malakoplakia is frequent in immunocompromised conditions, including organ transplantation, malignancy, diabetes mellitus, AIDS, and malnutrition, and in patients receiving immunosuppressive drugs (chemotherapy and steroids) [3–5]. Our patient had been poorly compliant with antiretroviral therapy and developed multiple systemic infections and T-cell lymphoma during the terminal stage, suggesting poor immune status. We could not establish the etiology for the gram-positive cocci found in the malakoplakia as no blood culture or invasive therapeutic interventions were performed after he agreed to hospice and palliative care.

Reported clinical presentations of patients have been nonspecific, such as abdominal pain, diarrhea, and fever [1,2]. These symptoms were masked in our patient because of the severe immunodeficiency and other systemic symptoms. The diagnosis was based on sheets of granular, eosinophilic histiocytes without atypia, inflammatory cells, and the sine qua non of malakoplakia, Michaelis-Gutmann bodies [4].

To our knowledge, this is the first case of malakoplakia of the liver reported in an AIDS patient. Unlike patients in previous reports, this patient was immunodeficient. Symptoms related to his multiple infections may have masked those of malakoplakia in the liver. Because it was an incidental finding on autopsy, we could not estimate the duration of the disease or determine its etiology retrospectively.

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