Malakoplakia of the Spleen: A Case Report

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Abstract
Malakoplakia is an uncommon chronic inflammatory disorder, which is characterized by the presence of histiocytes containing concentric concretions known as Michaelis-Gutmann bodies in a background of mixed inflammation. The urinary tract is the most commonly involved site. However, malakoplakia can be found in a wide range of other organs throughout the body. Its occurrence has been attributed to a defect in the bactericidal capacity of phagocytic cells, and it is usually seen in patients with some degree of immunologic compromise. A case of malakoplakia of the spleen in a patient with Crohn’s disease is reported in this article.

Keywords
spleen, malakoplakia, Crohn’s disease

Malakoplakia is an uncommon chronic granulomatous inflammatory disorder, which was first described in 1902 by Michaelis and Gutman.1 The name (from the Greek malakos, soft, and plakos, plaque) is derived from its characteristic gross appearance of soft, yellow-brown plaques or nodules. It is characterized by the presence of histiocytes containing basophilic concentric concretions known as Michaelis-Gutmann bodies in a background of mixed inflammation. The urinary tract is the most commonly involved site, followed by the gastrointestinal tract and other sites such as prostate, central nervous system, lung, and soft tissue.2 To our knowledge, involvement of the spleen has not been previously reported in the English literature. In this article, we report a case of malakoplakia of the spleen in a patient with Crohn’s disease.

Case Report
A 42-year-old man with a history of Crohn’s disease, psoriasis, asthma, and gastroesophageal reflux disease underwent ileocolic resection for an ileal stricture. Three months after surgery, he was found to have a subcapsular splenic hematoma. He underwent ultrasound-guided drainage of the hematoma and embolization of the splenic artery. He did well and was discharged from hospital. There was no bacterial growth in the aspirate. He re-presented 2 months later with recurrent splenic hematoma and abscess. Culture of this fluid was positive for Escherichia coli. He underwent open splenectomy with drainage of the abscess and lysis of adhesions after unsuccessful antibiotic treatment.

Pathologic Findings
The specimen received for pathologic examination was a 10.0 × 6.0 × 4.0-cm spleen weighing 241.9 g with scant adhered fatty tissue containing areas of hemorrhage, fibrosis, fat necrosis, and exudate collection. Serial sectioning revealed prominent white pulp with focal fibrosis and hemorrhage.

Microscopic examination showed foci of acute and chronic inflammation with fibrosis consistent with organizing abscess. There was a focal area showing sheets of foamy macrophages intermingled with lymphocytes, neutrophils, and plasma cells. Scattered intracellular and extracellular targetoid basophilic concretions were present (Figure 1).

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A microscopic nodule with similar features was also noted in the parasplenic soft tissue, possibly representing a replaced lymph node. Periodic acid-Schiff (PAS) stain with diastase treatment highlighted the intracellular and extracellular inclusions, consistent with Michaelis-Gutmann bodies, and these also stained positive with von Kossa and Prussian blue special stains (Figure 2). Electron microscopy demonstrated a histiocyte containing a Michaelis-Gutmann body with a homogeneous peripheral area and a dense calcified central core (Figure 3).

**Discussion**

Malakoplakia is a rare chronic granulomatous inflammatory disorder. The diagnosis is confirmed by the presence of histiocytes containing Michaelis-Gutmann bodies. These can also be extracellular and are better visualized by positive staining with PAS after diastase treatment, von Kossa (for calcium), and Prussian blue (for iron) stains. By electron microscopy, they appear as dense concentric crystalline central cores surrounded by a homogenous zone. They have been shown to consist of 94.6% organic material, thought to be partially digested bacteria, and 5.4% inorganic material including calcium, phosphorus, and iron.

The exact etiology of malakoplakia is still unknown. In affected patients, its appearance seems to be related to an immunocompromised state, which can be induced, for example, by concomitant infections such as human immunodeficiency virus, tuberculosis, and neonatal herpes viral infection. Other associated conditions include alcohol abuse, malnutrition, organ transplant, drugs (such as steroids or cytotoxic chemotherapy), malignancy, and other chronic diseases (such as diabetes mellitus, autoimmune disease, and sarcoidosis). Central nervous system malakoplakia has also been described in areas of cerebral infarction.
There has been a substantial body of evidence implicating the role of an infectious agent in the pathogenesis of malakoplakia. It is hypothesized that there is a defect in the host intracellular bactericidal function, leading to an accumulation of incompetent macrophages along with intracellular and extracellular bacterial residues. The most commonly associated organisms are gram-negative bacilli including, most frequently, *E. coli*, *Proteus mirabilis*, and *Klebsiella pneumoniae*. Staphylococcal and streptococcal infections, as well as more fastidious organisms such as mycobacteria and fungi, have also been rarely reported. Interestingly, the most commonly associated organisms to be reported in AIDS are quite unique, namely *Rhodococcus equi* and *Shigella boydii*.

The genitourinary system is the most commonly involved site of malakoplakia, approaching up to 75% of cases. Middle-aged and elderly patients are most often affected, with an average age of more than 50 years and a 4:1 female predominance. More recently, there has been an increased frequency of reported involvement of other sites. The most common organ system to be affected outside the genitourinary system is the gastrointestinal tract, including colon, terminal ileum, stomach, and appendix. Here, it is frequently associated with colorectal carcinoma. There have also been rare reports of intestinal malakoplakia associated with inflammatory bowel disease. Other reported sites of involvement include gallbladder, pancreas, soft tissue of the neck, central nervous system, middle ear, tongue, tonsils, conjunctiva, thyroid gland, trachea, lung, lymph nodes, bone, skin, breast, adrenal gland, and peritoneum.

With its recognition as an infectious process, treatment with antibiotics has now become the cornerstone of therapy of malakoplakia. Given the underlying intracellular bactericidal defect, antibiotics that can penetrate the cell wall, such as fluoroquinolones, trimethoprim-sulfamethoxazole, and rifampicin, have been effective. The withdrawal of immunosuppressive drugs, such as steroids or azathioprine, is also essential and can lead to significant clinical improvement. However, in some cases, surgical treatment may still be necessary.

In summary, to our knowledge, this is the first reported case of malakoplakia in the spleen in humans. In this case, the patient had a history of Crohn’s disease and developed a refractory *Staphylococcus* species and *E. coli*-associated splenic abscess after ileocecal resection. The predominant pathologic change in this case was organizing abscess, and there was only a small focus of malakoplakia. As a rare entity, we present this case and also alert to its possible presentation as only a focal finding in a more extensive background of nonspecific inflammation.

References


