Int J Clin Exp Med 2017;10(8):12663-12666
www.ijcem.com /ISSN:1940-5901/IJCEM0057631

Case Report

Retroperitoneal malakoplakia mimicking sarcoma: a case report

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Received May 17, 2017; Accepted July 21, 2017; Epub August 15, 2017; Published August 30, 2017

Abstract: This report describes a rare case of retroperitoneal malakoplakia in a 70-year-old woman. The patient underwent open nephrectomy under suspicion of a huge retroperitoneal malignancy invading the right kidney. The mass was attached to the capsule of the lower pole of the kidney. Microscopically, the mass was composed of solid sheets of mononuclear cells and variable numbers of chronic inflammatory cells. Round basophilic structures known as Michaelis-Gutmann bodies were found within the mononuclear cells and extracellularly in the stroma. The mononuclear cells were immunohistochemically positive for CD68, indicating that these cells were histiocytes. Because the patient was preoperatively diagnosed with retroperitoneal malignancy, surgery was performed. Malakoplakia should be included in the differential diagnosis of retroperitoneal masses.

Keywords: Malakoplakia, retroperitoneum, malignancy

Introduction

Malakoplakia is a rare histiocytic disease that is often misdiagnosed as a malignancy. It can affect any organ system, including the genitourinary tract, gastrointestinal system, bones, lungs, lymph nodes, and skin, but the collecting system of the urinary tract is most frequently involved. In most patients, malakoplakia is associated with urinary tract infections, with Escherichia coli and Proteus mirabilis being the most commonly identified etiologic agents [1]. Malakoplakia is thought to be caused by defects in the phagocytic activity of macrophages. About 40% of patients have conditions associated with immunosuppression, including solid organ transplants, autoimmune diseases, chronic systemic diseases, malignancy, alcohol abuse, and diabetes mellitus [1, 2]. This report describes an unusual case of retroperitoneal malakoplakia extending to the kidney and mimicking retroperitoneal sarcoma.

Case report

A 70-year-old woman with a 1-month history of right flank pain was transferred to Chungbuk National University Hospital under the clinical suspicion of malignancy. Her past medical history was not significant. A physical examination revealed direct tenderness in the right flank area. She had mild leukocytosis (white blood cells, $13.2\times10^3/mm^3$) and anemia (hemoglobin, 11.4 g/dL; hematocrit, 34.3 vol%). The results of liver and kidney function tests and urinalysis were within normal limits. Urine cultures were negative for organisms. An enhanced abdominal bone computed tomography (CT) scan revealed a huge heterogeneously enhanced mass along the right iliopsoas muscle extending to the lower pole of the right kidney and the back muscles (**Figure 1**).

Because the patient was suspected of retroperitoneal sarcoma infiltrating the kidney, open nephrectomy with mass excision was performed. Gross examination showed a lobulated multinodular yellowish-tan mass (4.5×3 cm) in the perinephric soft tissue and attached to the outside of the capsule of the lower pole of the right kidney (**Figure 2**). There was no evidence of direct extension of the mass to the kidney parenchyma. The cut surface of the mass was yellowish-white in color, friable, and soft. The kidney and ureter were unremarkable. Microscopically, the mass was composed of solid sheets of mononuclear cells with abundant eosinophilic granular cytoplasm and variable

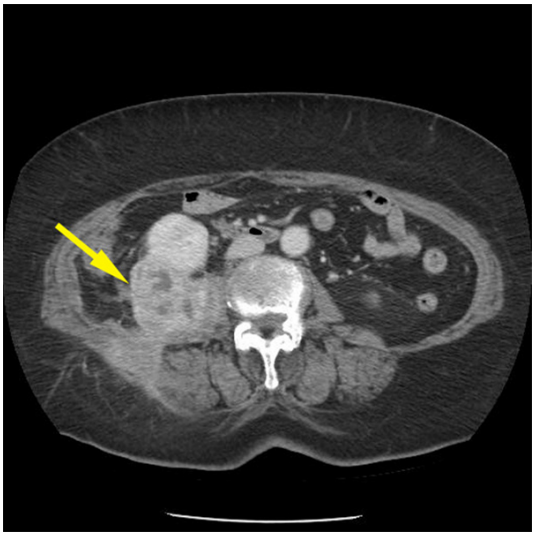

Figure 1. Contrast-enhanced computed tomography revealing a huge heterogenously enhancing lesion (arrow) along right iliopsoas muscle with extension to lower pole of right kidney and right back muscle.

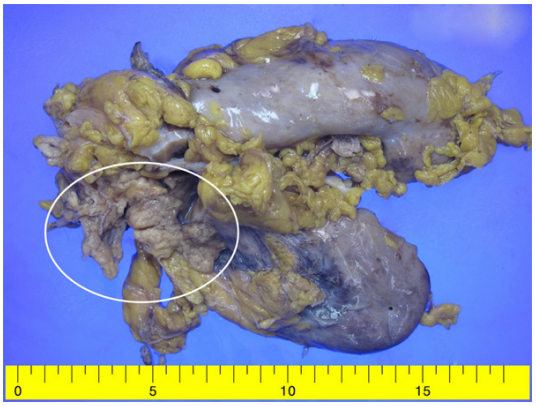

Figure 2. Photograph showing the gross appearance of the nephrectomy specimen. On evaluation, a lobulated multinodular yellowish-tan mass measuring 4.5×3 cm was identified in perinephric soft tissue attached to outside the capsule of lower pole of kidney.

numbers of chronic inflammatory cells (**Figure 3A**). Distinctive round basophilic structures with surrounding clear halos, known as Michaelis-Gutmann (M-G) bodies, were found within the mononuclear cells and extracellularly in the stroma. Some of these M-G bodies were laminated, some appeared homogeneous, and some had a dense central core with a targetoid appearance (**Figure 3B**). The mononuclear cells were confined to outside the perinephric fat and did not infiltrate the kidney. The histiocytes were positive for CD68 (**Figure 4A**), negative for cytokeratin, and positive for Periodic acid-Schiff (PAS) stained intracytoplasmic granules (**Figure 4B**).

After discharge from the hospital, the patient was treated with 500 mg/day levofloxacin alone. She remains alive and well 2 month after surgery.

Discussion

Malakoplakia is an unusual inflammatory disease, characterized by masses composed of histiocytes containing laminated intra- or extracytoplasmic inclusions, called M-G bodies. This condition, first described in 1902, presents as single or multiple tumor-like lesions and may occur in all organs. Malakoplakia is often difficult to diagnose, with the main differential diagnosis being malignancy [3].

Malakoplakia most frequently affects the urinary tract, with more than 50% of lesions occurring in the urinary bladder [4]. Other reported locations include the retroperitoneum, gastrointestinal tract, central nervous system, female genital tract, lungs, adrenal glands, spleen, pancreas, mesenteric lymph nodes, thyroid, eyes, skin, and bones [3]. Malakoplakia of the genitourinary tract is more common in women, whereas extraurinary malakoplakia is more frequent in men [3]. Age at diagnosis ranges from 6 weeks to 85 years, with a peak incidence in the fifth decade of life [5]. High rates of malakoplakia have been observed in patients with chronic urinary tract infection, immunocompromised conditions, and systemic diseases.

Malakoplakia results from a defect in the phagocytic function of histiocytes in response to Gram-negative bacteria. Non-digested bacteria accumulate in the cytoplasm and become mineralized by the deposition of calcium and iron on bacterial glycolipids [5]. These basophilic laminated concretions may be visualized by microscopy as pathognomonic M-G bodies. Most patients have positive urine cultures, usually with Gram-negative bacteria.

The treatment of malakoplakia depends on the extent of the disease and the underlying medical condition of the patient. Patients with bilateral or multifocal disease are most often treated with antibiotics such as fluoroquinolones and trimethoprim-sulfamethoxazole. Moreover,

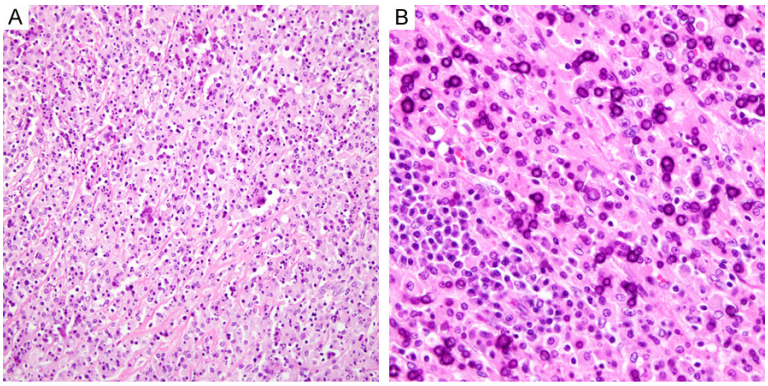

Figure 3. A: The mass was composed of mononuclear cells with abundant eosinophilic granular cytoplasm and various amounts of chronic inflammatory cells (magnification, ×200). B: The round basophilic structures with surrounding clear halos known as Michaelis-Gutmann bodies were identified (magnification, ×400).

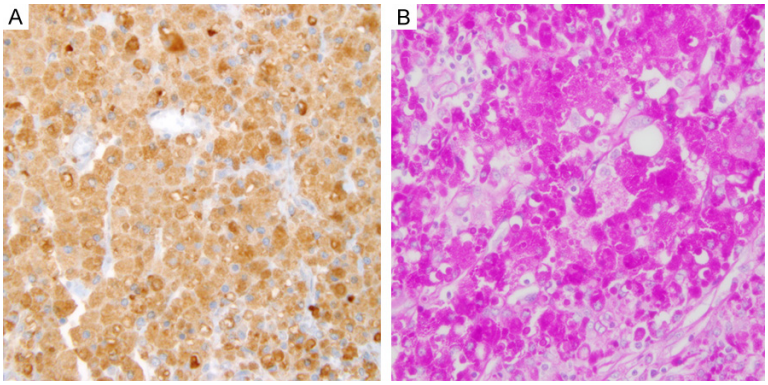

Figure 4. A: The immunostaining for CD68 was diffuse positive (magnification, ×400). B: Periodic acid-Schiff staining was positive for intracytoplasmic granules (magnification, ×400).

concomitant treatment with bethanechol chloride may correct the lysosomal defect. Surgical procedures are restricted to patients unresponsive to clinical therapy. Some patients respond to clinical and endoscopic treatment, whereas others may require aggressive surgical intervention [3]. The prognosis of patients with malakoplakia is usually good.

Diagnosis of malakoplakia is difficult, and it cannot be made exclusively in clinical or imaging ground. It is important to be aware that this entity may occur in abnormal locations and that it may mimic malignancy. Definitive diagnosis requires pathologic examination and is based on the pathognomonic findings described above. Preoperative biopsy may help in making clinical decisions and may reduce rates of unnecessary surgery.

At presentation, our patient had only a 1-month history of flank pain without any signs or symptoms of urinary tract infection. The results of urine culture were negative and there was no history of immunosuppression. The CT scan was not definitive but was indicative of malignancy or abscess formation.

Because retroperitoneal malakoplakia is rare, the patient was not preoperatively diagnosed with this condition. Based on a clinical suspicion of retroperitoneal sarcoma extending to the right kidney, open nephrectomy with mass excision was performed. The resected mass was attached to the capsule of the kidney without extension to the kidney parenchyma. Microscopically, the lesion was typical of malakoplakia. PAS staining was performed to better visualize the M-G bodies and immunohistochemical staining was performed to exclude other benign and malignant conditions.

Retroperitoneal malakoplakia was reported in a 20-year-old man who presented with a 3-month history of back pain, hematuria, lower urinary tract symptoms, and weight loss of 12 kg [3]. Imaging analysis showed a retroperitoneal mass infiltrating the urinary bladder. He was diagnosed with malakoplakia following a transurethral resection and was treated with fluoroquinolone, bethanechol, and vitamin C. A CT scan 12 months after treatment showed that the mass was reduced in size.

Malakoplakia is a chronic benign process that may present as a tumoral lesion of the retroperitoneum. Misinterpreting a large, rapidly growing malakoplakia as a tumor may lead to overtreatment, including unnecessary surgery. Our patient with retroperitoneal malakoplakia underwent an open nephrectomy based on the clinical suspicion of a sarcoma. Malakoplakia

should be included in the differential diagnosis of retroperitoneal masses.

Disclosure of conflict of interest

None.

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