Bladder Malakoplakia Simulating Neoplasm in a Young Girl: Report of a Case and Review of Literature

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ABSTRACT

Malakoplakia is a granulomatous disorder caused by infectious process. It was described by Von Hanseman in 1901 for the first time and then by Michaelis and Gutman in 1902. Although the most frequent site of involvement is genitourinary tract, various organs have been reported to be affected. The peak age incidence is about 50 years and it is rare in childhood. In this paper we report a case of bladder malakoplakia which to our knowledge is the youngest with isolated bladder malakoplakia that has been reported.

INTRODUCTION

Malakoplakia is a rare granulomatous disease of infectious etiology that most commonly is found in genitourinary tract. It is commonly observed in immunocompromised patients. Depending on the organ involvement, the patients may present in a myriad of ways and causing a huge diagnostic challenge. Malakoplakia is microscopically characterized by a collection of large mononuclear cells with abundant cytoplasm \(^1\). These cells are called Hanseman macrophages and are full of calcium and iron-laden lysosomal material that are known as Michaelis – Gutman bodies.

CASE REPORT

The patient is a 20 months girl that was brought for voiding dysfunction and repeated urinary tract infections and E. coli growth in urine culture. Urinalysis revealed pyuria. Blood profile, renal, and liver function tests were normal. Abdominal and pelvic ultrasound and C.T. Scan showed a mass in anterior part of the right lateral wall of the bladder, about 4 by 4 by 2.5 cm in size with suggestion for possibility of central necrosis (Figure 1). Cystoscopy showed a bladder mass, but biopsy was not informative and was suggestive for chronic cystitis. Exploration was done from a low-midline, retroperitoneal incision as the clinical diagnosis was a malignant neoplasm and the mass was resected with enough free margins (partial cystectomy).
Histopathology report (Figure 2) suggested malakoplakia. The patient received trimethoprim-sulfamethoxazole for 3 months after partial cystectomy and her 9 years post-operative period is uneventful.

**DISCUSSION**

Malakoplakia is usually seen in immunocompromized patients, but it can also be seen in immunocompetent individuals. Although the most common site of involvement is genitourinary tract, other common sites are gastrointestinal tract and retroperitoneum, but it can be seen everywhere in the body\(^2\). It is more common in males except for malakoplakia of genitourinary system that is more common in females. The peak age incidence is about 50 years and it is rare in childhood. The typical lesion of malakoplakia is grossly characterized by a soft yellow-brown mass or plaque with central ulceration and peripheral hyperemia. A patient with malakoplakia may present with a range of findings, but the standard criterion for diagnosis is pathologic evaluation. The pathologic findings are caused by defects in phagocytic degradative function of histiocytes in response to gram negative coliforms (E. coli or Proteus) that results in a chronic inflammatory process, followed by intracellular deposition of Calcium and Iron, a pattern that is known as Michaelis-Guttmann bodies. Large macrophages or von Hansemann cells with a variable inflammatory cells consisting mainly of lymphocytes, plasma cells and neutrophils are microscopic findings in malakoplakia. Although there is not a definite cause and effect relationship between coliforms and malakoplakia, many studies have shown an incidence of 89% to 93% coliform infections in patients with malakoplakia\(^3,4\).

Kajbafzadeh Am. and Baharnoori M. have reported a case of renal malakoplakia Simulating Neoplasm in a 10-year-old boy suffering from fever and headache for 20 days accompanied with poor condition and cachexia. An open biopsy of the mass was suggestive for malakoplakia and a trial treatment with bethanechol chloride, 12.5 mg three times daily, trimethoprim-sulfamethoxazole, one adult tablet per 12 hours, and ascorbic acid, 500 mg three times daily for 21 days managed the disease without surgical intervention\(^5\).
Amar Shah and Harish Chandran reported a case of malakoplakia presenting as multiple bladder polyps in an 11-year-old boy with no response to long-term antibiotic treatment, they performed surgical excision of the polyps and resolved his problem (6). Also they proposed surgical excision as an alternative form of management of this rare lesion. Surgical excision as an alternative treatment also has been suggested for very large lesions which complete eradication of them may be impossible by medical therapy alone (7).

Kuldeep Sh. And coworkers reported spontaneous perforation of the bladder in a 9-year-old female with coexistence of Xanthogranulomatous cystitis with malakoplakia (8). Raghavaiah NV. And coworkers have reported a case of nephrogenic adenoma of urinary bladder associated with malakoplakia in a 12 years female child, associated with recurrent Escherichia coli urinary tract infection, but to our knowledge our patient is the youngest with isolated bladder malakoplakia that has been reported(9)

REFERENCES

Figure 1: C.T.Scan of the pelvis
Figure 2: Histopathology of the bladder mass suggestive for Malakoplakia

Figure 2(a): Michaelis-gutmann body. Figure 2(b): Michaelis-gutmann body. Figure 2(c): Michaelis-gutmann body. Figure 2(d): Michaelis-gutmann body. Figure 2(e): Michaelis-gutmann body.