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CASE REPORT

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Malakoplakia of bladder in childhood

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Abstract Malakoplakia is a granulomatous inflammatory disease affecting the genitourinary tract. It is rare in childhood. We report a case of malakoplakia presenting as multiple bladder polyps in an 11-year-old boy. The child did not respond to long-term antibiotic treatment, and subsequent surgical excision of the polyps resolved his problems. We propose surgical excision as an alternative form of management of this rare lesion.

Keywords Malakoplakia · Bladder polyps · 11-year-old boy

The term malakoplakia is derived from the Greek words ''malakos,'' meaning soft, and ''plakos,'' meaning plaque. Michaelis and Gutmann first described this granulomatous inflammatory condition in 1902 [1]. Though commonly occurring in the genitourinary tract, malakoplakia can affect other organs of the body, including the skin. Our patient presented with frequent urinary tract infections with multiple bladder polyps and was subsequently diagnosed with malakoplakia.

Case report

An 11-year-old boy was referred with a history of recurrent urinary tract infections. He had a history of frequent urine infections, most of which grew *Escherichia coli*. Haematology and serum biochemistry were normal. An ultrasound scan showed a bladder polyp. The kidneys and ureters were normal. He had been operated on at the age of 5 years for repair of megalourethra and at the age of 7 years for a left pyeloplasty and ureteric reimplantation for left pelviureteric junction obstruction with lower ureteric stenosis.

Cystoscopy was carried out, which showed a 4-mm polyp arising from the posterior wall of the bladder and multiple yellowish nodules along the left lateral wall. The lesion was biopsied and showed changes of chronic inflammation. Two years later he again presented with urinary infections and terminal haematuria. Cystoscopy revealed two polyps, 0.4 cm and 1 cm in size, on the posterior bladder wall, which were biopsied (Figs. 1, 2). Histopathological examination showed mixed inflammatory cell infiltrate with histiocytes containing the characteristic Michaelis–Gutmann bodies of malakoplakia. The patient was started on long-term antibiotic prophylaxis. The following year he again had several urinary tract infections, which cultured *E. coli*. Follow-up cystoscopy showed multiple polypoid masses in the bladder. A 1-cm mass was found arising from the previous biopsy site on the posterior bladder wall, and three small 0.5-cm masses were found arising from the lateral bladder wall. The masses were excised cystoscopically. Histopathological examination confirmed malakoplakia. There was no evidence of malignancy. The child is presently well and on antibiotic prophylaxis. A follow-up cystoscopy is planned.

Discussion

Malakoplakia commonly affects the genitourinary tract, followed by the gastrointestinal tract and the retroperitoneum in decreasing order of frequency. It has also been reported in other sites, including the conjunctiva, tonsils, adrenal glands, spleen, pancreas, brain, lung, and skin [2]. Urinary tract malakoplakia is more common in females, whereas the extraurinary tract lesions are more frequently seen in males. The peak incidence is at 50 years, and children are rarely affected. The youngest reported case was in a 6-week-old child with malakoplakia of the adrenal and colon [3].

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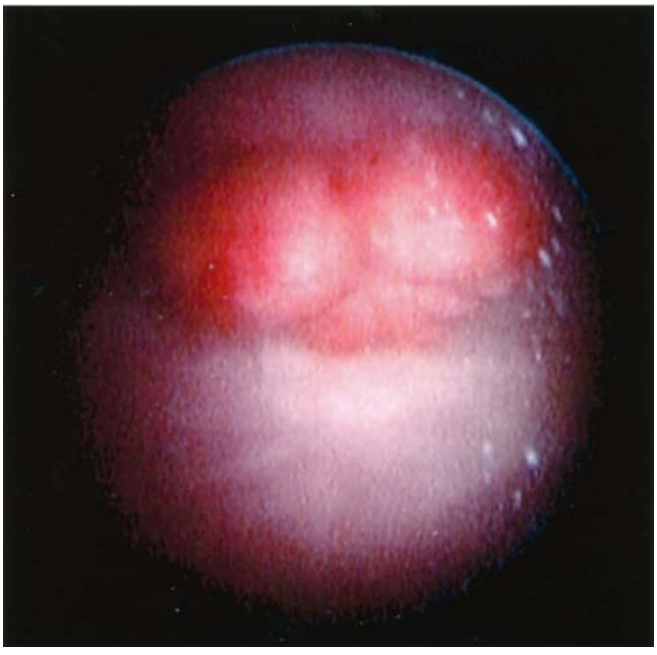

Fig. 1 Cystoscopic view of the bladder showing multiple bladder polyps

The lesions of malakoplakia characteristically appear as soft yellow-brown plaque with central ulceration and peripheral hyperemia. Malakoplakia is believed to result from inadequate killing of bacteria by macrophages or monocytes that exhibit defective phagolysosomal activity. The partially digested bacteria accumulate in the monocytes or macrophages and lead to deposition of calcium and iron on residual bacterial glycolipid. The presence of a resulting basophilic inclusion structure, the Michaelis–Gutmann body, is considered pathognomonic for malakoplakia. Many possible causes of malakoplakia have been proposed, which include prolonged systemic corticosteroids, tuberculosis, sarcoidosis, fungal and viral infections, and neoplasia. Some studies suggest a low cGMP/cAMP ratio to be the cause of defective phagocytosis, resulting in incomplete bacterial digestion.

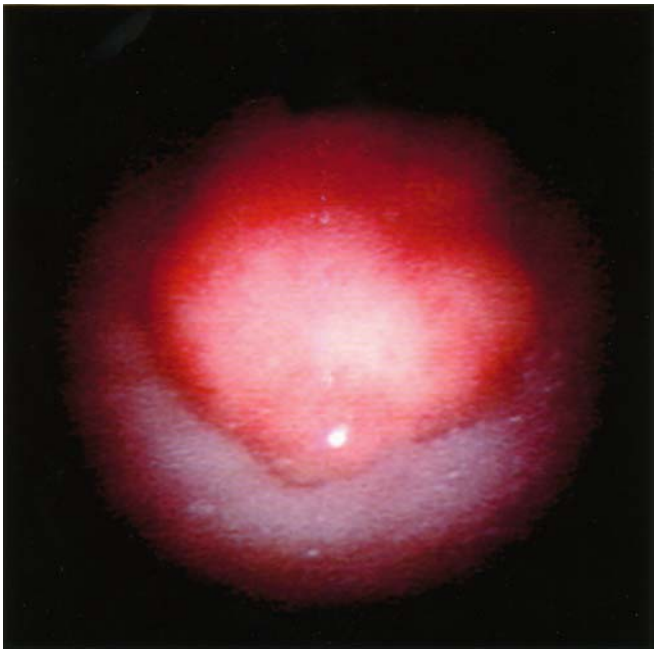

Fig. 2 Cystoscopic view of the bladder showing a bladder polyp

In the present case, the only striking feature was a long history of repeated urinary tract infections with *E. coli*. Different studies have reported an incidence of 89–93% coliform infections in patients with malakoplakia [4, 5]. Of these, 72% were *E. coli* infections. A cause–effect relationship between coliform organisms and malakoplakia has not yet been established, and the presence of an additional factor has been suggested. Lewin et al. [6] proposed the theory of unusual strains of *E. coli*, whereas Stanton and Maxted [4] suggested an immune deficiency syndrome or an autoimmune disease as an additional aetiology. McClure [7] found several diseases unrelated to the urinary tract in patients with malakoplakia. The presentation of malakoplakia as multiple bladder polyps is very uncommon.

The medical treatment of malakoplakia aims to improve the macrophage function by means of cholinergic agonists, e.g. bethanechol [8] and antimicrobials for bacterial elimination. The cholinergic agonists are believed to raise the intracellular cGMP concentrations [9], which stimulate synthesis of tumour necrosis factor [10]. Our attempt to treat the child with long-term antimicrobials was unsuccessful. Possible causes for this failure are poor penetration of the drug into the macrophages or its low activity in the phagolysosomes of the cells [11]. The child has been doing very well since his operation and continues on antibiotic prophylaxis.

Though long-term antibiotic treatment has been proposed, no major studies have been reported to show the cure of malakoplakia with antibiotics. We recommend surgical excision of the lesion as an additional tool to treat this rare lesion.

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