

CASE REPORT

Colonic malakoplakia in a child: report of a case and review of the literature

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ABSTRACT

Malakoplakia is an uncommon inflammatory condition usually affecting the genitourinary tract, which has been associated with infections, tumours and immunocompromised states. We report a case of malakoplakia in the colon of a 5-year-old girl with a history of bloody diarrhea. Clinically and macroscopically malakoplakia can simulate tumours or abscesses and can cause diagnostic difficulties.

Key words: Malakoplakia, Colon, child, infantile.

INTRODUCTION

Malakoplakia is a chronic inflammatory disorder. It was first described by Michaelis-Gutmann in 1902.¹ In 1903, it was named 'malakoplakia' by Von Hansemann.² The term malakoplakia derives from the Greek term 'malakos'(soft) and plakion (plaque). Malakoplakia has been reported in people with wide age spectrum (6 weeks to 88 years), but paediatric cases are sparse.³ It occurs most frequently in the genitourinary tract (about two third of the cases) but is also diagnosed in many other organs. It is claimed that the gastrointestinal tract (GI tract) being the second-most-common site.³ In the bowel, descending colon, sigmoid, and rectum are usually involved. Here we describe a case of malakoplakia of the colon in a 5-year-old Iranian girl.

CASE REPORT

A 5-year-old girl was admitted with a history of bloody diarrhea for about one month. Diarrhea was bloody and mucoid in appearance, and occurred 5-7times daily. On physical examination the child was pale and edematous, and her weight was 17.5 kg. No organomegaly was detected. Developmental state was normal.

Laboratory test results were haemoglobin=7 g/dl, erythrocyte sedimentation rate=80 mm/h, white blood cells= $17 \times 10^9/l$, plateletes= $870 \times 10^9/l$, C-reactive protein=3+and serum albumin=2.5 g/dl. Tuberculin skin test was non reactive. Stool examination revealed white blood cells =25-30and red blood cells=many, stool and urine cultures were negative. Abdominal sonography showed no abnormality. Upper endoscopy revealed reflux esophagitis. Colonoscopy showed multiple yellow plaque and masses from rectum to

cecum. There was no erythema, erosion or ulcer. Colonic biopsy was performed. Histologic examination showed colonic mucosa with infiltration of large macrophages in lamina propria with abundant pink cytoplasm, containing typical Michaelis-Guttman bodies and also infiltration of lymph mononuclear cells (Fig. 1,2).

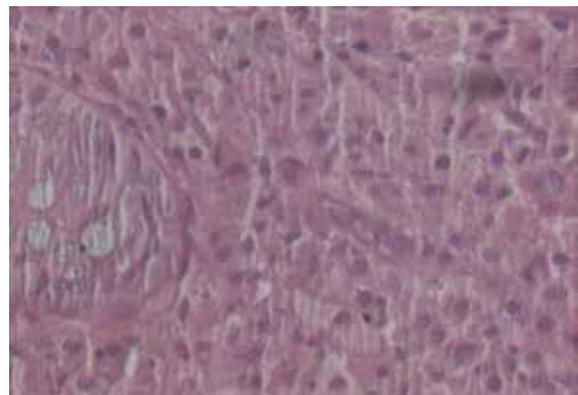


Figure 1, Section shows colonic mucosa with infiltration of large macrophages in lamina propria with abundant pink cytoplasm, containing typical Michaelis-Guttman bodies(H&E x40).

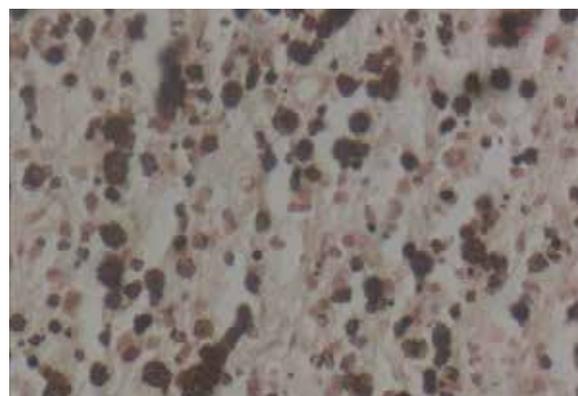


Figure 2, Michaelis-Guttman bodies stain for calcium(Von kossa staining.x40).

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The diagnosis was malacoplakia. Thereafter the patient evaluated for immunologic state. The immunologic tests showed normal level of immunoglobulins (IgA=1.4, IgG=18, IgM=1.2, IgE=9.7) but the percent of CD3⁺ and CD4⁺ lymphocytes were less than normal (27% and 15.7% respectively) (normal value:CD3+lymphocyte=55-82% and CD4+ lymphocyte=25-57%). The patient received intravenous alb-umin and packed cell. In addition we started methylprednisolon (15-20mg/kg/day for 3day), ciprofloxacin (250 mg twice a day for 2 weeks), cotrimoxazole (1tab twice a day) bethanechol (5mg twice a day) vit c (250 mg/day), ferrous sulfate (1 tab/day) and folic acid (1 tab/day). Diarrhea was stopped and anaemia improved. The next lab data (three weeks after treatment) were ESR=10, haemoglobin=11gr/dl & serum albumin=3.5gr/dl. Colonoscopy showed regression of the lesions. She is well in follow-up visits.

DISCUSSION

Malacoplakia is a rare chronic inflammatory disease with unique histologic features that involves different organs such as the urinary tract and gastrointestinal system.^{4, 5} In 1958, the first description of malakoplakia outside the urinary tract was reported.⁶ Now through searching the literature, there are accumulating reports of malakoplakia occurring in different organs other than the urinary tract. Among them the GI tract is the most common and it most commonly involve the colon and the rectum.⁷ It was in 1965 that colonic malakoplakia was first reported by Turner and Lattes.⁸ Malakoplakia is reported in the age range from 6 weeks to 88 years and there is a slight male predominance. Our patient was a 5-year-old girl. This case is interesting because of its rarity in children. Mouzan et al reported two brothers with confirmed colonic malakoplakia.⁹ Marino et al. explained 5 children with intestinal malakoplakia during a period of 10 years.¹⁰ Although the exact aetiology of malakoplakia is unclear, three possible mechanisms have been suggested. These mechanisms include microorganisms (especially *E.coli*), an altered immune response and an abnormal macrophage response because of defective lysosomal function.¹¹ As a result the moderately digested bacteria accumulate in macrophages and leads to a deposition of iron and calcium. Malakoplakia is commonly associated with immunocompromised status, systemic disorders, carcinoma and autoimmune disorders.¹ For example Sameer S Shaktawat described a case of malakoplakia of the appendix in association with inflammatory bowel disease.¹² F Karasavidou et al reported a 64-year-old man with colonic adenocarcinoma accompanied with colonic malakoplakia.¹³ Peter TW Kim et al. explained a case of colonic malakoplakia in a 58-year-old woman, with a history of liver transplantation.¹⁴ But there are examples of MLP in previously healthy subjects. Farahmand et al.¹⁵ described 3 children with intestinal malakoplakia but the immunity exploration of these cases were normal Malakoplakia of the colon can be

either segmental or diffuse. The gross appearance is usually a polypoid mass although a multinodular or stenotic presentation are not rare. In our case colonoscopy showed multiple yellow plaque and masses from rectum to cecum. Diagnosis is essentially by histological examination. All malakoplakias have the identical morphological features. It is important to be aware of the existence of this entity especially in unusual locations. Clinically the differential diagnosis of colonic malakoplakia includes Crohn enterocolitis, tuberculosis, and malignancy. Microscopically, colonic malakoplakia must be differentiated from Whipple disease, infectious or noninfectious granulomas such as tuberculosis, sarcoidosis, and finally histiocytic storage diseases. Also, pathologists should be alert to the possibility of more than one pathology being present in the specimen, because this may influence the therapeutic decision. A diagnosis of malakoplakia prompts precise work up for the possible wide aetiology (infections, immunosuppressed states, etc), which may remain unapparent. Our patient underwent careful investigation and we found that the percent of her CD3⁺ and CD4⁺ lymphocytes were less than normal (27% and 15.7% respectively). We suspect that the altered cellular immunity was the most reasonable trigger for the development of this disorder in our case. Antimicrobial agents are usually used for the treatment of malakoplakia. Quinolone antibiotics (e.g., ciprofloxacin) and sulfonamides (e.g., trimethoprim-sulfamethoxazole) are more popular. Bethanechol (to correct the lysosomal defect) and ascorbic acid have also been used in the treatment of patients with malakoplakia. Ascorbic acid has been used to improve the microtubular and lysosomal activity in monocytes. The presented patient received intravenous albumin, packed cell and methylprednisolon (15-20mg/kg/day for 3 day) ciprofloxacin (250 mg twice a day for 2 weeks), cotrimoxazole 1 tab twice a day, betanecohol 5mg twice a day vit c 250 mg/day, ferrous sulfate 1 tab/day and folic acid 1 tab/day. Now she is good.

CONCLUSION

Although malakoplakia usually affecting the genitourinary tract, examples of malakoplakia outside the urinary tract are reported. It is important to be aware of the existence of this entity especially in unusual locations. A diagnosis of malakoplakia prompts detailed investigations for the diverse etiology (infections, immunosuppressed states, etc), which may remain elusive.

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