Heliyon 9 (2023) e20780

Contents lists available at ScienceDirect

Heliyon

journal homepage: www.cell.com/heliyon

Rectal malakoplakia mimicking advanced rectal cancer: A case report

Xiangyu Liu [a], Chenming Yu [b,1], Zhuo Zhao [c], Yiping Zheng [d], Xin Chen [d], Dandan Zhou [d,*]

[a] Department of Radiology, The Fourth Affiliated Hospital, Zhejiang University School of Medicine, Yiwu, China
[b] Department of Interventional Radiology, Lishui District People's Hospital, Nanjing, China
[c] Department of Gastroenterology, The First Hospital of Jilin University, Changchun, China
[d] Department of Radiology, The First Hospital of Jilin University, Changchun, China

ARTICLE INFO

Keywords:
Malakoplakia
Michaelis-gutmann bodies
Rectum
FDG PET/CT
Case report

ABSTRACT

Background: Malakoplakia is a rare acquired chronic infectious granulomatous condition, that is characterized by the accumulation of large granular macrophages containing basophilic inclusion bodies in the cytoplasm termed Michaelis-Gutmann (MG) bodies. Malakoplakia most commonly involves the genitourinary system, and the second most commonly affected site is the gastrointestinal tract. Rectal malakoplakia is an unusual entity that is difficult to diagnose due to its diverse clinical manifestations and radiological findings that are similar to different diseases and advanced cancers.

Case description: A 61-year-old male patient presented with difficulty in urination and defecation that started 4 months prior, along with a weight loss of 10 kg. Abdominal computerized tomography (CT) scanning revealed diffuse lesions of the perirectal region with multiple lymphadenopathies and involvement of the bladder, prostate, bilateral seminal vesicles, and left ureter. 18F-FDG PET/CT MIP showed intense FDG uptake in the rectal region, and a diagnosis of an occupying lesion was proposed. Colonoscopy and histological examination of rectal lesion biopsies showed the characteristic features of malakoplakia.

Conclusion: Malakoplakia of the rectum with lymph node involvement and adjacent organ extension has been extensively misdiagnosed in clinical practice, and mimics malignancy radiologically. It is of great importance for radiologists to be aware of malakoplakia when making the differential diagnosis of benign and malignant mass lesions of the rectum, although the radiologic findings are nonspecific. Endoscopic evaluation and pathologic examination of a biopsy should be recommended to make the correct diagnosis, which may prevent unnecessary surgical resection.

1. Introduction

Malakoplakia (also acknowledged as von Hansemann's disease) is an extremely uncommon chronic infectious granulomatous lesion characterized by dense histiocytic infiltration. It was first reported by Michaelis and Gutmann in 1902, and one year later, the

* Corresponding author. Department of Radiology, The First Hospital of Jilin University, Changchun, China.
E-mail addresses: lxy592088@163.com (X. Liu), ycm199407@163.com (C. Yu), zhaozhuo831115@jlu.edu.cn (Z. Zhao), zhengyp22@mails.jlu.edu.cn (Y. Zheng), cxin21@mails.jlu.edu.cn (X. Chen), zhoudan0928@jlu.edu.cn (D. Zhou).
[1] Co-First author.

https://doi.org/10.1016/j.heliyon.2023.e20780
Received 13 April 2023; Received in revised form 4 October 2023; Accepted 6 October 2023
Available online 6 October 2023

term “malakoplakia” coined by Hansemann was derived from the Greek “malakos” (soft) “plakos” (plaque) [1]. Malakoplakia can affect all organs and tissues of the whole body, but most commonly occurs in the urogenital tract, with the bladder most frequently affected, followed by the prostate, kidney, ureter, testes, and epididymis. The gastrointestinal tract was involved second only to the urinary system, with the colon and rectum being the most common sites of involvement. Many other organs, including the skin, central nervous system, lung, bone, female genital tract and thyroid gland may also be rarely affected [1,2]. Malakoplakia in the urinary system is reported most commonly, and several studies have reported malakoplakia in the gastrointestinal tract. Yu-Chun Ma et al. (2022) described a case of colonic malakoplakia with the peritoneal cavity and retroperitoneum extension mimicking advanced colonic malignancy [3]. Scott Berger et al. (2022) reported a case of rectal malakoplakia in an immunocompromised woman with interstitial lung disease [4]. These studies indicate that malakoplakia more frequently develops in immunocompromised patients, with a higher preponderance in females. To our knowledge, Malakoplakia of the rectum is truly an unusual entity, with few cases reported in the literature. We report a case of rectal malakoplakia in a male patient whose tumour had spread into the retroperitoneal cavities, involving the bladder, prostate, bilateral seminal vesicles, left ureter, anterior abdominal wall and multiple lymph nodes, and the lesion presented as an advanced cancer radiologically [2,3].

2. Case description

A 61-year-old male patient had difficulty in urination and defecation 4 months previously, along with a weight loss of 10 kg from symptom onset and complained of progressive aggravation of symptoms with no fever, cough, expectoration, nausea or vomiting. The patient underwent cervical vertebra surgery and lower limb artery surgery more than 10 years prior. He denied having a history of viral hepatitis and HIV infectious diseases and a history of tuberculosis. According to the result of colonoscopy in a local hospital, the patient was considered to have rectal stenosis; intestinal tuberculosis could not be excluded; and abdominal CT examination demonstrated rectal occupation with lymph node and spermatic cord invasion.

After admission, routine blood tests showed decreases in red blood cells and haemoglobin (HGB at 9.3 g/dL), and inflammatory markers such as C-reactive protein (CRP) were mildly elevated to 2.47 mg/dL. Urine analysis showed white blood cell (WBC) and RBC counts of 526.80/μL and 80.20/μL, respectively. Serum CEA, CA19-9 and CA125 levels were within the normal ranges. Physical examination revealed no definite abnormal findings except for a palpable abdominal mass without tenderness.

A plain CT scan of the abdomen suggested diffuse lesions of the prostate, bilateral seminal vesicles, and perirectal areas, which were considered occupying lesions; there was uneven thickening of the bladder wall, thickening and dilatation of the left ureter, and moderate hydronephrosis in the left kidney (Fig. 1a and b). Enhanced CT of the colon revealed uneven enhancement of the lesions with multiple lymphadenopathies and invasion of adjacent organs, which were considered as lymphomas (Fig. 1c and d). 18F-FDG PET/CT MIP showed intense FDG uptake in the rectal region (Fig. 2). Fused PET/CT and axial, coronal, and sagittal CT images demonstrated multiple FDG-avid soft tissue density lesions involving the rectum, prostate, bilateral seminal vesicle glands and left ureter. Areas of FDG uptake in the anterior pelvic abdominal wall were observed (Fig. 3).

Colonoscopy revealed several flat polypoid lesions up to 5 cm in size from the descending colon to the rectum with a smooth surface, a 0.5*0.6 cm elevated mucosal lesion and a cupped mucosal lesion with an eroded surface in the rectum (Fig. 4a, b and 4c). Histological examination of rectal lesion biopsies indicated dense histiocyte infiltration with round laminated basophilic inclusion

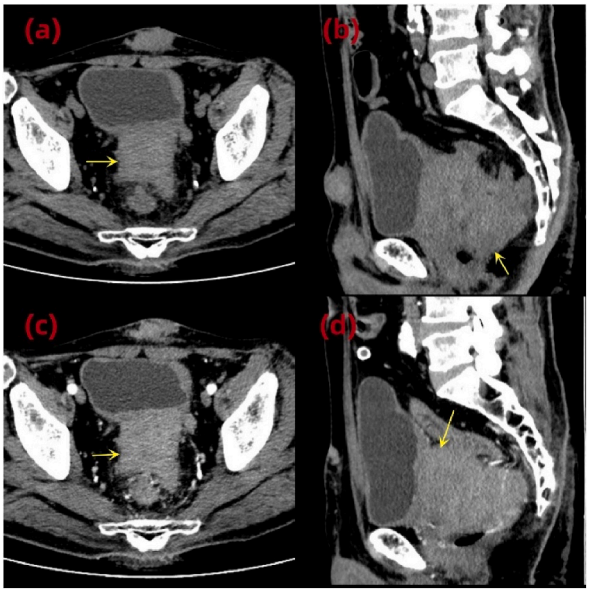

Fig. 1. Plain CT scan of the abdomen shows diffuse lesions of the perirectal, prostate, and bilateral seminal vesicles involving the bladder wall and the left ureter (a and b, arrow). Enhanced CT of the colon shows uneven contrast-enhancing lesions (c and d, arrow).

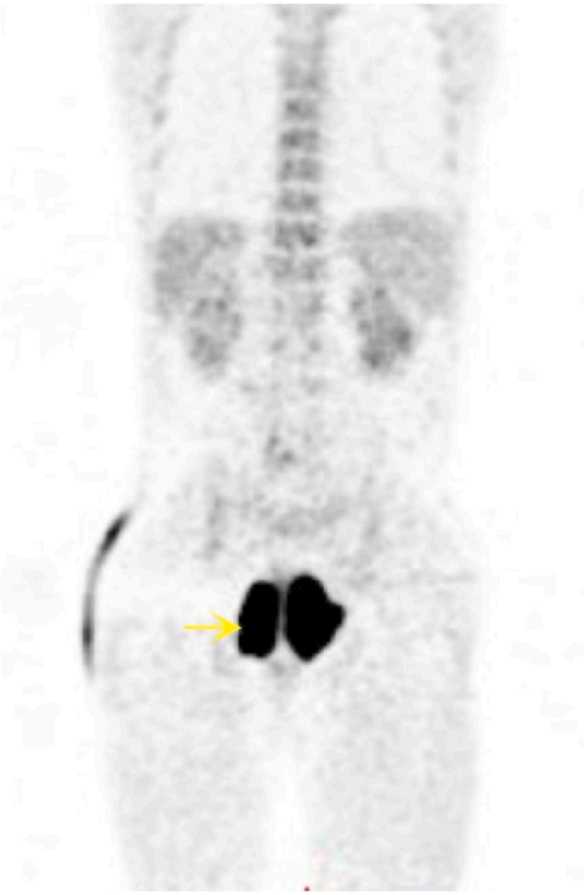

Fig. 2. PET/CT MIP shows areas of intense FDG uptake in the rectal region. (arrow).

bodies in the cytoplasm known as Michaelis-Gutmann bodies, which were pathognomonic features of malakoplakia, without evidence of malignancy (Fig. 5a). Immunohistochemistry showed the following findings:β-catenin (epithelial +), CD163 (+), SDHB (+), Ki-67 (2 %), and CD68 (+) (Fig. 5b). Special stains indicated PAS (+) (Fig. 5c). In addition, a CT-guided biopsy for the right inguinal lymph node was performed to eliminate the clinician's concern of the possibility of malignancy, and pathology showed dense infiltration of histiocytes. Taken together, a diagnosis of rectal malakoplakia with retroperitoneal involvement was confirmed.

3. Discussion

Malakoplakia is a rare acquired granulomatous condition, and is 4 times more prone to develop in females than in males. Its incidence peaks in middle-aged individuals, but the disease can also occasionally occur in children [1]. Malakoplakia is a rare disease that is difficult to diagnose due to its diverse clinical manifestations and radiological findings that are similar to different diseases and advanced cancers. In particular, literature regarding Malakoplakia of the rectum is limited. In this article, we report a case of rectal malakoplakia, including its clinical, radiological, and microscopic features.

The clinical presentation and signs of rectal malakoplakia are not specific, ranging from asymptomatic to diarrhoea, abdominal pain, abdominal discomfort, blood in the stool, constipation, anorexia, and intestinal obstruction [5,6]. The clinical symptoms of our patient were weight loss and dysuria, because the lesion involved the left ureter and caused obstruction.

The diagnosis of malakoplakia is mainly made by histologic examination [7]. Endoscopically, the lesions are focal or diffuse, and are varied in size and shape. The endoscopic findings revealed three typical patterns: thickened areas of mucosal erythema, multinodular or massive conditions that may resemble polyps and cancers, and large mass lesions [5,8,9]. Malakoplakia usually presents as soft yellow to brown mucosal plaques in the early stages or as grey to tan lesions with different shapes, surrounding congestion and central depression in the late stages [6].

Histologically, it is defined by the characteristic feature of accumulated histiocytes (termed von Hansemann cells or Hansemann histiocytes) with abundant 5–15 mm granular basophilic inclusions that are periodic acid-Schiff positive and diastase resistant, and Michaelis-Gutmann bodies [10,11]. MG bodies are the calcified detritus of incompletely digested bacteria mineralized by iron and calcium deposits within phagolysosomes and are thus positive for von Kossa calcium staining or iron staining [3]. It is interesting that Malakoplakia can occasionally be locally aggressive, as seen in the presented case, with an imitation of lymph node spread and

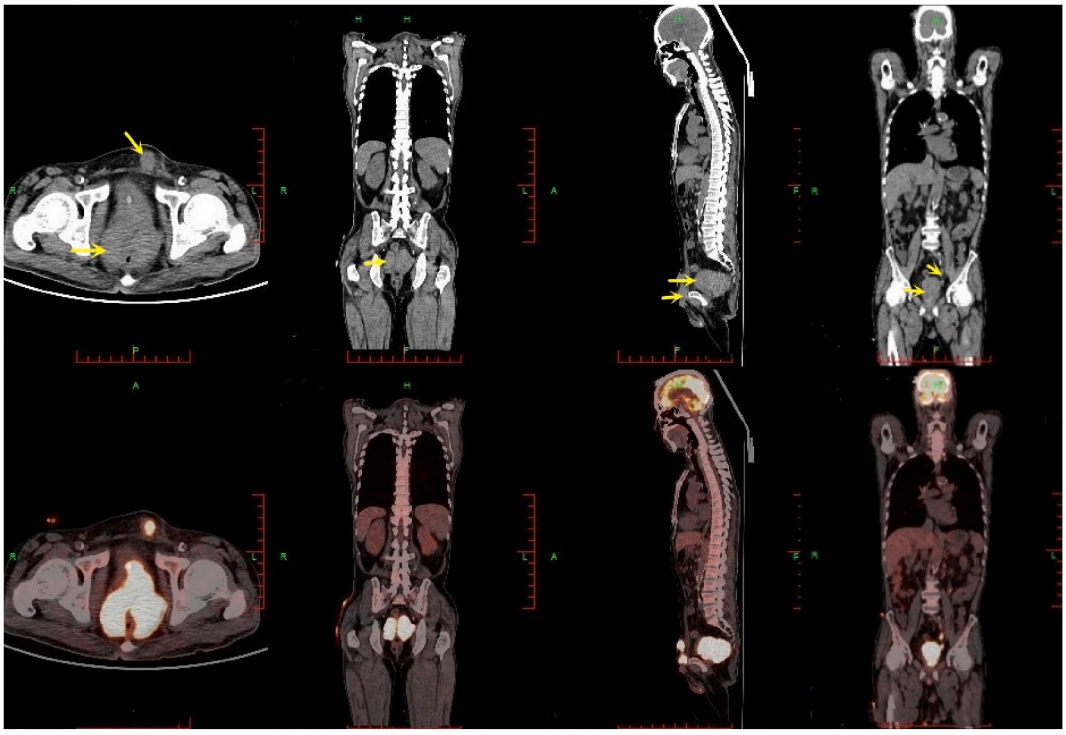

Fig. 3. Fused PET/CT and axial, coronal, and sagittal CT images: multiple FDG-avid soft tissue density lesions were seen in the rectum, prostate, bilateral seminal vesicle glands and left ureter. Areas of FDG uptake in the anterior pelvic abdominal wall were noted.

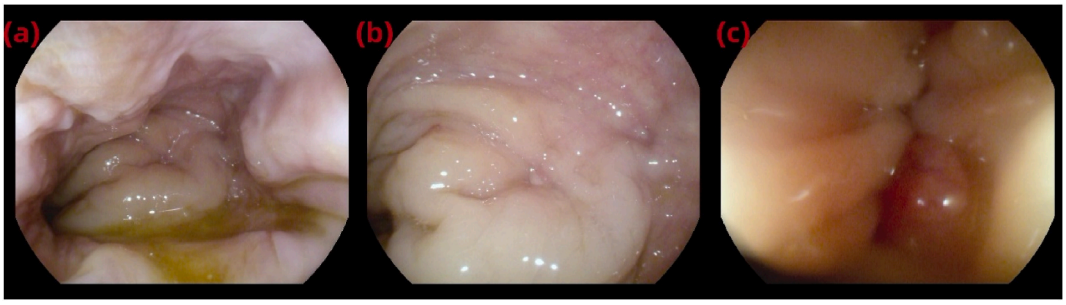

Fig. 4. Colonoscopy shows flat elevated mucosal lesions and polypoid mucosal lesions (a, b and c).

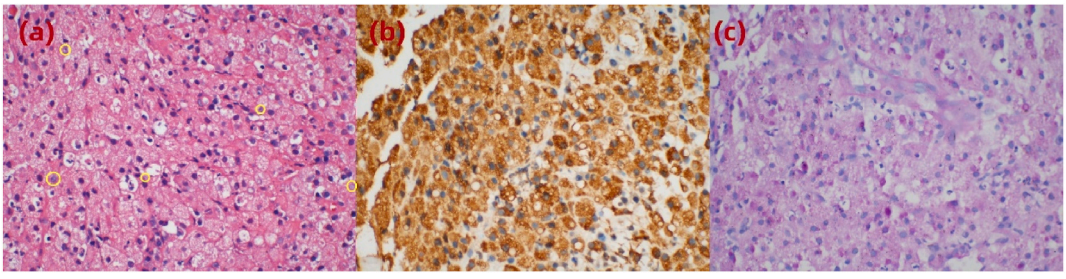

Fig. 5. Special histological stains show abundant macrophages (von Hansemann cells) with scattered cytoplasmic Michaelis-Gutmann bodies (circled) (a, H and E stain, x40). Immunohistochemistry showed $CD68^{+}$ (b, IHC, x40). Michaelis-Gutmann bodies and cytoplasm were purplish red (c, PAS, x40).

invasion of adjacent organs in another organ-confined tumour. The reasons may be that the accumulated von Hansemann cells infiltrate the gastrointestinal tract and extend into the retroperitoneum, which may imitate an advanced cancer clinically [12–14].

The aetiology and pathogenesis of the disorder is not fully understood, but three possible pathogenetic mechanisms have been proposed. First, microbial infection, such as infections with *Escherichia coli* (more than 2/3), *Mycobacterium tuberculosis*, Proteus, *Staphylococcus aureus*, etc. Second, it also can involve abnormal lysosomal phagocytic process of microorganisms by macrophages or monocytes. It is currently believed that low cGMP levels and reduced release of glucuronidase in monocytes or macrophages lead to lysosomal dysfunction, which makes microorganisms incompletely digested and cause the microorganisms to accumulate in lysosomes. Third, there is immune deficiency of the body. Malakoplakia mostly affects patients with an immunocompromised status, such as in patients with diabetes, cytotoxic chemotherapy, renal transplantation, tuberculosis, malignancy and AIDS [8,15].

Medical treatment of malakoplakia currently consists of appropriate antibiotic therapy and mitigation of immunosuppression. Antibiotics, such as quinolones or trimethoprim-sulfamethoxazole, obtain high concentrations in macrophages and are effective in killing bacteria [16]. As an alternative, cholinergic agonists such as bethanechol can increase the intracellular cGMP/cAMP ratio in macrophages, thus improving lysosomal bactericidal function [17]. Patients with malakoplakia usually have good results with pharmacologic treatment because it is a self-limiting and benign disease. Surgery may be required for therapy when pharmacologic treatment is ineffective [11].

4. Conclusion

In summary, our study presents a rare case of malakoplakia of the rectum with lymph node involvement and adjacent organ extension, which mimics malignancy radiologically and/or clinically. However, studies of lymph node involvement by malakoplakia have rarely been reported. This case highlights that it is of great importance for radiologists to be aware of malakoplakia when making the differential diagnosis of benign and malignant mass lesions of the rectum, although the radiologic findings are nonspecific. Endoscopic evaluation and pathologic examination of a biopsy should be recommended to make the correct diagnosis, which may prevent unnecessary surgical resection.

Data availability statement

No, the authors do not have permission to share data.

Statement

When the diagnosis of rectal malakoplakia was confirmed, we advised the patient to receive treatment, but he refused and requested to be discharged. As a result, some items in the CARE checklist were not applicable in my study.

CRediT authorship contribution statement

Xiangyu Liu: Writing – review & editing, Writing – original draft, Investigation, Data curation. **Chenming Yu:** Supervision. **Zhuo Zhao:** Methodology, Formal analysis, Data curation. **Yiping Zheng:** Investigation, Data curation. **Xin Chen:** Formal analysis, Data curation. **Dandan Zhou:** Writing – review & editing, Visualization, Validation, Supervision, Resources, Project administration, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

[1] Z. Wang, J. Ren, Clinical analysis of renal failure caused by malakoplakia: a case report and literature review, Front. Med. 9 (2022), 770731, https://doi.org/10.3389/fmed.2022.770731.
[2] S.S. Trepeta, S. Trikha, D.D. Alterman, CT of colonic malakoplakia in a patient with AIDS, AJR Am. J. Roentgenol. 171 (3) (1998) 637–638, https://doi.org/10.2214/ajr.171.3.9725288.
[3] Y.C. Ma, S.W. Wang, K.C. Chang, Colonic malakoplakia with retroperitoneal extension mimicking advanced colon cancer, J. Gastrointest. Surg. 26 (11) (2022) 2393–2395, https://doi.org/10.1007/s11605-022-05392-z.
[4] S. Berger, C. Marginean, M. Kaur, A case of rectal malakoplakia in an immunocompromised woman with chronic colonic bacterial infections, Clin. Gastroenterol. Hepatol. 20 (12) (2022) A27–A28, 10. 1016/j.cgh.2022.06.004.
[5] G.M. Yousef, B. Naghibi, M.M. Hamodat, Malakoplakia outside the urinary tract, Arch. Pathol. Lab Med. 131 (2) (2007) 297–300, https://doi.org/10.5858/2007-131-297-MOTUT.
[6] K.H. Hyun, H.D. Shin, D.H. Kim, Malakoplakia in a healthy young female patient, Korean J. Intern. Med. (Engl. Ed.) 28 (4) (2013) 475–480, https://doi.org/10.3904/kjim.2013.28.4.475.
[7] L. Cipolletta, M.A. Bianco, F. Fumo, P. Orabona, F. Piccinino, Malacoplakia of the colon, Gastrointest. Endosc. 41 (3) (1995) 255–258, https://doi.org/10.1016/s0016-5107(95)70351-9.
[8] M.J. Stanton, W. Maxted, Malacoplakia: a study of the literature and current concepts of pathogenesis, diagnosis and treatment, J. Urol. 125 (2) (1981) 139–146, https://doi.org/10.1016/s0022-5347(17)54940-x.

[9] L.C. Gustavo, M.E. Robert, L.W. Lamps, S.P. Lagarde, D. Jain, Isolated gastric malakoplakia: a case report and review of the literature, Arch. Pathol. Lab Med. 128 (11) (2004) e153–e156, 10.5858/200 4-128-e153-IGMACR.
[10] J.W. Park, D.H. Baek, S.J. Lee, Multiple polypoid lesions in the sigmoid colon, Gastroenterology 158 (3) (2020) 482–484, https://doi.org/10.1053/j.gastro.2019.09.021.
[11] A. Mitchell, A. Dugas, Malakoplakia of the colon following renal transplantation in a 73 year old woman: report of a case presenting as intestinal perforation, Diagn, For. Pathol. 14 (1) (2019) 22, 10.1186/s 13000-019-0799-z.
[12] L. Hubbard, S. Iriana, A. Carey, R. Odrobina, M. Sossenheimer, D. Rogers, Radiographic and endoscopic features of pancreaticoduodenal malakoplakia, Pancreas 49 (3) (2020) 455–460, 10.1097/MP A.0000000000001497.
[13] Y. Zhang, K. Byrnes, D. Lam-Himlin, M. Pittman, M. Pezhouh, R.S. Gonzalez, et al., Gastrointestinal malakoplakia: clinicopathologic analysis of 26 cases, Am. J. Surg. Pathol. 44 (9) (2020) 1251–1258, https://doi.org/10.1097/pas.0000000000001491.
[14] M.J. Irarrázaval-Mainguyague, M. Cabreras, S. Oksenberg, M.A. Pulgar, F. Rojas, M. Álvarez, et al., Malakoplakia mimicking a locally advanced colorectal neoplasm, J. Surg. Case Rep. 2021 (6) (2021), https://doi.org/10.1093/jscr/rjab225 rjab225.
[15] K. Pillay, R. Chetty, Malakoplakia in association with colorectal carcinoma: a series of four cases, Pathology 34 (4) (2002) 332–335, https://doi.org/10.1080/0031302027601204 81.
[16] M. Lee, H.M. Ko, A. Rubino, H. Lee, R. Gill, S.M. Lagana, Malakoplakia of the gastrointestinal tract: clinicopathologic analysis of 23 cases, Diagn. Pathol. 15 (1) (2020) 97, https://doi.org/10.1186/s13000-020-01013-y.
[17] M. Webb, J.R. Pincott, W.C. Marshall, L. Spitz, B.A. Harvey, J.F. Soothill, Hypogammaglobulinaemia and malakoplakia: response to bethanechol, Eur. J. Pediatr. 145 (4) (1986) 297–302, 10.1007/bf0 0439404.