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CASE REPORT

MALACOPLAKIA OF THE CECUM: REPORT OF A CASE

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SUMMARY

A case of malacoplakia of the cecum was studied by light and electron microscopy. The findings were typical. The report stresses the rarity of this condition, and its pathogenesis is discussed.

RESUMO

Malacoplaquia do cego: relato de um caso

Um caso de malacoplaquia do cego foi estudado através de microscopia óptica e electrónica. Os achados foram típicos. O relato acentua a raridade desta condição, e o seu mecanismo patogenético é discutido.

INTRODUCTION

Gastrointestinal malacoplakia is an uncommon condition, there being 38 reported cases in the world literature.[1] The involvement of the cecum is extremely rare, there being only 6 documented cases.[1] Malacoplakia has often been misdiagnosed as a neoplastic lesion in biopsy specimens.[2, 3] It has been more frequently diagnosed as an incidental finding in autopsy. However it can be easily recognized due to its well-defined morphological characteristics. A better recognition of this entity can be reached only through continued study and reporting.

The aim of this paper is to report a case of cecal malacoplakia associated with colonic diverticulosis and paracoccidioidal (South American blastomycosis) osteomyelitis, and presenting as a tumoral mass in the right iliac fossa.

CASE REPORT

AM, white male, 56 years old, was admitted to the University Hospital of the Medical School of Ribeirão Preto on April 17, 1980 because of a paracoccidioidal (South American blastomycosis) pharyngeal abscess. At the age of 53 years he had been submitted to a cardiac revascularization surgery. Two and one years prior to admission he was submitted to surgical drainage and antibiotic therapy because of paracoccidioidal osteomyelitis of the right clavicle and the 6th left rib.

On admission the physical examination disclosed a palpable, movable and painful tumoral mass in the right iliac fossa. The barium enema of the colon revealed mucosal irregularities and wall stiffness in the cecum and diverticulosis of the colon. In the evolution a fracture of the 3rd cervical vertebra due to paracoccidioidomycosis occurred and arthrodesis of C_2-C_4 was performed on May 15, 1980. In the post-operative period the patient developed bilateral bronchopneumonia and acute renal failure, and died on July 13, 1980.

Autopsy disclosed multiple ulcerations of the cecal mucosa, a tumoral mass continuous with the posterior wall of the cecum and colonic diverticulosis. The mass, measuring 10 cm in its greatest dimension, was dark yellow, extended from the cecum to the right iliac fossa, and showed a necrotic center. The other autopsy findings included bilateral bronchopneumonia, enlarged abdominal lymph nodes, paracoccidioidal osteomyelitis of cervical vertebrae, three patent saphenous vein-coronary artery bypass grafts, multiple foci of myocardial fibrosis and calcified and ulcerated atheromas of thoracic and abdominal aorta. The light microscopic examination of the cecum showed a polymorph inflammatory process. The cecal wall was infiltrated with

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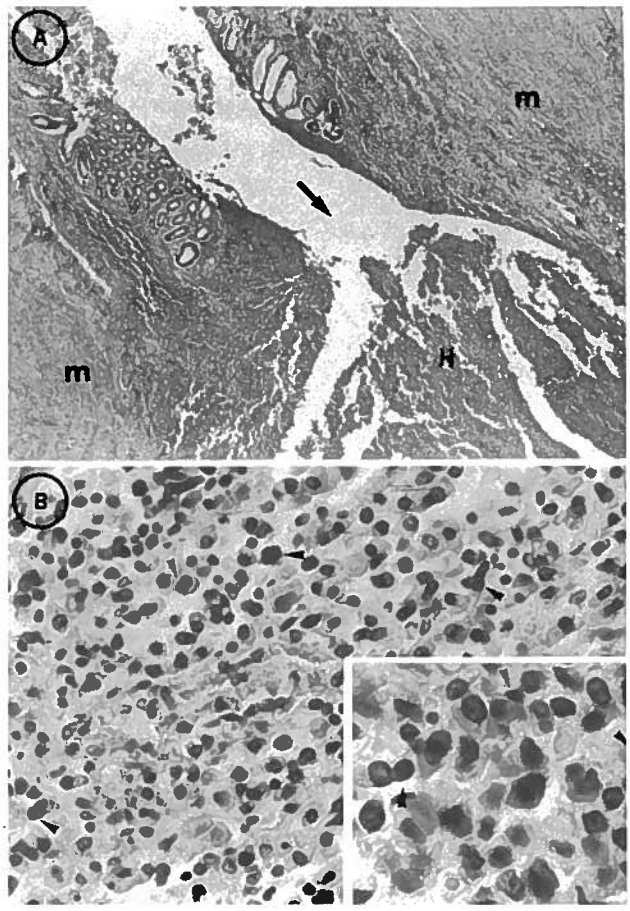

Figura 1: (A) Erosion of mucosa presumably at the mouth of a diverticulum (arrow) and large aggregate of histiocytes (H) spreading outside the cecal wall. m, muscularis externa (HE, x34). (B) Peripheral zone of the lesion consisting of an inflammatory exudate composed of large histiocytes and a few plasma cells and lymphocytes, Michaelis-Gutmann bodies in varying stages of development are seen within and among histiocytes (HE, x205; Inset, x520).

large masses of histiocytic cells with a small nucleus and abundant granular to vacuolated osmiophilic cytoplasm, polymorphonuclear leucocytes and a few plasma cells and lymphocytes spreading outside the cecal wall (Fig. 1 *a*, *b*). The overlying mucosa showed necrosis and ulcerations. The tumoral mass was predominantly composed of large histiocytes. These histiocytes, either in the cecal wall or in the retrocecal mass, showed the characteristic cytoplasmic calcified inclusions known as the Michaelis-Gutmann bodies (Fig. 1 *b*) allowing the diagnosis of malacoplakia.

Small fragments of the specimens of the malacoplakia, fixed in 10% formalin, were rinsed overnight in phosphate buffer, transferred to 2.5% glutaraldehyde solution in phosphate buffer for 4 hours, postfixed in 1% osmic acid for 2 hours, and embedded in araldite. Ultrathin sections were double stained with uranyl acetate and lead citrate and examined in a Zeiss EM 109 electron microscope. Many Michaelis-Gutmann bodies in different stages of development were seen in the ultrastructural examination: early forms, consisting of an aggregate of membranous lamellae, the typical Michaelis-Gutmann bodies, composed of alternating concentric zones of deposition of crystaline material and central calcification, and the transitional forms, made of concentric zones of crystaline material with a central core of membranous lamellae (Fig. 2).

DISCUSSION

Malacoplakia is a rare chronic inflammatory process characterized by proliferation of large histiocytes with a granular or vacuolated eosinophilic cytoplasm (Hansemann cells) containing rounded target-shaped calcified structures known as the Michaelis-Gutmann bodies. These bodies are also seen in the interstitial space. This condition is more frequently found in the urinary tract, where it was described for the first time.[3] However, it has been reported in other locations such as skin,[4] testis,[5] epididymis,[6] endometrium and broad ligament,[7] brain,[8] lung,[9] bone,[10] and gallbladder.[11]

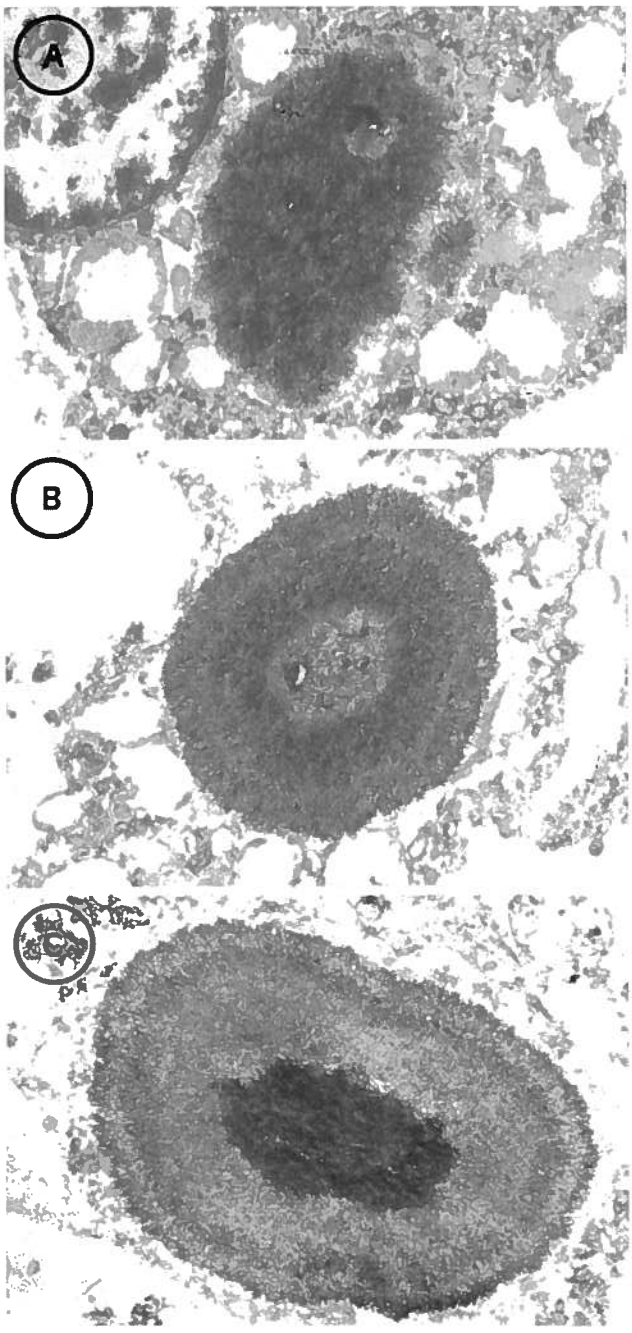

Figura 2: Electronmicrographs showing early (A), transitional (B), and typical (C) Michaelis-Gutmann bodies (x11 550).

Malacoplakia was first described by Michaelis and Gutmann[12] and the name was given by von Hansemann.[13] Michaelis and Gutmann suggested it to be a neoplastic process. The lesions often occur in patients with coexisting debilitating diseases such as tuberculosis, sarcoidosis, fungal and viruses infections and malignancies.[14] There is increasing evidence suggesting that they represent the result of an inherited or acquired abnormal macrophage response to bacterial infection.[15, 16] A frequent association with gram-negative bacteria, mainly *E. coli*, has been reported. The macrophage dysfunction is probably related with the phagolysosomal digestion of ingested material represented by microorganisms or inflammatory cells.

Since malacoplakia is a rare lesion, it has hardly been remembered in the differential diagnosis of tumoral processes. This occurred in the present case. The clinical finding of a palpable tumoral mass in the right iliac fossa associated with a chronic debilitating fungal disease (South American blastomycosis) and diverticulosis of the colon calls for the necessity of keeping in mind the possibility of less common pathologies in the differential diagnosis. Inflammation of a cecal diverticulum with subsequent pericolic spread would explain at least the location of the malacoplakia.

Three histologic forms have been described for malacoplakia, and they presumably represent transitional forms: early, granulomatous and fibrosing.[17] The classic lesion that permits the diagnosis is the granulomatous form. This probably explain, at least in part, the small number of reported cases and consequently why malacoplakia has been considered a rare condition.

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