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Pancreatic Malakoplakia in a Patient with Chronic Pancreatitis: A Case Report

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Case Report

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Abstract

Introduction: Malakoplakia is a granulomatous disease that most frequently involves the genitourinary system and is commonly associated with immunosuppressed patients. However, these lesions can affect other organs, including the pancreas. Its presentation can mimic other pancreatic pathologies, such as neoplasms or chronic pancreatitis, making its diagnosis challenging. Early and precise diagnosis of the disease may avoid unnecessary surgery and improve outcomes.

Patient presentation: A 39-year-old woman who presented with abdominal pain underwent a distal pancreatectomy and splenectomy, and the pathology revealed rare malakoplakia in the context of chronic pancreatitis. Notably, the patient had no immunosuppression or infection.

Conclusion: We report a rare case of pancreatic malakoplakia in a patient with no immunosuppression or infection to raise awareness of this condition and further reviewing findings from previously reported cases of pancreatic malakoplakia in the literature.

Introduction

Malakoplakia, characterized by defective phagolysosomal function in macrophages, is a granulomatous disease. The term “Malakoplakia,” first coined by von Hansemann in 1903, means “soft plaque”[1]. The pathognomonic histological feature of malakoplakia is the presence of Michaelis-Gutmann bodies, which were first identified and described by Michaelis and Gutmann in 1902[2].

Malakoplakia most commonly affects the urinary tract in immunocompromised individuals but has also been reported in those with a normal immune system, as well as in other organs, such as the gastrointestinal tract, lungs, and skin[3–8]. Pancreatic involvement is extremely rare, with only a few cases documented in the literature.

This case report describes a 39-year-old woman with a background of chronic pancreatitis and no immunosuppression who was diagnosed with pancreatic malakoplakia following distal pancreatectomy and splenectomy. The objective of this report is to raise awareness of this rare pathology and its diagnostic challenges, particularly in patients with underlying pancreatic conditions, and to review previously reported cases of pancreatic malakoplakia in English-language literatures.

Case Presentation

An afebrile 39-year-old woman with a history of alcoholic pancreatitis and gastroesophageal reflux disease (GERD) presented to our institute with diffuse, paroxysmal abdominal pain, which occurred 2–3 times per week and was exacerbated by eating. The patient had remained sober for 28 days since the onset of her symptoms. Physical examination revealed mild tenderness in the upper abdomen. Lactate: 3.6 mmol/L (0.5–1.6 mmol/L); WBC: 21.84 k/cumm (4.8–10.8 k/cumm); hemoglobin: 11.4 gm/dL (12.0–16.0 gm/dL); platelets: 55 k/cumm (140–440 k/cumm); Glucose: 206 mg/dL (70–100 mg/dL). There was no evidence of an underlying immunocompromised state. Blood culture and urinalysis were negative for bacteria or yeast. Contrast-enhanced CT of the abdomen revealed thick-walled fluid collections measuring 4.8 × 5.1 cm inferior to the body and tail of the pancreas, with an additional 1.2 cm pseudocyst superior to the pancreatic tail (Fig. 1). The patient underwent distal pancreatectomy and splenectomy due to concerns of severe chronic pancreatitis with possible necrosis. Intraoperative findings include extensive chronic pancreatitis with necrotic areas. There were no visible pancreatic masses or obvious malignancies.

On gross examination of the pancreatic specimen, the surface was fibrotic, with a soft, necrotic area abutting the resection margin measuring 2.0 × 1.3 × 1.0 cm. On light microscopy, sections from the pancreatic tail revealed extensive malakoplakia characterized by sheets of histiocytes containing basophilic Michaelis-Gutmann bodies (Figs. 3&4), with a background of chronic pancreatitis (Fig. 2).

At the 3-month follow-up, the patient reported significant improvement in her symptoms, with no further episodes of abdominal pain.

Discussion

Malakoplakia is characterized by defective phagolysosomal activity. The undigested bacterial fragments eventually become mineralized with iron and calcium phosphate, reflected by targetoid, PAS-D-positive Michaelis-Gutmann (MG) bodies. They represent calcified lysosomes containing bacterial remnants. Von Kossa’s staining is the most common method to highlight these intracytoplasmic inclusions[3]. In previously reported cases (Table 1), five out of eight showed bacterial infection, most commonly *Escherichia coli* infection [9–13], although *Klebsiella sp., Corynebacterium Equi, Streptococcus salivarius*, *and Enterococcus faecium* have also been reported. Two patients experienced significant reduction or resolution of pancreatic masses with conservative antibiotic treatment, underscoring the benign nature of malakoplakia[9, 12]. One patient had biopsy-confirmed malakoplakia and serology evidence of infection, emphasizing the importance of biopsy for management [12]. Despite its benign nature, five patients underwent surgery [14, 10, 15, 11, 13], with four involving pancreatectomy[14, 10, 11, 13]. Notably, one of the four patients with FNA-confirmed malakoplakia underwent Whipple’s procedure due to disease progression regardless of antibiotic treatment[11], suggesting the limits of conservative therapy. Although, malakoplakia is considered a benign process; the treatment should be customized according to specific clinical scenario. If a patient is stable, biopsy may be more beneficial as it might avoid invasive therapy overall.

Table 1
Summary of Pancreatic Malakoplakia in English-Language Literature

Malakoplakia in pancreas									
Author	Age	Gender	Imaging	Site	Malignancy	Immunodeficiency	Infection	Other features	Treat
Zuk,R.J., et al[14] 1990	45	F	CT: a probable pancreatic neoplasm	Head of pancreas [a]	Moderately well-differentiated adenocarcinoma	No	No	Metastasized to liver post Whipple's surgery	Whip proce
Sinclair-Smith, C., et al[17] 1975	6 weeks	M	None given	Adrenal and colon with spread into tail of pancreas [b]	No	Yes	No	Systemic involvement: Thymic and lymphoid hypoplasia	None
Colby, T.V. [10] 1978	49	M	None given	8-cm-diameter left retroperitoneal mass invading the tail of the pancreas [b]	No	suspicious	Yes, *Klebsiella* sp. In addition, *corynebacterium Equi.* in biopsies	Systemic involvement: Weight loss and anemia	lapar
Kulatunga, A., et al[15] 1987	75	M	None given	8 X 9 cm mass in caudal region of the pancreas [a]	No	No	No	Lesion identified at elective cholecystectomy	Chole and r resec panc reser
Guha, S., et al[11] 2015	59	M	Endoscopic ultrasound: 33 X 32 mm mass	Head of the pancreas [a]	No	No	Yes, *E. coli* in ascites	Systemic involvement: Abdominal pain, fatigue, and weight loss	Whip proce
Nuciforo, P.G., et al[13] 2003	74	M	CT and MRI: ovoidal mass measuring 4x 2 cm	Isthmus and the tail of the pancreas [a]	No	No	Yes, TEM showing bacterial-like structures	History of DM. Low grade fever and abdominal pain	Dista panc with lymp disse
Hubbard, L., et al[12] 2020	66	M	MRI: 10.3 cm mass	Head of the pancreas and duodenum [a]	No	Yes	Yes, *Escherichia coli*, streptococcus salivarius group	Systemic involvement: weight loss, fatigue s/p orthotopic heart transplantation	Ceftri metro
Afzal, R.M., et al[9] 2023	74	M	CT: infiltrating mass	Head of the pancreas [a]	No	No	Yes, *E. coli* and *Enterococcus faecium* in biopsies.	Fevers, chills, pseudocyst in pancreatic head	Amox clavu
Lilong G, et al Present study	39	F	CT: fluid collections measuring 4.8 x 5.1 cm	Body and tail of the pancreas [a]	No	No	No	Abdominal pain, Pseudocyst in pancreatic tail	panc and s
a. Primary pancreatic involvement									
b. Secondary pancreatic involvement									

Among the eight documented cases, five were found to involve primarily the pancreas[9, 11, 12, 15, 13, 14], similar to our case, which developed in the context of chronic pancreatitis and pseudocyst formation. Interestingly, one patient presented with moderately well-differentiated pancreatic adenocarcinoma, suggesting that malakoplakia can be associated with malignancy [14]. This association has been documented in the gastrointestinal tract, liver, and stomach[16]. The development of malakoplakia in patients with malignancies may be a histiocytic response to cancer or inflammation, although the exact cellular mechanisms involved remain unclear. In our review, with the exception of one patient with malignancy[14], two patients were immunosuppressed, and one was born with thymic and lymphoid hypoplasia[17, 12]. The immunosuppressed state associated with malignancy may play a role in the development of malakoplakia. As our patient was not immunosuppressed, showed no evidence of infection, remained afebrile, had negative blood culture and uranalysis, and did not receive antibiotic treatment, the pathogenesis of malakoplakia was unclear. Although the likelihood of an underlying malignancy in our patient was low, comprehensive evaluation of the lesions was undertaken to exclude a coexistent neoplasm. The exclusion of malignancy is imperative before pursuing a non-surgical management strategy in such cases given the association between malakoplakia with neoplasm[18, 19]. Lastly,

it is essential to raise awareness among surgeons that pancreatic malakoplakia, although exceedingly rare, should be considered in the differential diagnosis in patients with chronic pancreatitis and possible pancreatic pseudocysts.

Malakoplakia often presents as a mass, mimicking cancer and complicating the diagnosis, especially when it is extensive or accompanied by systemic symptoms. Four patients presented with systemic symptoms such as weight loss, anemia, fatigue, and low-grade fever[10–12, 17]. One patient even experienced recurrence at another site later in life[10]. The imaging findings could be very predictable on magnetic resonance (MR) which would be expected of a granulomatous mass, resulting from abundant foamy histiocytic reaction[12, 20–22]. However, our case differed in that no mass was observed, and imaging revealed a pseudocystic lesion with possible necrosis.

Overall, based on the clinicopathologic analysis of nine cases here, preoperative suspicion for malakoplakia should be amplified when a constellation of clinical and radiological findings is present including immunosuppressed status, recurrent and resistant infections- particularly *E. coli* or gram-negative bacteria- and presence of mass lesion on imaging studies. Although these features may raise clinical concerns for malakoplakia, a definitive diagnosis relies on histopathological evaluation of sampled tissue.

When it comes to differential diagnoses, conditions characterized by histiocytic or granulomatous infiltration should also be considered. Chronic granulomatous inflammation such as mycobacterial and fungal infections, can frequently mimic the entity, as these lesions often exhibit aggregates of epithelioid cells and giant cells. However, they lack the diagnostic Michaelis–Gutmann bodies. Additionally, nonspecific histiocytic infiltrates within the wall of a pseudocyst can also resemble malakoplakia, but they do not display the characteristic inclusions. Rosai–Dorfman disease may enter the differential due to its histiocytic proliferation, but it is distinguished by the hallmark features of emperipolesis and by the presence of enlarged non-Langerhans histiocytes with abundant eosinophilic cytoplasm, oval nuclei, and prominent nucleoli rather than foamy histiocytes with laminated inclusions. Rarely, signet ring cell carcinoma can mimic malakoplakia when malignant cells with cytoplasmic mucin droplets form targetoid inclusion. Nonetheless, the presence of nuclear atypia and epithelial marker expression facilitates accurate distinction. Furthermore, metastatic clear cell renal cell carcinoma may mimic the foamy histiocytes of malakoplakia due to its clear cytoplasmic morphology. However, careful evaluation of cytologic atypia and immunohistochemistry allows separation of the two.

Conclusion

Pancreatic malakoplakia is a rare but significant differential diagnosis in patients with chronic pancreatitis who present with mass-like lesions or pseudocysts. This case highlights the diagnostic challenges and the importance of histopathological confirmation. A review of previously reported cases in English-language literature underscores the complexity of treatment and etiology.

Abbreviations

GERD
gastroesophageal reflux disease
WBC
white blood cell
CT
computed tomography
PAS-D
periodic acid-Schiff with diastase
FNA
fine-needle aspiration
MR
magnetic resonance
MRI
magnetic resonance imaging

Declarations

i. **Funding:** Not applicable

ii. **Conflicts of interest/Competing interests:** Author Lilong G., Author David L., and Author Chadi H. declare that they have no conflicts of interest.

iii. **Ethics approval:** All procedures performed in studies involving human participants were in accordance. with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards

iv. **Consent to participate:** Informed consent was obtained from all individual participants included in the study.

v. **Written Consent for publication**: Informed consent was obtained from all individual authors and patients included in the study.

vi. **Availability of data and material:** Not applicable

vii. **Code availability:** Not applicable

viii. **Authors' contributions:** Author L.G. collected the data and drafted the manuscript; Author D.L. contributed to the revision; Author C.H. contributed to the analysis and revision of the manuscript.

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Figures

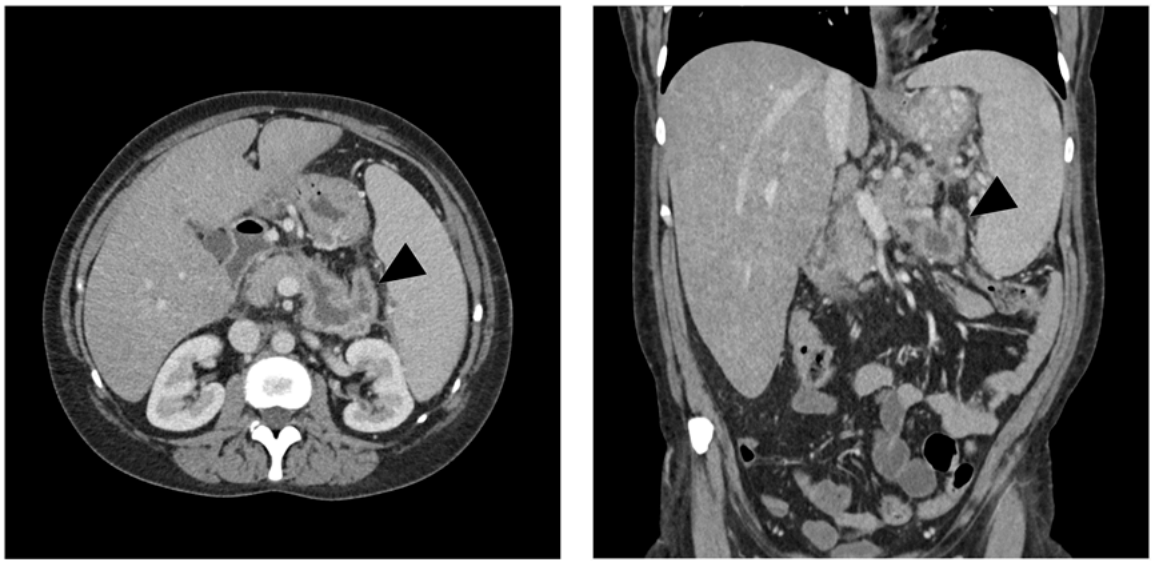


Figure 1

Figure 1

Abdominal CT revealed athick-walled, amorphous collection inferior to the body and tail of the pancreas measured 4.8 ×5.1 cm (A). Additionally,1.2 cm of fluid was collected superior to the tail of the pancreas (B).

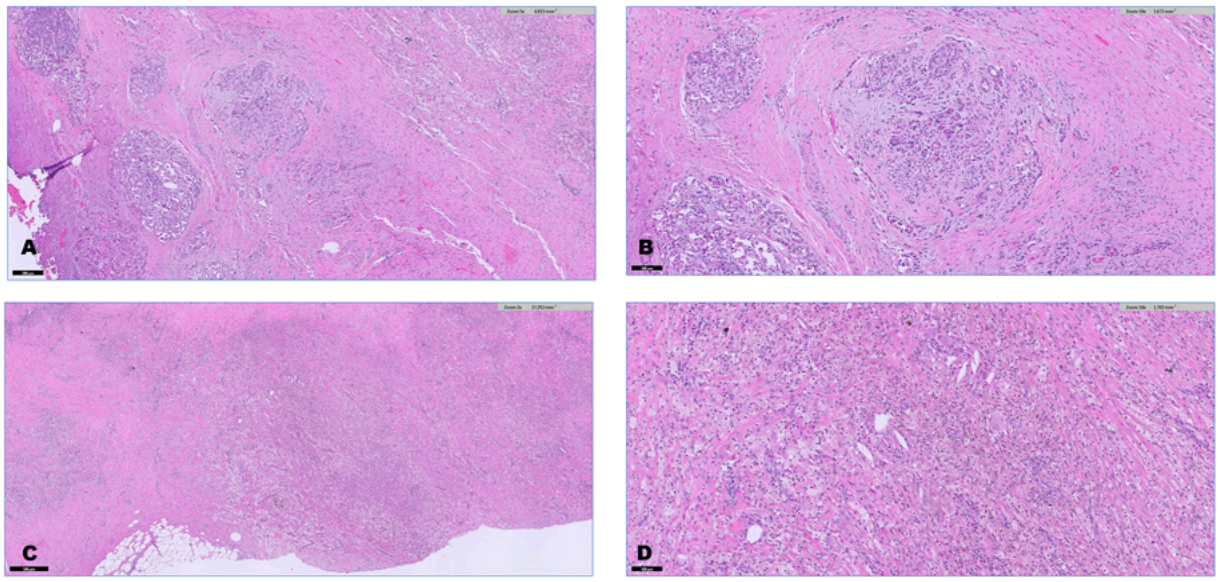


Figure 2

Figure 2

Histological examination (H&E) revealed chronic pancreatitis with intralobular fibrosis, loss of acinar tissue (A, 5X; B, 10X) and pseudocyst formation with crystal clefts and inflammatory infiltration in the cyst wall (C, 2X; D, 10X).

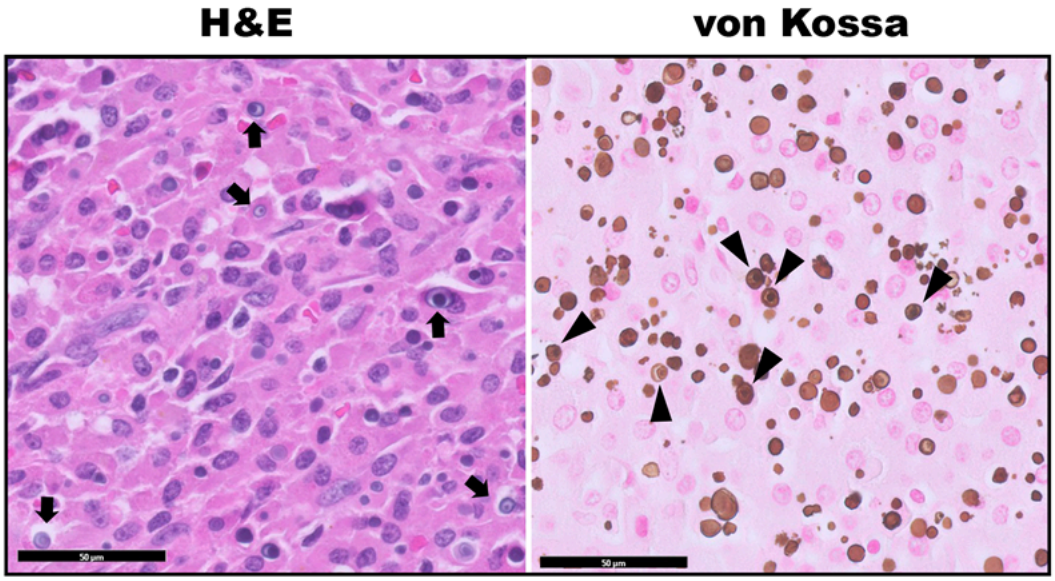


Figure 3

Figure 3

The H&E section demonstrates targetoid histiocytes (arrows); von Kossa staining highlights diffuse intracytoplasmic targetoid inclusions (arrowheads) known as Michaelis-Gutmann bodies. (A, H&E, 40X; B, von Kossa stain, 40X).

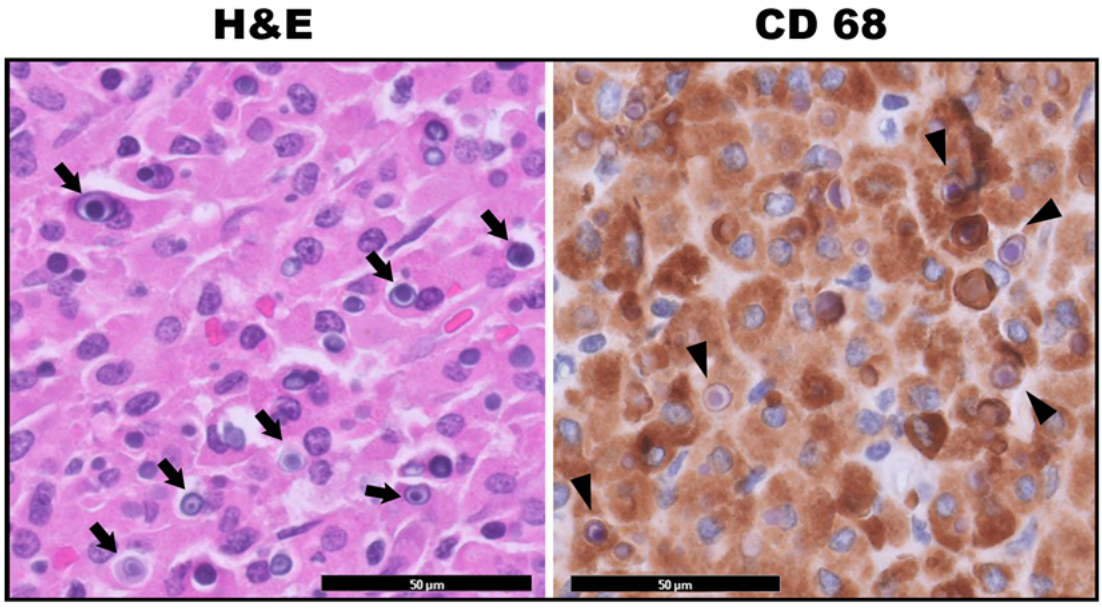


Figure 4

Figure 4

Histological examination (H&E) revealed chronic pancreatitis with intralobular fibrosis, loss of acinar tissue (A, 5X; B, 10X) and pseudocyst formation with crystal clefts and inflammatory infiltration in the cyst wall (C, 2X; D, 10X).