



MALAKOPLAKIA OF GALL BLADDER

A CASE REPORT AND A LITERATURE REVIEW

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INTRODUCTION

MALAKOPLAKIA



Malakoplakia (MP) is a rare granulomatous disorder that results from phagolysosomal defect, mostly involving urinary tract.



Malakoplakia in gall bladder (GB) is **extremely rare**; comprehensive search revealed only 8 case reports.



Malakoplakia clinically mimics other lesions like xanthogranulomatous cholecystitis and GB carcinoma.




Therefore, recognition of MP in GB is important for surgeons and histopathologists, in order to set a proper plan for management and avoid unnecessary extensive surgery.



PATHOGENESIS

Although more than a century has passed since Malakoplakia's original recognition, **the exact pathogenesis has not been fully recognized.**



SUGGESTED HYPOTHESIS FOR MP PATHOGENESIS

HYPOTHESIS NO. 1

Malakoplakia is thought to result from defective phagolysosomal activity which may be due to decreased intracellular concentration of cGMP which results in deficient fusion of lysosomes with phagosome.

Partially digested bacteria accumulate in monocytes or macrophages and lead to the deposition of calcium and iron on residual bacterial glycolipid, resulting in accumulation of inclusion structures in cytoplasm of histiocytes, the Michaelis–Gutmann bodies which are considered to be pathognomic of malakoplakia.

The defect may exhibit the expression of a genetic disorder or an improper immune response like that observed in alcohol abuse, malnutrition, post organ transplant, immunosuppressive drugs such as steroids or cytotoxic agents, malignancy and chronic diseases such as diabetes mellitus and autoimmune disease.

HYPOTHESIS NO. 2



PATHOGENESIS


Malakoplakia has been described also **in association with several microorganisms:** *E. coli* usually seen in cases of urinary MP.




PATHOGENESIS

In our case:

The patient was nondiabetic female, free of any of reported risk factors → the matter which refers to the possibility of **multifactorial pathogenesis** of this disorder.





PRESENTATION & MEDICAL DECISION

There are only few reported cases of malakoplakia in the gall bladder, wherein, it is usually presented as:

- wall thickness as in the following case
- mass lesion

Open cholecystectomy is indicated due to diagnostic suspicion of carcinoma as in the following case.



CASE REPORT OBJECTIVE

- This case report presents a case of gall bladder wall thickness, clinically suspected to be carcinoma and then revealed as MP on histopathology.
- The condition is discussed in the context of other causes of gall bladder wall thickness.



CASE PRESENTATION

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A 65 year-old nondiabetic female patient, presented with a complaint of **upper abdominal discomfort**.



Abdominal examination showed **palpable non-tender mass in the right hypochondrium**.



Signs and symptoms of acute pancreatitis were negative.

CASE PRESENTATION



Investigations:

- Blood analysis, liver function, renal function tests, Serum amylase and lipase were within normal levels.
- **Abdominal U/S** revealed contracted gall bladder with multiple stones and wall thickening.



Clinically:

GB carcinoma was suspected.



Operation:

Open cholecystectomy was performed and GB specimen was referred to histopathological examination (HPE).



HISTOPATHOLOGY

GROSS PICTURE

- Gall Bladder specimen measuring **10x6x4 cm with thickened wall.**
- Dissection showed **greenish velvety mucosa with areas of ulceration.**
- No masses were detected.
- **Three blackish stones** were also delivered.



MICROSCOPIC PICTURE

- Sections from gall bladder wall revealed **subtotally denuded mucosal lining with thickening of the wall.**
- Ulcerated area and lamina propria showed **sheets of foamy macrophages with rounded, concentrically layered intracytoplasmic inclusions (Michaelis-Gutmann bodies)**
- There was no evidence of granuloma, polyps, dysplasia or malignancy .

(MICHAELIS-GUTTMANN BODIES)

FIGURE 1. MEDIUM POWER PHOTOMICROGRAPH SHOWING SHEETS OF HISTIOCYTES INCLUDING THE CHARACTERISTIC MICHAELIS-GUTTMAN BODIES, (H&E X200).

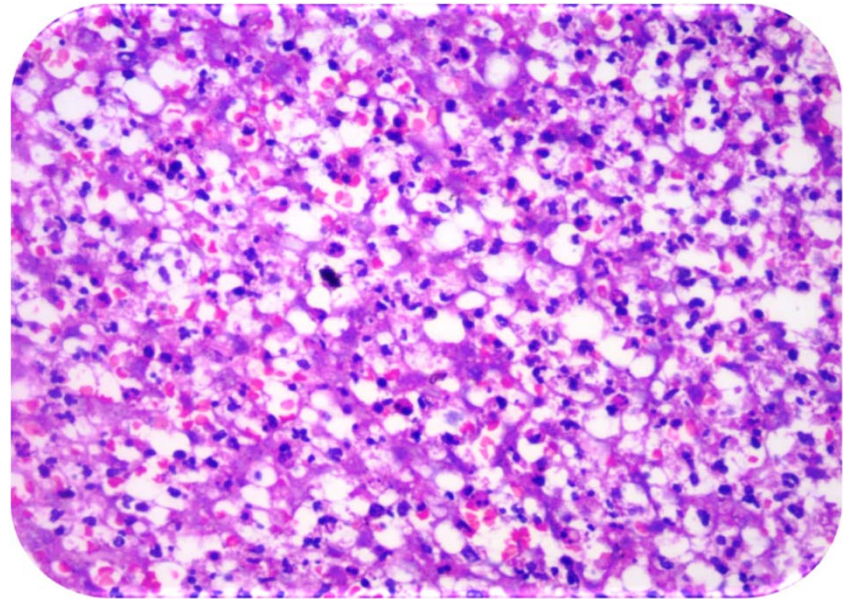
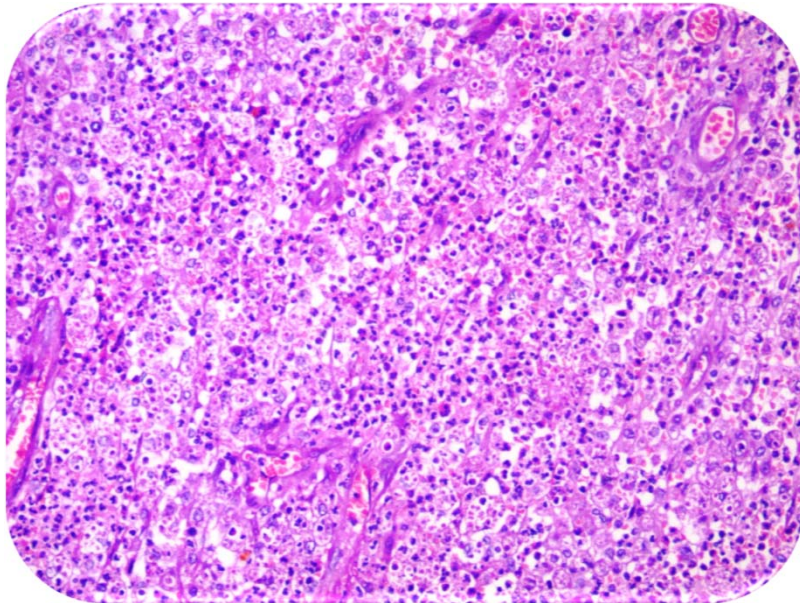


FIGURE 2. HIGH POWER PHOTOMICROGRAPH SHOWING THE CHARACTERISTIC MICHAELIS-GUTTMAN BODIES, (H&E X400).



MICROSCOPIC PICTURE

FIGURE 3. HIGH POWER PHOTOMICROGRAPH SHOWING THE CHARACTERISTIC MICHAELIS-GUTTMAN BODIES POSITIVELY STAINED BY PERIODIC ACID SCHIFF STAIN, (PAS X₄₀₀).

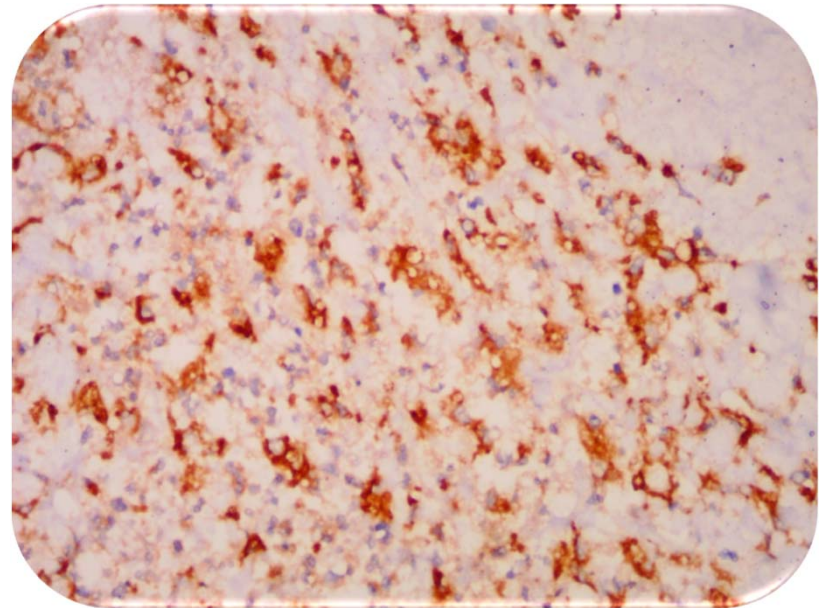
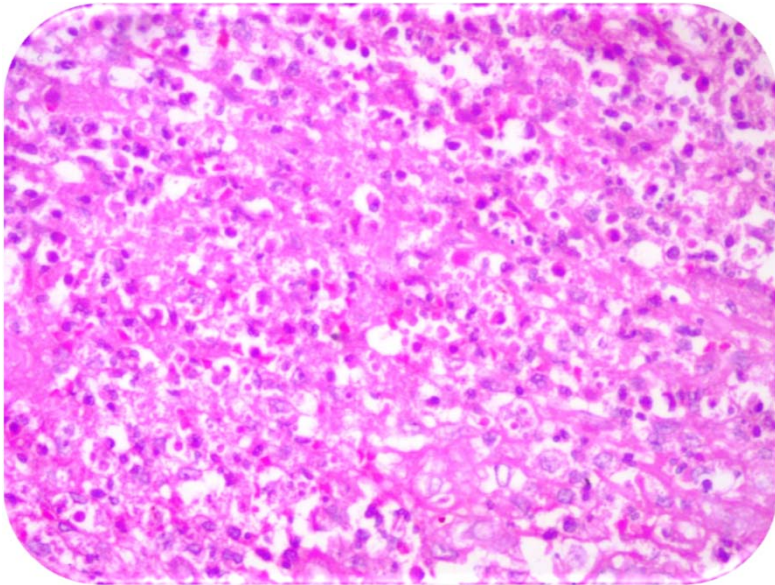


FIGURE 4. HIGH POWER PHOTOMICROGRAPH SHOWING CD68 STRONG EXPRESSION IN HISTIOCYTES SHEETS, (CD68X₄₀₀).



MP MIMICKERS

MP MIMICKERS

HPE is necessary to differentiate MP from its mimickers:

Xanthogranulomatous cholecystitis and malakoplakia that can present in a similar pattern. Both are thought to be part of spectrum of chronic inflammatory pathology, with difference that malakoplakia is more aggressive and shows the presence of both intracellular and extracellular Michaelis–Gutmann bodies as in our case.

Gallbladder involvement in autoimmune pancreatitis as a part of IgG4-associated systemic disease is also a common mimicker.

In our case, this was excluded by lack of characteristic laboratory investigations and by the absence of characteristic histopathological findings of this disease.

MP OUTCOME & MEDICAL APPROACH

Although malakoplakia is considered as a benign pathological process that requires local excision, it has been observed that many of these cases have aggressive course including high disease recurrence with fistulisation and poor response to antibiotics.

More specific therapy with drugs that concentrate in macrophages such as Bethanechol, a choline agonist, has been recommended in combination with antibiotics and surgery with the principle that Bethanechol may correct the decreased cGMP levels that are believed to interfere with successful phagolysosomal activity.

MP OUTCOME

ascorbic acid has been used also to increase the cGMP and cAMP levels in monocytes, which may offer an effective strategy for therapy, though it is still under trial.

Discontinuation of immunosuppressive drug therapy is usually needed to treat malakoplakia effectively

Attempts of curative surgery would appear to be supported in all cases of gall bladder MP and its mimickers; gall bladder carcinoma and xanthogranulomatous cholecystitis due to the difficulty of distinguishing them radiologically.

OUTCOME IN THIS CASE



- The patient's postoperative period was non-eventful. She was given broad-spectrum antibiotics.
- She is symptom free and on regular follow up.



CONCLUSION

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Diagnosis of MP presenting as GB wall thickness is a diagnostic dilemma faced by surgeons, radiologists and pathologists.

The conclusion of this case report is to stress upon keeping MP in mind as a potential differential diagnosis for GB carcinoma and granulomatous cholecystitis.

Recognition of MP in GB and differentiating it from its mimickers by histopathology is important for surgeons in order to set a proper plan for management and avoid unnecessary extensive surgery.



THANK YOU



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