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Malakoplakia of the Urogenital Tract

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ABSTRACT

Malakoplakia is a rare, granulomatous condition most commonly found in the genitourinary tract. It can present in a myriad of ways depending on the organ involved, thus presenting a huge diagnostic challenge. We present 4 patients with genitourinary malakoplakia, who manifested with recurrent urinary tract infection (UTI) and hematuria in all except one, who presented with hydronephrosis secondary to a large pelvic mass. We discuss the need for a high index of suspicion and careful scrutiny of histology to order to avoid misdiagnosis as simple long term antibiotics are an effective treatment in all but those with large pelvic masses.

Introduction

Malakoplakia is defined histopathologically by Michaelis–Gutmann bodies and whilst the etiology is uncertain, it is believed to be associated with defective macrophage function. It can occur in almost any part of the body but is most commonly found in the genitourinary tract. Because it is so rare and can present in a number of different ways, malakoplakia poses a difficult diagnostic challenge. We report four cases of malakoplakia identified from the uro-pathology database at Queen Elizabeth Hospital Birmingham (QEHB) over the last 10 yrs.

Case histories

All patients were women with a mean age of 55 years (range: 49–65 yrs) with all suffering from asthma, menstrual irregularities and lower urinary tract symptoms (LUTS). The presenting symptomatology was recurrent UTI and hematuria in all except one who presented with upper tract obstruction secondary to a large pelvic inflammatory mass. Upper tract investigations were normal in the three patients with recurrent UTI. Cystoscopy and bladder biopsies were undertaken which on histological assessment showed that the lamina propriae was infiltrated by foamy macrophages containing Michaelis–Gutmann bodies. These patients were treated with low dose prophylactic antibiotics with symptomatic improvement and no further UTIs. Mean follow up was 50 months (range: 25–108 months). One of the patients has stopped her antibiotics after one year with no recurrence of symptoms. The lesions remained static on flexible cystoscopy after treatment.

The patient with pelvic malakoplakia is a 49 yr old lady with Type II diabetes, obesity, asthma and congestive cardiac failure. She initially presented with an incidental finding of deranged renal function and underwent imaging with computerized tomography (CT) and magnetic resonance imaging (MRI) as shown in Fig. 1 and Fig. 2. These revealed abnormal soft tissue at the aortic bifurcation and pre-sacral area forming a dense fibrotic inflammatory reaction involving a large left ovarian cyst, the uterus, bladder, small bowel and both ureters causing bilateral hydronephrosis. The initial thought was that of intra-abdominal malignancy or retroperitoneal fibrosis but further histology was compatible with a diagnosis of malakoplakia. The patient was due to have laparotomy for removal of ovarian mass and ureterolysis, with pre-operative bilateral ureteric stenting, but was deemed unfit at pre-operative assessment. The patient received retrograde ureteric stents meanwhile, leading to an improvement in renal function. The patient has received long term antibiotics which included a combination of rifampicin, ciprofloxacin and amoxicillin, with repeat CT showing a minor degree of shrinkage in size of the pelvic mass and some symptomatic benefit.

Discussion

Malakoplakia was first reported by Michaelis and Gutmann[1] in 1902, but it was Professor von Hansemann who first identified the condition a year earlier. It is defined histologically by von Hansemann histiocytes and Michaelis–Gutmann bodies.[2] Von Hansemann cells are ovoid histiocytes which contain intracytoplasmic

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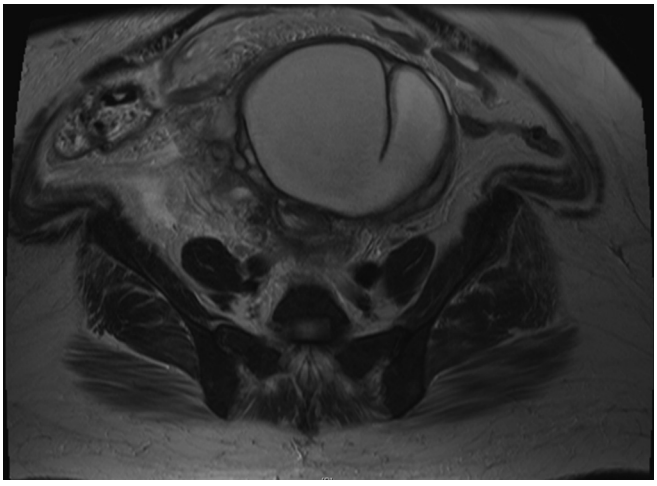

Figure 1. MRI pelvis of patient with pelvic malakoplakia showing large pelvic mass.

bodies: Michaelis–Gutmann bodies. They have specific staining characteristics, being gram-negative and positive for alizarin red and von Kossa stain (calcium), Perls' stain and Prussian blue (iron), and Periodic Acid-Schiff stain. Fig. 3 shows histology taken from one of the patients demonstrating H&E staining characteristics of Michaelis–Gutmann bodies. Whilst etiology is unclear, it is believed that malakoplakia occurs due to defective bacterial digestion, leading to calcium and iron accumulation on the residual bacterial glycolipid. This is found within the pathognomonic Michaelis–Gutmann bodies.

The genitourinary tract is most commonly involved area in patients with malakoplakia, mostly affecting the bladder, prostate, ureter, kidney, female genital tract and retroperitoneal tissue. In our series the most common site was the bladder (three patients) and the fourth patient had a large pelvic inflammatory mass. The presentation of malakoplakia of the genitourinary tract can be very varied depending on the area involved and is often not diagnosed until symptoms have persisted for quite some time.

Malakoplakia of the bladder usually presents with recurrent UTIs, lower urinary tract symptoms or hematuria.[3] Typically *Escherichia coli* is grown in urine culture and upon flexible cystoscopy, yellow brown soft plaques with a central umbilication are the most frequent finding. Malakoplakia of the bladder has also been found in association with bladder tumors[3] with or without a history of infection. This makes the accurate identification of malakoplakia especially important in the context of a suspicion of malignancy.

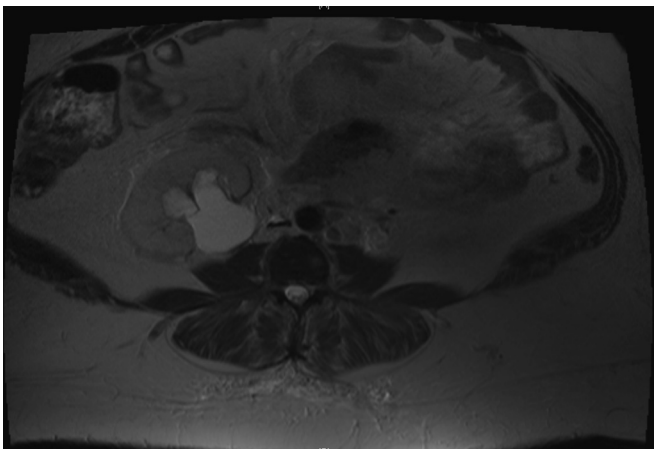

Figure 2. MRI pelvis of patient with pelvic malakoplakia showing hydronephrosis of right kidney.

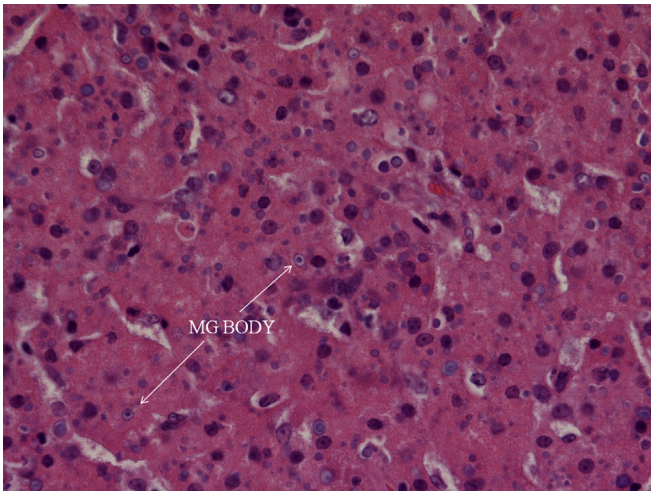

Figure 3. 400× High power hematoxylin and eosin (H&E) stain showing sheets of foamy macrophages and Michaelis–Gutmann (MG) bodies.

Malakoplakia arising from retroperitoneal organs has the potential to spread throughout the retroperitoneum resulting in extensive pelvic malakoplakia. This most commonly occurs in malakoplakia of the bladder and the female genital tract. The mechanism of pelvic mass formation is unknown. It is presumably due to aggressive immune reaction to a urinary tract pathogen which causes scar tissue around the urogenital organs which then involves adjacent viscera forming a mass. The consequences depend on the organs involved but can cause renal failure if both ureters are compressed such as the patient in our series with extensive pelvic malakoplakia. Extensive pelvic malakoplakia can often be mistaken for invasive cancer[4] on CT and so careful scrutiny of biopsies are vital.

Treatment of malakoplakia is mostly medical with surgical intervention sometimes being necessary. There are no widely established guidelines for the medical treatment of malakoplakia but most approaches involve antibiotics[5] which work intracellularly such as quinolones (mainstay of treatment), trimethoprim and rifampicin to aid the defective phagolysosomal mechanism found in malakoplakia. These can be used long-term at low doses to prevent recurrence. Moreover, as immunosuppression has been implicated in malakoplakia, patients on immunosuppressants may need to discontinue them depending on the risk to benefit ratio. It is interesting to note that all 4 of our patients with malakoplakia had a history of asthma. Whilst this may be a coincidence, this could perhaps be tentatively explained by the fact that asthma is a hypersensitivity immune disorder and immune deficiencies have been associated with malakoplakia.

Surgical treatment may be necessary depending on the organ(s) affected. Vesical malakoplakia may occasionally require transurethral resection if the lesion is large or obstructing the ureters, in addition to nephrostomy/ureteric stenting. Patients with extensive pelvic malakoplakia will often need large, complex abdominal surgery especially if the bowel is involved.

Conclusion

Malakoplakia of the urogenital tract is a rare condition which requires careful histological scrutiny to diagnose correctly. We illustrate with these 4 cases that most patients, once accurately diagnosed, can be successfully treated with long term low dose antibiotics. However, patients with large pelvic masses may require further surgical intervention.

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