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CASE REPORT

Testicular malakoplakia

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Malakoplakia may affect any part of the body, but it is most likely found in the urinary tract in susceptible patients. We present a case of testicular malakoplakia in a male patient who presented with a history of a swollen, painful right testis and a progressively enlarging ulcerating lesion overlying the right hemi-scrotum. After minimal response to a course of antibiotic therapy, we proceeded to a right orchidectomy with high ligation of the spermatic cord and a right hemiscrotectomy with a good result. Histopathological analysis demonstrated Von Hansemann histiocytes and Michaelis-Gutmann bodies, pathognomonic of malakoplakia.

Keywords: malakoplakia, tumour, ulcer, testes, orchitis, orchidectomy, hemiscrotectomy, urinary tract

Case presentation

A 44-year-old HIV-positive African male (CD4 count 55 cells/ul) presented with a six-month history of a swollen, painful right testis and a progressively widening ulcerative lesion of the scrotal skin overlying his right testis. The patient volunteered that he had received treatment at a local primary healthcare facility for possible right testicular infection six months prior with only minimal improvement in symptoms. During the physical examination, a 10 × 8 cm, well-demarcated ulcerative lesion was noted in the skin of the right hemi-scrotum (Figure 1). The underlying testes and epididymis were hard and tender, and adherent to the ulcerative lesion. The left testis, penis and perineum appeared normal, and no regional nodes were palpable. His white cell count was 6.38 × 10^9/L, C-reactive protein 13 mg/L and testicular tumour markers all within the normal range. Scrotal ultrasound revealed an enlarged right testis (46 × 37 mm) with distortion of its normal architecture and multiple poorly-vascularised, fluid-filled areas suggestive of necrosis (Figure 2). The left testis appeared unremarkable. A wedge biopsy of the ulcerative scrotal skin lesion revealed extensive xanthogranulomatous inflammation, with no evidence of dysplasia or malignant change. The Ziehl Neelsen stain was negative for mycobacteria. A course of antibiotics had a minimal response, and we proceeded with a right orchidectomy with high ligation of the spermatic cord and a wide excision of the scrotal ulcer by right hemiscrotectomy. The testis was found to have been entirely replaced by a soft, flesh-textured tan-yellow mass (Figure 3), and histopathology noted an inflammatory infiltrate comprising lymphocytes, plasma cells and Von Hansemann histiocytes, as well as extensive areas of fibrosis and necrosis. Numerous intra- and extracytoplasmic laminated concretions (Michaelis-Gutmann bodies) were pathognomonic of malakoplakia. The diagnosis was confirmed by positive Von Kossa and Prussian blue stains for Michaelis-Gutmann bodies. Although the Ziehl-Neelsen stain was negative for mycobacteria, the mucicarmine and Grocott's stains proved negative for fungi. The patient's postoperative course was

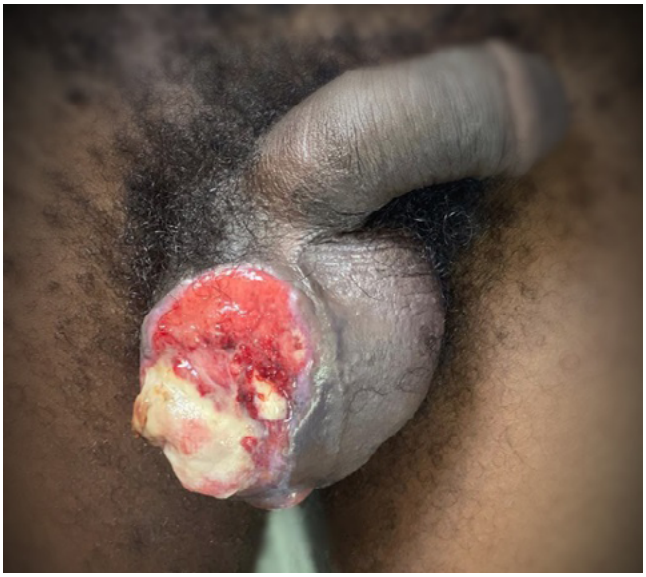

Figure 1: Photograph showing a 10 × 8 cm, well-demarcated ulcerative lesion on the right hemi-scrotum

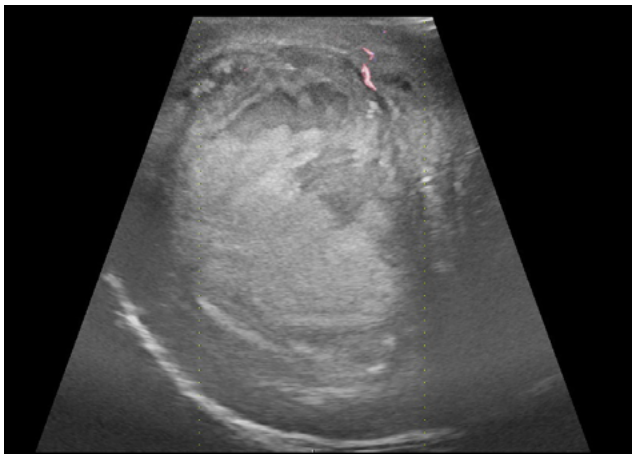

Figure 2: Sonogram of the right testicle demonstrating partially confluent curvilinear areas of hypoechogenicity, which demonstrate no significant vascularity

Figure 3: Photograph showing the bivalved testis almost entirely replaced by a soft, flesh-textured tan-yellow mass

uneventful and he was fully recovered at discharge. On follow-up 12 months later, there was no evidence of recurrence.

Discussion

Malakoplakia (from the Greek word meaning "soft plaque") is a rare granulomatous inflammatory disease described first by Michaelis and Gutmann in 1902 and Von Hansemann in 1903. It was only decades later, in 1958, that the first case of testicular malakoplakia was described.[1] It most commonly involves the urinary tract but can also occur in any other part of the body, including the genital tract, gastrointestinal system, bones, lungs, lymph nodes, skin and retroperitoneum, and in these cases, it is associated with considerable morbidity.[2] Involvement of the testes is uncommon, and patients with testicular disease present either with a painless, intrascrotal mass simulating a testicular tumour or with recurrent epididymo-orchitis.[3,4]

The exact pathogenesis of malakoplakia is unknown and likely multifactorial. Factors implicated in its pathogenesis include the altered phagocytic function of macrophages, gram-negative infection, and an abnormal immune response.[5] Reported cases have mostly occurred in middle-aged men who are immunosuppressed, debilitated or suffering from chronic medical conditions.[6,7] An imbalance between cyclic adenosine monophosphate (cAMP) and cyclic guanosine monophosphate (cGMP) is thought to cause deficient lysosomic degranulation in phagocytes. This impairs their ability to completely digest bacteria, mainly *Escherichia coli*.[8] Partially-degraded bacteria or bacterial fragments then form a nidus for the intracellular deposition of iron and calcium phosphate crystals. Periodic acid–Schiff (PAS) positive histiocytes containing such lamellated iron and calcium inclusions, called Michaelis-Gutmann bodies, are pathognomonic of the disease. The calcium in these bodies is identified with a Von Kossa stain and the iron is identified with a Prussian blue stain.

The differential diagnosis of testicular malakoplakia includes idiopathic granulomatous orchitis, Leydig cell tumour, other testicular tumours and inflammatory processes such as tuberculosis, histoplasmosis and cryptococcosis. Intratubular multinucleated giant cells are found in idiopathic granulomatous orchitis, whereas in malakoplakia, giant cells are mostly absent. Leydig cell tumours may contain mononucleated or binucleated cells with abundant cytoplasm. Reinke crystalloids (absent in malakoplakia) are identified in up to 40% of cases of Leydig cell tumours (in which Michaelis-Gutmann bodies are absent).[9]

Medical therapies for malakoplakia have had limited success. Antibiotics are the mainstay of treatment, particularly those effective against *Escherichia coli*. Antibiotics that are taken up and concentrate in macrophages, such as fluoroquinolones, are the first choice. Other agents with reported efficacy are rifampicin, co-trimoxazole and gentamicin.[3,6] If successful, these antibiotics can be used for a longer period at lower doses to prevent a recurrence. Other medications such as bethanechol, a choline agonist, and ascorbic acid are thought to increase intracellular cGMP and restore lysosomal activity against bacteria. Discontinuing immunosuppressive medication may be an option if the benefit outweighs the risk to the patient.[7] Despite its benign nature, testicular malakoplakia often ends in surgery. An orchidectomy is usually the outcome if there is an inadequate response to medical therapy and to exclude testicular malignancy. Our patient had minimal improvement on antibacterial treatment and underwent an orchidectomy and hemiscrotectomy, which proved both diagnostic and therapeutic.

Although malakoplakia rarely involves more than one anatomical site in the same patient, subsequent radiological investigations and a cystoscopy ruled out other areas of genitourinary involvement in our patient. The prognosis after an orchidectomy is usually good, and recurrence is rare. However, some authors advocate that patients should be followed up periodically to monitor for a possible recurrence.[10] The exact nature of follow-up (i.e. frequency and the length of follow-up) has not yet been determined.

Testicular malakoplakia is a rare condition that should be considered in the differential diagnosis of patients with diffuse enlargement and sonographic parenchymal abnormalities of the testis, especially in an immunocompromised patient.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

No ethical approval was required by the institution.

Informed consent

Written informed consent was obtained from the patient for the anonymised information and the accompanying images to be published in this article.

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References

1. Haukohl R, Chinchinian H. Malakoplakia of testicle - report of a case. Am J Clin Pathol. 1958;29:473-8. https://doi.org/10.1093/ajcp/29.5.473.
2. Son SM, Woo CG, Lee HC, Yun SJ, Lee OJ. Retroperitoneal malakoplakia mimicking sarcoma: a case report. Int J Clin Exp Med. 2017;10(8):12663-6.
3. Hina S, Hasan A, Iqbal N, Shabbir MU, Sheikh AAE. Malakoplakia of the urinary bladder and unilateral ureter. J Coll Physicians Surg Pakistan. 2019;29(6):582-4. https://doi.org/10.29271/jcpsp.2019.06.582.
4. Kostakopoulos A, Giannakopoulos S, Demonakou M, Deliveliotis C. Malakoplakia of the testis. Int Urol Nephrol. 1997;29(4):461-3. https://doi.org/10.1007/BF02551114.
5. Bandrés SÁ, Guarch Troyas RM, Cebrián Lostal JL, et al. Malacoplaquia testicular: Aportación de un nuevo caso y revisión de la literatura científica. [Malacoplakia of the testis: a new case report and a literature review.] Actas Urol Esp. 2009;33(9):1028-31. https://doi.org/10.1016/S0210-4806(09)72905-5.
6. Venyo A. Malakoplakia of testis and epididymis: a review of the literature. J Med Sci Technol. 2015;4(1):62-8. https://doi.org/10.9734/BJMMR/2015/17310.
7. Morgan TA, Rabban JT, Filly RA. Testicular malakoplakia: a rare sonographic mimic of malignancy. J Clin Ultrasound. 2015;43(3):199-202. https://doi.org/10.1002/jcu.22239.
8. Mahesh MS, Bhat S, Jose A. Malakoplakia of testis: a case report. J Med Sci Clin Res. 2017;05(02):18153-5. https://doi.org/10.18535/jmscr/v5i2.147.
9. Bostwick D, Cheng L. Urologic Surgical Pathology. 4th ed. London: Elsevier Inc.; 2020.
10. Dubhashi SP, Kumar H, Kulkarni V, Seleman AM. Malakoplakia of the testis. Surg Sci. 2014;5(4):233-5. https://doi.org/10.4236/ss.2014.55040.