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Case Report

Malakoplakia in sclera of the left eye- A rare case report

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ABSTRACT

“Malakoplakia” comes from Greek words malakos (soft) and plakos (plaque). It is a rare histiocytic disease that may occur in all organs, most frequently involving GU tract, particularly bladder. Other organs involved are gastrointestinal tract (most commonly colon, followed by stomach and duodenum), central nervous system and female genital tract. Malignancy and other forms of immunosuppression are predisposing factors. Malakoplakia in eye and orbit region are extremely rare and are usually appraised by the pathologist on histopathology sections with a few mimickers as differential diagnosis. Histopathological features showed. Collections of histiocytes with granular eosinophilic cytoplasm accumulated beneath the surface epithelium. Few cells showed round, concentrically layered intracytoplasmic inclusions known as Michaelis–Gutmann (MG) bodies which are characteristic of malakoplakia. The MG bodies are basophilic and are demonstrated by Periodic Acid–Schiff (PAS) stain and stains for iron and calcium. Rare histiocytic disease that can simulate a neoplasm. Gross appearance: Single or multiple white-yellow soft raised plaques on the mucosal surface. Pathological diagnosis/entity rather than a clinical impression and warrants special stains to arrive at the histological conclusion. More common in immunocompromised (HIV, renal transplant recipients) and women. Noted in > 50 years of age. Malakoplakia is presently regarded as a defect in the host macrophage (phagolysosomal) response to a bacterial infection, usually from gram-negative coliform bacilli. Malakoplakia should be kept as one of the differential diagnosis by the pathologist when dealing with ophthalmic specimen.

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1. Introduction

Malakoplakia (MPL) is a rare inflammatory disease of unknown etiology. The name has been derived from Greek malakos means soft, plakos-plaque.¹ It is known to affect both genders (especially women) and mostly people of younger age group.² MPL is a chronic granulomatous disease and also precedes or accompanied by an immunosuppressive state especially in old age and also can occur before. It is a benign condition and also self-limiting with mostly having good prognosis.² The disease was first described by Von Hansemann in 1901, and then a

year later Leonor Michaelis and Carl Gutmann declared the morphological characteristics of the disease.^{3,4}

Being a rare histiocytic disease it is known to occur in all systemic organs and known to occur commonly in the genitourinary tract, particularly bladder followed by gastrointestinal tract

It is known to co-exist with underlying malignant conditions especially in the genitourinary system. Though the etiopathogenesis of MPL is not fully known, impairment in bactericidal activity of macrophages had been widely accepted as an underlying mechanism in pathogenesis.

The definite diagnosis of MPL is usually arrived by histopathological studies of the affected tissue. Malakoplakia in eye and orbit region are extremely rare. To

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our knowledge only three cases of malakoplakia involving the eye had been reported that too among elderly patients.⁵ Literature states that Malakoplakia in orbital region is present with ulceration could carry away the clinician towards infective conjunctivitis and at instance ophthalmic tumours.

In the present case report, we encountered a middle-aged male presenting with swelling in left eye, otherwise asymptomatic case.

2. Case History

A thirty-nine years old male patient came with a complaint of swelling in left eye since 1 year. There was no history of watering or discharge from eyes.



Fig. 1: Swelling in the left eye-nasal side of sclera.

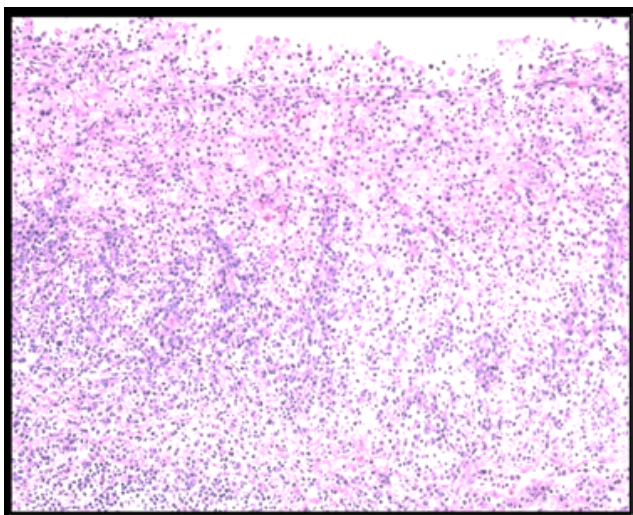


Fig. 2: Microscopic appearance showing mixed population of inflammatory cells and sheets of histiocytes in aggregates, H&E4X

Ultrasound procedure was done which revealed a scleral nodule in the left eye.

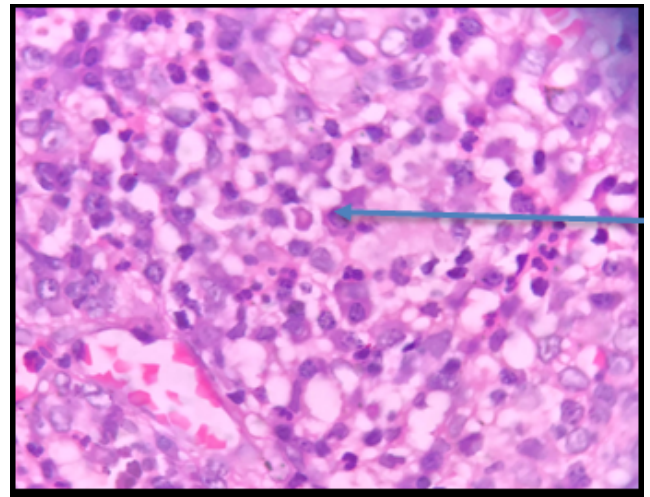


Fig. 3: Microscopic appearance shows histiocytes and inflammatory cells, Michaelis Guttman bodies. H&E (oil immersion)

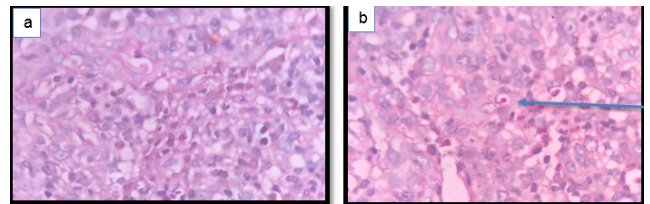


Fig. 4: Pas stain: **a,b:** Microscopic rounded basophilic intracytoplasmic inclusions that contain calcium and iron deposits, H&E (oil immersion)

Laboratory test done showed total count of 7700 cells/cu.mm with 45.9% neutrophils, 38.9% lymphocytes, 10.9% eosinophils, 0.8% basophils, 3.5% monocytes and platelet count of 281000/cu.mm and hb of 12.0 g/dl.

2.1. On Local Examination of left eye

Vision was normal, the posterior segment was normal and there was a firm tender nodule with overlying dilated conjunctival and episcleral vessel and minimal chemosis. Clinically infective conjunctivitis was suspected and due to the persistent nature of the swelling, left eye lamellar excision of scleral nodule was performed under LA for histopathological evaluation.

3. Discussion

Malakoplakia is a rare granulomatous disease that occurs commonly in the urinary tract, also seen in gastrointestinal tract, central nervous system, tongue and female genital tract.^{1,2} Grossly, malakoplakia can present as soft tan yellow plaques and nodules or even extensive bands. The lesion is usually solitary but also can be multiple.^{3,4}

The intracellular accumulation of partially degraded bacteria leads to development of granulomatous reaction and the pathognomonic Michelis Gutman bodies(MG)-calcified, basophilic, intracellular inclusions that stains positive for periodic acid Schiff stain which often appear as targetoid or owl's eye lesions.⁶ Because the bactericidal activity of macrophages is impaired, it may lead to infections, sepsis and bacteremia. Patients with malakoplakia are often found to be infected with bacteria which is *Escherichia coli*.^{7,8} Other bacterias in patients include *Klebsiella pneumoniae*, *Aerobacter aerogenes*, *Rhodococcus equi*, *Mycobacterium intracellulare*, *Corynebacterium* species, *Proteus* species, *Pseudomonas* species, *Acinetobacter* species, *Staphylococcus* species, *Streptococcus* species, and *Enterococcus* species.⁷

Histologically, it shows collections of histiocytes with granular eosinophilic cytoplasm accumulated beneath the surface epithelium (Figure 2). Few cells show round, concentrically layered intracytoplasmic inclusions known as Michaelis–Gutmann bodies(characteristic of malakoplakia)(Figure 3). The MG bodies are basophilic and demonstrated by periodic acid–Schiff(PAS) positive and stain for iron and calcium(Figure 4 a,b). The granulomatous process was accompanied by chronic inflammatory infiltrate (lymphocytes and plasma cells) and also acute inflammatory infiltrate (neutrophils). Stains used are PAS, Perls (iron) stain and von Kossa (calcium) for Michaelis-Gutmann bodies. The treatment depends on medical therapy in the form of multiple antibiotics that are concentrated in macrophages that is quinolone and trimethoprim-sulfamethoxazole.⁸

Malakoplakia needs to be differentiated from other infectious diseases, neoplastic and reactive/reparative processes. Infections to consider include tuberculosis, Whipple's disease, lepromatous leprosy, fungus such as *Cryptococcus*, and parasites like leishmaniasis. Special stains for microorganisms and tissue culture are necessary to differentiate.⁹

4. Conclusion

To conclude, Malakoplakia is a chronic inflammatory disease though very rare, still known to occur in an otherwise asymptomatic patient. Malakoplakia should be kept as one of the differential diagnosis by the pathologist while dealing with ophthalmic specimen in middle aged eye swelling especially when many histiocytic aggregates are encountered on histology. Clinical suspicion of malakoplakia should be raised in non-immunocompromised

patients presenting with asymptomatic eye swelling.

5. Conflict of Interest

The authors declare that there is no conflict of interest.

6. Source of Funding

None.

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