Cutaneous Malakoplakia - A Rare Clinical Entity

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Abstract

Introduction: Malakoplakia is a rare chronic granulomatous disease of infectious etiology. While any organ may be affected, it commonly involves the urogenital tract. Cutaneous Malakoplakia is even rarer.

Case presentation: We report a case of cutaneous malakoplakia in a patient suffering from Hansens disease.

Conclusion: Cutaneous malakoplakia is a rare clinical disorder, diagnosis of which can be confirmed on skin biopsy and treatment includes wide local excision and medical management including use of sulphonamides.

Keywords: Malakoplakia, Cutaneous Malakoplakia, Michaelis Gutmann body.

Introduction

Whenever a patient with multiple discharging sinuses presents to the surgeon a differential diagnosis of Koch's or Leprosy is primarily considered due to its high prevalence in India. Our patient being a known case of Leprosy, the same was considered as a provisional diagnosis. However, skin biopsy was suggestive of Cutaneous Malakoplakia.

Malakoplakia , a term derived from the Greek, meaning "soft plaque" [1,2], was first described in 1902 by Michaelis and Gutmann [3]. A rare granulomatous disease of infectious etiology triggered by bacteria, it occurs most often in subjects affected by primary or secondary immunodeficiency and results from abnormal macrophage function in response to a bacterial infection. Partially digested bacteria accumulate in monocytes or macrophages and lead to the deposition of calcium and iron on residual bacterial glycolipid. This results in formation of Michaelis Gutmann body which is pathognomonic for malakoplakia.

Case Presentation

A 45 year old diabetic , hypertensive male presented with complaints of multiple discharging sinuses over the suprapubic region since 1-2 months. The patient was a known case of Hansen's disease on medication which included Thalidomide 400 mg HS and Prednisolone 5mg twice a day. The patient was on this treatment on and off since the past 2 years. On physical examination, multiple discharging sinuses were seen in the suprapubic regionwith a yellowish and seropurulent discharge(Fig.1). There was local induration and the area was minimally tender on palpation. There were no palpable inguinal lymph nodes and there was no associated history of fever or weight loss. Routine hematological and biochemical investigations were within normal limits. Local ultrasonography was suggestive of heterogeneously ulcerated subcutaneous soft tissue thickening over the pubic region with multiple solid nodules, and lymphadenopathy. A culture and sensitivity of pus from the area showed growth of E.coli. for which the patient was started on appropriate intravenous antibiotics according to sensitivity reports. Accordingly, the patient was started on injectable Ciprofloxacin 500 mg IV 12 hourly. A preliminary incisional skin biopsy was performed which was suggestive

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of presence of Michaelis Gutman bodies typical of Malakoplakia. A complete surgical debridement was then carried out using a transverse elliptical suprapubic incision and the excised tissue sent for histopathological examination. Wide margins measuring approximately 1 cm. were sent seperately for evaluation of adequate excision. The histopathology was suggestive of skin covered tissue bits with dense infiltration of histiocytes and reticular nuclei along with lympholplasmic cells going deep into the subcutaneous tissue. Michaelis Gutmann bodies noted in the cytoplasm of histiocytes (Fig.2). The margins were reported as clear of disease. The patient was advised to discontinue Prednisolone and started on Sulphamethoxazole-Trimethoprim along with Ciprofloxacin for two weeks. The wound was left open to heal with secondary intention and , regular dressings were done. The wound healed completely uneventfully.

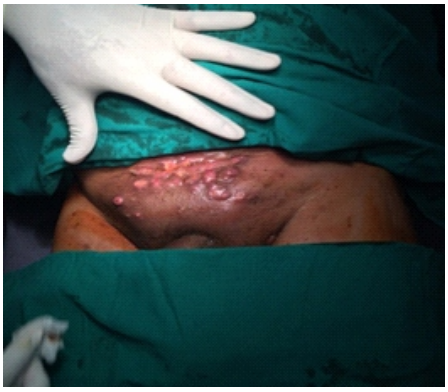

Figure 1. Pre operative photograph

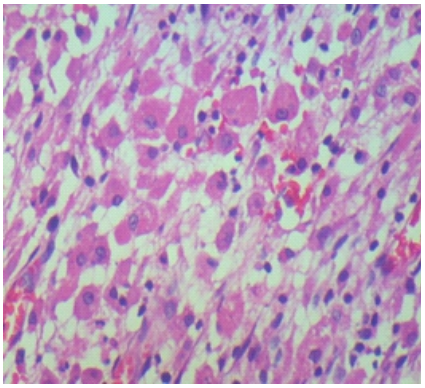

Figure 2. H&E slide showing Michaelis Gutmann body

Discussion

Malakoplakia results from inadequate killing of bacteria by macrophages or monocytes following defective phagolysosomal activity due to defect of the destructive capacity of macrophages following endocytosis which is considered to be the central event. Reduced intracellular cGMP level causes inadequate microtubular function and lysosomal activity, leading to an incomplete elimination of bacteria from monocytes and macrophages. Partially digested bacteria accumulate in monocytes or macrophages with subsequent deposition of calcium and iron on residual bacterial glycolipid. Disturbed phagosome-lysosome fusion has been suggested as the major cause; however it is still not clear why this disorder happens, and this hypothesis has not been unequivocally accepted [4-6]. While the most common site involved is the urogenital tract (75% of cases), less common sites include the gastrointestinal / respiratory system, retroperitoneum, thyroid, lymph nodes, bones/joints, middle ear and brain and skin [7]. Cutaneous presentation of Malakoplakia is extremely rare and was first reported in by Leclerc and Bernier in 1972 [8]. It is usually found in immunocompromised patients though it has also been reported in individuals without immunodeficiency as well. Our patient who was on prednisolone for a long duration when he reported to us therefore belonged to the first category. The condition is more prevalent in females as compared to males [9]. Approximately, 90% of patients have coliform bacteria detected in urine, blood, or tissue (E. Coli was seen on pus culture in our patient), suggesting an infectious cause but Klebsiella, Proteus, Pseudomonas, Mycobacterium avium, Mycobacterium tuberculosis, Shigella, Staphylococcus aureus and Enterococcus sp have also been detected on occasions [10,11]. Rhodococcus equi is the most commonly implicated microbe in HIV-infected patients [12]. Although, no typical clinical presentation is described, skin presentation varies from papules, plaques, nodules, abscesses with or without fluctuation, and fistula to ulcers, cystic and polypoid masses [13].

Differential diagnosis includes other infectious diseases or neoplastic and reactive/reparative processes. Infections which should be considered include tuberculosis, Whipple's disease, lepromatous leprosy, fungus (Cryptococcus), and parasites (leishmaniasis). Special stains for microorganisms and tissue culture are necessary. Reactive and neoplastic processes include Langerhans cell histiocytosis, fibrous histiocytoma, lymphoma, granular cell tumor, xanthoma, foreign-body granuloma, hemophagocytic syndromes, and sarcoidosis [14].

Discontinuation of immunosuppressives and treatment of HIV forms the main line of treatment. Surgical management includes excision of skin lesions and drainage of abscesses. Bethanechol, a choline agonist, corrects the decreased cGMP levels which has been implicated in the pathogenesis of malakoplakia. Quinolones and Sulphamethoxazole-Trimethoprim are used as they concentrate within the macrophages.

Conclusion

Malakoplakia is a rare disorder of granulomatous origin. Cutaneous Malakoplakia is even rarer and can result in chronicity since it is often treated as any other maculopapular skin lesion without a proper diagnosis. The condition can be suspected on clinical history and confirmed on skin biopsy. Management includes wide local excision at the earliest followed by medical treatment where the use of sulphonamides is universal. Stoppage of drugs causing immunodeficiency, if any, is mandatory.

Clinical Message

Cutaneous malakoplakia should be considered as a rare differential diagnosis in cases with multiple cutaneous discharging sinuses, especially in immunocompromised patients after other possibilities especially tuberculosis have been ruled out.

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