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Case Report

Malakoplakia of the Vulva Following Lung Transplantation

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Abstract

Malakoplakia is a rare histiocytic inflammatory disorder that predominantly affects the urinary tract and rarely the female genital tract. The pathogenesis is hypothesized to be a result of defective bacterial degradation within dysfunctional phagolysosomes, resulting in formation of the pathognomonic Michaelis-Gutmann bodies. Immunocompromised patients are more commonly affected. We present a case of malakoplakia of the vulva and left mainstem bronchus in a 67-year-old postmenopausal immunosuppressed female following bilateral lung transplantation. Multiple painful nodular lesions were present on the labia, which did not respond to multiple courses of antibiotic therapy. Histologic examination revealed a diffuse histiocytic infiltrate with associated lymphocytes, neutrophils, and pathognomonic Michaelis-Gutmann bodies. The patient also complained of progressive cough and was found to have concomitant bronchial malakoplakia, diagnosed after endobronchial biopsy of a nodule at the anastomotic site. Both areas were cultured and grew Escherichia coli, the organism most frequently implicated in malakoplakia. After surgical excision, the vulvar lesions did not recur. Malakoplakia is a diagnostic entity that the pathologist should consider in the differential diagnosis when encountered with histiocytic inflammation in the female genital tract. This entity can mimic gynecologic malignancies and can frequently persist and recur if not treated appropriately. In the gynecologic tract, it is a rare occurrence. We present an unusual case of combined malakoplakia involving vulva and lung.

Keywords: Malakoplakia; Vulva; Lung transplant

Case Presentation

A 67-year-old woman with a past medical history of interstitial lung disease, status post bilateral lung transplant, presented with persistent painful bilateral labial lesions, which first appeared three months post-transplant. The lesions were biopsied and initially diagnosed as an abscess. Additional lesions developed and cultures grew Escherichia coli; however, despite treatment with multiple antibiotics and tapering of immunosuppressive agents, the lesions never resolved. A subsequent pelvic exam revealed three to five erythematous papules on bilateral labia measuring up to 4 mm in greatest dimension without spontaneous drainage (Figure 1). The patient underwent complete excision of all vulvar lesions and the specimens were received in surgical pathology.

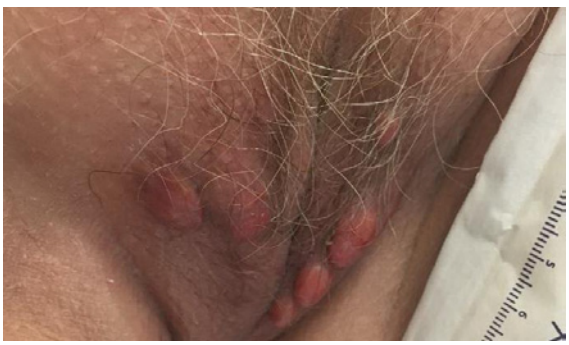

Figure 1: Multiple erythematous, firm, bilateral labial nodules.

Microscopic examination revealed a nodular proliferation of sheets of histiocytes occupying the dermis with admixed lymphocytes and neutrophils, underlying an unremarkable epidermis (Figures 2 & 3). Numerous targetoid, basophilic Michaelis-Gutmann bodies were conspicuously seen within the proliferation, highlighted by von Kossa and periodic-acid Schiff stains (Figure 4). Special stains for infectious organisms were negative. A diagnosis of malakoplakia of the vulva was made.

Interestingly, approximately one month prior to the excision of the vulvar lesions, the patient complained of an increasing cough and a bronchoscopy revealed a nodule present in the left mainstem bronchus at the anastomotic site (Figure 5). A biopsy of the lesion at the bronchial anastomotic site was diagnostic of malakoplakia. A recent bronchoalveolar lavage prior to bronchoscopy was positive for E. coli. The patient was treated with a ten-day course of amoxicillin-clavulanate.

Urine cultures also grew E. coli and the patient was treated with two weeks of intravenous ceftriaxone after excision of the vulvar lesions. No additional lesions have developed.

We present an unusual case of simultaneous vulvar and pulmonary malakoplakia following bilateral lung transplantation.

Discussion

Malakoplakia is a rare, benign histiocytic disorder that

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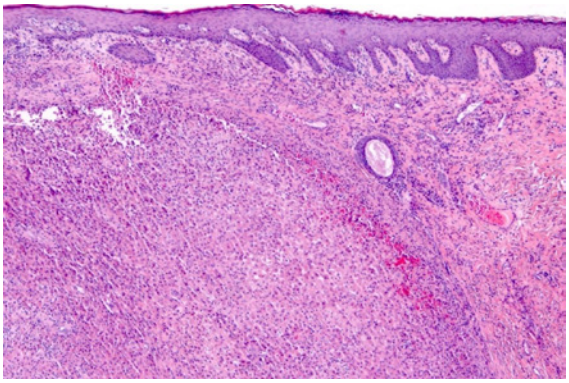

Figure 2: Nodular proliferation of granular histiocytes within the dermis underlying a relatively unremarkable epidermis (H&E, 5x).

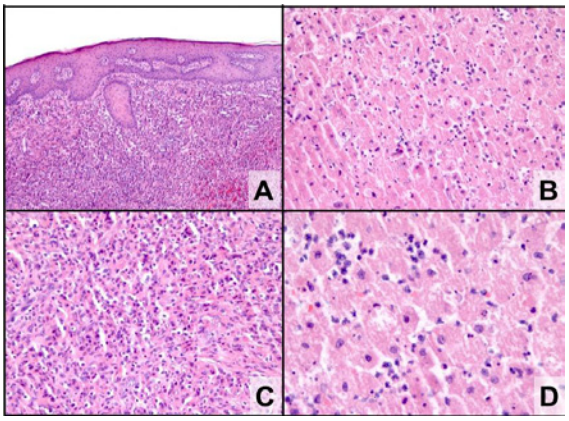

Figure 3: Malakoplakia of the vulvar dermis underlying unremarkable epidermis (A) (H&E, 5x). Granular histiocytes compose most of the cellularity (B) (H&E, 20x). Numerous Michaelis-Gutmann bodies are identified within the histiocytic inflammation (C) (H&E, 40x). Acute inflammation is also observed admixed with the histiocytes (H&E, 40x).

predominantly affects the urinary tract. Less than 500 cases have been reported in the literature [1]. It is well known that malakoplakia more commonly affects immunocompromised patients, specifically solid organ transplant recipients [1]. It was initially described in 1901 by von Hansemann and is also known as von Hansemann's disease [2]. Malakoplakia is a Greek-derived term and stands for "soft" (malakos) "plaque" (plakos) [2]. Histologically, malakoplakia is characterized by a histiocytic infiltrate with pathognomonic Michaelis-Gutmann bodies: 5-10 μm, targetoid, concentric, basophilic intracellular and extracellular structures. The Michaelis-Gutmann bodies are characteristically positive for von Kossa's calcium stain, Periodic Acid Schiff (PAS) stain, and iron stain [2].

The pathogenesis of malakoplakia is not well understood; however, recent theories suggest that the disease is an outcome of a bactericidal defect within the histiocyte lysosomes. The defective lysosomes have ineffective microtubule assembly due to a lack of Cyclic Guanosine Monophosphate (cGMP), which prevents the normal degranulation of lysosomes, an essential step in killing bacteria [2]. Lamellar deposition of organic and inorganic materials, such as calcium, iron, and phosphorus, form the classic Michaelis-Gutmann bodies [1,3]. The most commonly implicated organisms are Escherichia coli,

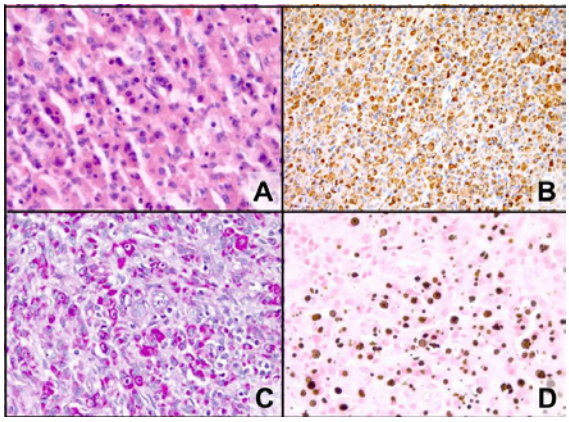

Figure 4: High power view of the inflammatory proliferation where numerous targetoid Michaelis-Gutmann bodies are identified (A) (H&E, 40x). CD68 stain highlighting abundant histiocytes (B) (CD68, 20x). Periodic acid-Schiff (C) and Von Kossa calcium stains (D) highlight the Michaelis-Gutmann bodies (PAS and Von Kossa, 40x).

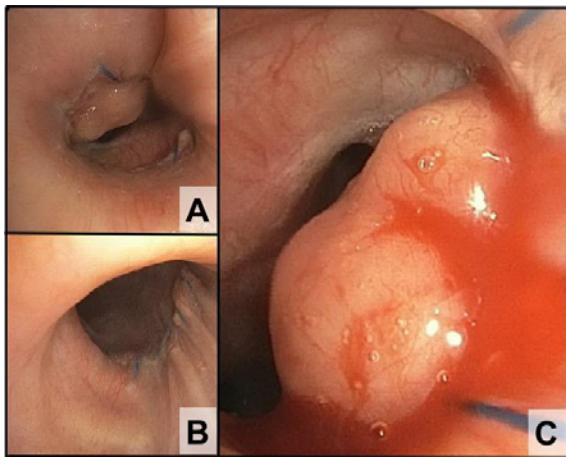

Figure 5: Bronchoscopy demonstrating nodules present at the anastomotic site of the left mainstem bronchus (A and C). Right mainstem bronchus with unremarkable anastomotic site (B).

Staphylococcus aureus, Pseudomonas aeruginosa, Mycobacterium tuberculosis, Proteus sp., and Rhodococcus equi [1,4]. Of these, Escherichia coli is by far the most common causative organism [3]. E. coli is also the most frequently identified bacterial culprit of urinary tract infections [5], which could explain why malakoplakia is most commonly seen in the urinary tract.

Malakoplakia has been described in numerous organ systems in addition to the urinary tract, including the gastrointestinal tract, skin, lungs, central nervous system, and bone, among others [1]. These lesions have been described less commonly in the female genital tract, including the uterus, cervix, vagina, and, rarely, the vulva [6] (Table 1). Women with genital tract malakoplakia are most commonly post-menopausal with an average age of 60 years [7]. Of all of the female genital tract organs, the vagina is the most common site involved and vaginal bleeding is usually the presenting symptom [7,8]. Vulvar malakoplakia has only been previously reported four times in the literature [6,9-11]. We present an unusual case of simultaneous vulvar and pulmonary malakoplakia following bilateral lung transplantation.

Table 1: Reported cases of malakoplakia affecting the female genital tract.

Case	Reference	Year	Age	Location	Symptoms/History
1	Rao [13]	1969	40	Broad ligament, inguinal area, endometrium	Vaginal bleeding
2	Van der Walt, et al [14]	1973	65	Vagina	Vaginal bleeding
3	Scheiner, et al [8]	1975	29	Fallopian tube	--
4	Arul and Emmerson [11]	1977	75	Vulva	Ulceration; Rheumatoid arthritis
5	Thomas, et al [15]	1977	60	Endometrium	Vaginal bleeding; Rheumatoid arthritis; Sjogren's syndrome
6	Lin, et al [16]	1979	49	Vagina	Hepatitis
7	Cremer and Busanny-Caspari [17]	1979	56	Vagina	Vaginal bleeding
8	Chalvardjian, et al [7]	1980	61	Left ovary, fallopian tube	Vaginal bleeding
9	Chalvardjian, et al [7]	1980	64	Cervix, vagina	Vaginal bleeding
10	Chalvardjian, et al [7]	1980	84	Vagina	Vaginal bleeding; Urothelial carcinoma
11	Wahl [18]	1982	72	Cervix	Vaginal bleeding; Rheumatoid arthritis
12	Wahl [18]	1982	66	Cervix	Vaginal bleeding
13	Willen, et al [19]	1983	71	Uterus and cervix	Vaginal bleeding
14	Molnar and Polliak [20]	1983	69	Endometrium	Diabetes mellitus
15	Chen and Hendricks [8]	1985	83	Cervix, pelvis	Vaginal bleeding; Xanthogranulomatous pyelonephritis
16	Chadha, et al [21]	1985	72	Endometrium	Vaginal bleeding
17	Falcon-Escobedo, et al [22]	1986	--	Cervix	--
18	Paquin, et al [23]	1986	49	Bartholin's gland	Asymptomatic
19	Paquin, et al [23]	1986	61	Bartholin's gland	Tender swelling labium minus; Parkinson's disease
20	Klempner,et al [24]	1987	50	Ovary (left adnexa)	Neurofibromatosis type I
21	Kawai, et al [25]	1988	88	Endometrium	--
22	Stewart and Thomas [26]	1991	--	Cervix and endometrium	Vaginal bleeding; Rheumatoid disease
23	Bessim, et al [10]	1991	44	**Vulva**, vagina	Fever, vaginal bleeding; Alcohol abuse
24	Koetsawang, et al [27]	1992	67	Endometrium, omentum	Vaginal bleeding
25	Fishman, et al [28]	1993	84	Vaginal cuff	Remote history cervix squamous cell carcinoma
26	Shaikh, et al [29]	1994	28	Uterus	Severe lower abdominal pain, fever
27	Baez-Giangreco, et al [30]	1994	50	Uterus, cervix, vagina	Vaginal bleeding
28	Kogulan, et al [6]	2001	29	**Vulva**, vagina	Vaginal bleeding and purulent cutaneous nodules; Ureterolithiasis
29	Chou, et al [31]	2002	47	Uterus, ovary, fallopian tube	Diabetes mellitus
30	Agnarsdottir, et al [9]	2004	69	Cervix	Uterine prolapse
31	Agnarsdottir, et al [9]	2004	49	**Vulva**	Vulvar lesion
32	Ramdial, et al [32]	2008	36	Cervix	Acquired Immunodeficiency Syndrome (AIDS)
33	Ramdial, et al [32]	2008	27	Cervix	Acquired Immunodeficiency Syndrome (AIDS)
34	Mirfazaelian, et al [33]	2015	57	Uterus	Vaginal bleeding
35	Jenkins and Reyes, current case	2017	67	**Vulva**, left main stem bronchus	Painful vulvar lesions; Bilateral lung transplant

This entity is important for both clinicians and pathologists to recognize and consider in the differential diagnosis when a patient is suspected to have an infectious process, especially when this involves the urogenital tract. Michaelis-Gutmann bodies are not always identified, especially in the early or late stages of malakoplakia [12]. Malakoplakia should be included in the differential diagnosis when a post-menopausal woman presents with vaginal bleeding or lesions of the vagina, vulva, and cervix. Large tumor-like lesions can be mistaken for gynecologic malignancies of the cervix, uterine adnexa, vagina, and vulva. Thus, malakoplakia is important to recognize both clinically and histologically, as malakoplakia can be cured with surgical or medical therapy.

Conclusion

Malakoplakia is a rare histiocytic inflammatory disorder caused by defective degradation of bacteria by phagolysosomes. Malakoplakia involving the female genital tract, especially the vulva, is rare. We present a case of persistent, painful vulvar malakoplakia and simultaneous bronchial malakoplakia in an immunosuppressed patient after bilateral lung transplant. The vulvar lesions were

unresponsive to antibiotic therapy alone, and surgical excision was necessary. Malakoplakia is an important inflammatory condition to recognize because it can mimic gynecologic malignancies and is usually cured with excision or antibiotic therapy. Pathologists must also be highly suspicious of malakoplakia when confronted with a specimen composed of sheets of foamy histiocytes and an inflammatory background, even if Michaelis-Gutmann bodies are not identified. Correlation with cultures and clinical history is essential.

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