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CASE SERIES

Pigmented Extramammary Paget's Disease of the Axilla: Two Case Reports and a Literature Review

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Abstract: Extramammary Paget's disease is a rare intraepithelial adenocarcinoma that mainly occurs in areas rich in apocrine sweat glands, such as the vulva, perianal region, and, less commonly, in the axillary skin. Owing to the varied manifestations of extramammary Paget's disease, it is often misdiagnosed. Particularly, pigmented extramammary Paget's disease (PEMPD) can be mistaken for superficial spreading melanoma, Bowen's disease, pigmented superficial basal cell carcinoma, or seborrheic keratosis. Here, we report two cases of women with axillary pigmented plaques who were confirmed as PEMPD through histopathological and immunohistochemical analyses. One of these patients was only 31 years old, which makes her the youngest reported case of PEMPD to date. All patients underwent Mohs micrographic surgery, and no recurrence or metastasis was observed postoperatively. PEMPD is a rare clinical variant that can resemble other pigmented lesions, both clinically and histopathologically. Therefore, this report underscores the importance of accurate differential diagnosis through immunohistochemistry.

Keywords: pigmented extramammary Paget disease, axilla, histopathology, immunohistochemistry, Mohs micrographic surgery

Introduction

Extramammary Paget's disease (EMPD) is a rare malignant tumor that typically occurs in areas rich in apocrine glands. Among the Asian population, male patients are more commonly affected by EMPD.[1,2] The most common sites of involvement are the scrotum and penis in men and the vulva in women; however, less commonly, other areas including the perianal, axillary, and umbilical regions may be involved. In some cases, the disease can be multifocal, involving more than one site. It often presents with infiltrative erythema, accompanied by scabs and scales, which sometimes resemble other skin conditions such as eczema. As the disease progresses, changes such as the development of sclerodermoid macules and lichenoid papules may occur.[3] Histologically, EMPD is characterized by the presence of Paget cells in the epidermis, which has a round, pale vacuolated cytoplasm and large pleomorphic nuclei. Hematoxylin–eosin staining revealed abundant, clear cytoplasm in these cells, which may appear eosinophilic. Paget cells can occur singly or in clusters. Tumor cells may exhibit pigmentation, or non-neoplastic dendritic melanocytes may colonize the affected epidermis.[4] Most cases of EMPD are in situ, typically demonstrating a slow progression. However, once Paget cells invade the dermis with an infiltration depth >1 mm, regional lymphatic metastasis and distant metastasis often occur,[5] which significantly impact patient survival and prognosis. Pigmented extramammary Paget's disease (PEMPD) is a rarer clinical variant characterized by a pigmented rash or plaque, often affecting only one side of the body.[6] The presence of pigment granules in the cytoplasm of tumor cells can lead to its misdiagnosis as melanoma. Immunohistochemical staining is thus essential for distinguishing Paget cells from melanoma cells in diverse epidermal proliferative lesions.

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Case Presentation

Case 1

A 47-year-old woman presented to our clinic with a black plaque in her left axilla that had been present for 5 years. Initially, three light brown small papules appeared in her left axilla without any obvious cause, and they did not cause any significant discomfort. Therefore, she did not seek medical attention. However, the rash gradually enlarged and merged, darkening in color. The patient developed mild pruritus three months prior, which persisted despite self-administered topical miconazole nitrate (2% cream) application for one week, prompting subsequent dermatological evaluation at our institution. Upon physical examination, a deep brown plaque approximately 1.5×1.3 cm in size was detected in the left axilla. The plaque showed an uneven surface, uneven color, and fairly distinct boundaries with slightly raised edges (Figure 1A).

Case 2

A 31-year-old woman presented to our clinic with a plaque in her right axilla that had been present for the past 2 years. Initially, the patient noticed a light-brown papule in her right axilla that developed without an obvious cause. She did not experience any particular discomfort and hence did not seek any medical attention, until later when the lesion gradually enlarged. Physical examination revealed a brown patch measuring 1.6×1.6 cm in the right axilla. The plaque displayed a rough surface with slightly darker edges and clear boundaries (Figure 1B).

Both the patients underwent detailed clinical examinations, showing no similar lesions elsewhere on the body. Breast and axillary lymph node palpation demonstrated no abnormalities. The patient did not have a personal or familial history of cancer. To confirm the diagnosis, we performed skin biopsies on both patients. Histopathological examination (Figure 2A) revealed typical features of Paget's disease, a disorganized intraepidermal proliferation of single and nested epithelioid cells with pagetoid migration, involving the full thickness of the suprabasal epidermis. The tumor cells exhibited conspicuous cytologic atypia, including abundant cytoplasm containing prominent fine brown melanin pigment and enlarged, irregular pleomorphic nuclei with occasionally conspicuous. Dermal invasion was not identified. The tumor cells exhibited positivity for CK7 and CK18 (Figure 2B and C), while being negative for S100, Melan-A, and CK20 (Figure 2D–F). This immunoprofile confirmed the diagnosis of PEMPD. Axillary and breast lymph node ultrasound scans, mammography, and chest computed tomography were conducted to exclude any associated underlying malignancy. After confirming that all skin lesions were primary, both the patients underwent Mohs micrographic surgery under local anesthesia and were followed up for >1 year. None of the cases showed any recurrence or postoperative complications.

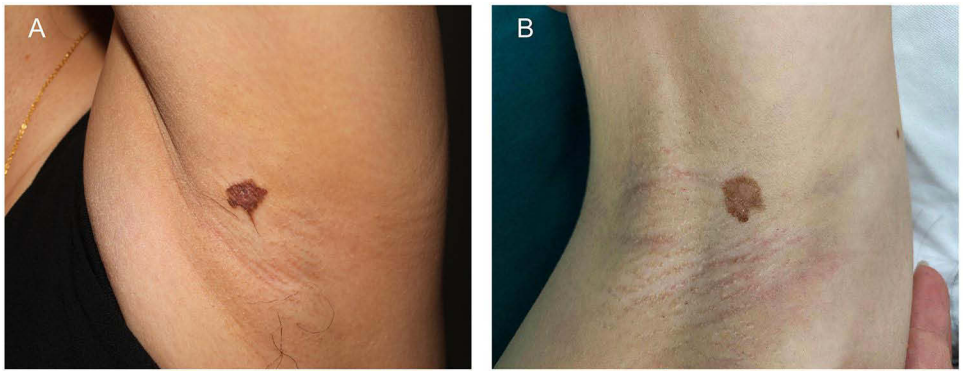

Figure 1 The skin lesions of 2 patients. (**A**) Dark brown plaque with slightly raised edges in the left axilla. (**B**) Brown plaque in the right axilla with slightly darker edges.

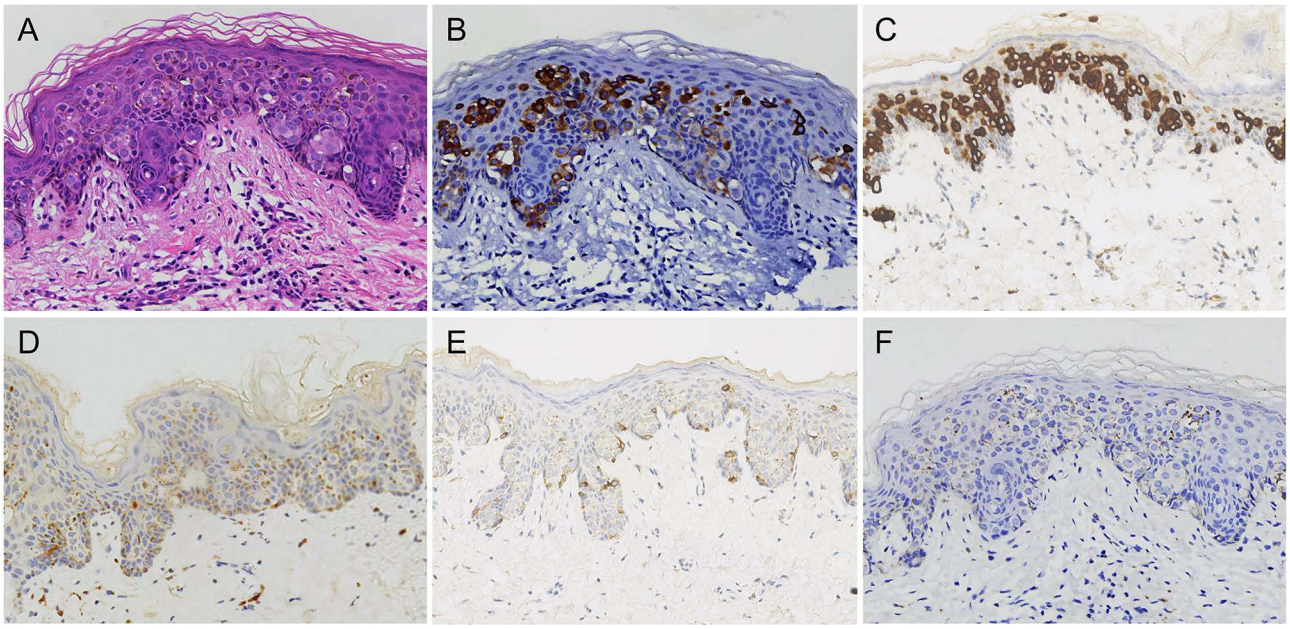

Figure 2 HE staining and immunohistochemical staining of Case 2. (**A**) Scattered and nested tumor cells are distributed in the spinous layer. The tumor cells were characterized by round, pale, vacuolated cytoplasm, and large pleomorphic nuclei. Pigment granules can be seen in the cytoplasm of some tumor cells but also in the cytoplasm of surrounding epidermal keratinocytes. The immunohistochemical staining of CK7 (**B**) and CK18 (**C**) was positive for neoplastic Paget cells in the epidermis. The tumor cells were negative for S100 (**D**), Melan-A (**E**), and CK20 (**F**) staining. The populations of non-neoplastic intraepidermal dendritic melanocytes were highlighted by melanocytic markers.

Discussion

PEMPD is a very rare clinical-pathological variant of EMPD, and understanding its disease features is crucial for obtaining accurate diagnosis. We reviewed articles published on PubMed from 2000 to 2024 using the keywords "Pigmented extramammary Paget's disease", and found 20 cases of PEMPD in total, out of which 4 cases were not included in the present analyses due to lack of sufficient diagnostic evidence (Table 1).[4,7–20] Among these cases, 75% (n=12) were female patients. The age of onset ranged from 43 to 74 years, with an average age of 59.33 ± 10.43 years. The most common site of occurrence in females was the vulvar region. Conversely, in male patients, the age of onset ranged from 65 to 83 years, with an average age of 75.75 ± 7.72 years, with the scrotum and axilla being the most common sites of lesions. Only five cases of PEMPD were recorded in the axillary region, which included three female patients. In our report, both the cases occurred in the axillary region and were of female patients. Among the cases with known disease duration, the longest is 30 years and the shortest is 4 months. The lesions mostly appear as uneven pigmented patches (dark-red, brown, or black) and are sometimes accompanied by erythematous scaling, which generally worsens throughout the disease. The most reliable immunohistochemical markers used to confirm Paget cells were CK7 (81.25%, n=13), CEA (43.75%, n=7), and CAM5.2 (37.50%, n=6). The most reliable markers used to rule out melanocytic tumors were HMB-45 (93.75%, n=15), S100 (68.75%, n=11), and Melan-A (43.75%, n=7). Of the 16 patients, four had underlying tumors, including colonic adenocarcinoma, prostatic adenocarcinoma, and breast cancer, with no significant correlation with PEMPD. None of the patients had recurrence or metastasis. Surgery was the chosen approach in cases where the treatment methods were provided.

Similar to typical EMPD, axillary EMPD usually presents with well-defined erythematous patches or plaques. Brownish to pigmented patches are seen in some cases as a subtype, termed pigmented EMPD.[20] Hypopigmentation was also observed in some patients. It may occur unilaterally, bilaterally, or simultaneously with the genital area. A recent retrospective study of 16 axillary EMPD cases revealed that unilateral lesions were more common, with a female predominance, while male patients tended to develop multiple lesions.[21] Compared to anogenital EMPD, axillary lesions are more frequently asymptomatic and thus prone to delayed diagnosis.[22] In axillary EMPD, the presence of an underlying carcinoma was reported in 35% (8/23) of cases.[10] In contrast, axillary PEMPD exhibits no significant differences in disease characteristics, typically presenting as

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Table 1 Pigmented Extramammary Paget Disease

Study	Sex/ Age	Duration (Y/M)	Clinical Appearance	Location	Positive Stains	Negative Stains	Underlying Carcinoma	Treatment	Initial Diagnosis
Chiba et al[7]	F/52	30Y	Brown-black plaque	Vulva	CAM5.2, CEA	S100, HMB-45	Breast carcinoma	/	BD
Chiba et al[7]	F/70	10Y	Scaly pigmented and erythematous lesion	Vulva	CAM5.2, CEA	S100, HMB-45	No	/	BD
Gumurdula et al[8]	F/63	/	/	Vulva/ perineal	CEA	HMB-45, MART-1	No	/	SSM
Petersson et al[9]	F/43	/	Erythematous and brown-black patch	Vulva	CK7	CEA, HMB-45, Melan-A, S100	No	/	/
Hilliard et al[10]	M/ 79	/	Pigmented plaque	Axilla	CK7, CAM5.2, EMA, CEA	CK903, CK20, S100, MART1, HMB-45, MITF	No	Operation	Melanoma
Vincent et al[4]	F/63	/	/	Abdomen	CK7	CK20, S100, MITF, Melan-A, HMB-45	Colonic adenocarcinoma	Operation	PEMPD
Lentini et al[11]	F/50	4M	Brown plaque with uneven color	Vulva/ perineal	CK7, PAS	HMB-45	No	Operation	/
Coras-Stepanek et al[12]	M/ 83	5-10Y	Irregular hyperpigmented plaque	Scrotum	CK7, CEA	S100, Melan-A, HMB-45, CK20	No	Operation	PEMPD
Wang et al[13]	M/ 76	/	Variegated brown plaque	Axilla	Alcian blue, PAS, CK7, CEA	CK20, S100, MelanA, HMB-45, HMWCK (34BE12)	Prostatic adenocarcinoma	Operation	SK
Ladak et al[14]	F/63	/	Scaly hyperpigmented plaque	Axilla	CAM5.2, EMA, CK7, HER2	P63, CEA, S100, Melan-A, HMB-45, ER, PR, CK20	No	Operation	Pigmented naevi
De la Garza Bravo et al[15]	F/51	6M	Irregular pigmented brown patch	Thigh	CK7, CK8/18, P63	CAM5.2, CK5/6, CEA, SOX-10, HMB-45, antityrosinase,	Breast carcinoma	Operation	PEMPD
Nagano et al[16]	F/46	5Y	Dark brown to black plaque with irregular border	Perineal	CK7, CEA, CAM5.2	S100, HMB-45	No	Operation	SSM
Wang et al[17]	M/ 65	3Y	Dark erythema, some areas are grayish brown	Scrotum	CK7, CerbB-2, EMA, CEA, PAS, CK20, Ki-67 (40%+)	K5/6, ER, PR, P16, P53, Melan-A, HMB-45, S100	No	Operation	Eczema
Kiavash et al[18]	F/74	8Y	Scaly erythema and pigmented plaque	Abdomen	Ber-Ep4, CAM5.2, CK7, GCDFP-15	CEA, CK20, S100, MART-1, HMB-45, MITF, antityrosinase	No	/	SK
Chuchvara et[19]	F/67	5Y	Multi-colored patch (dark brown, light brown, and white)	Axilla	CK7, CK8/18	Melan-A, SOX-10	No	/	Melanoma
Zhang et al[20]	F/70	4Y	Pink-to-brown plaque	Axilla	EMA, CK7	HMB-45	No	Operation	PEMPD

Abbreviations: CEA, carcinoembryonic antigen; CK, cytokeratin; EMA, epithelial membrane antigen; ER, estrogen receptor; GCDFP, gross cystic disease fluid protein-15; HMB-45, human melanoma black-45; HMWCK, high molecular weight cytokeratin; MART-1, melanoma antigen recognized by T cells 1; MiTF, microphthalmia-associated transcription factor; PAS, periodic acid-Schiff; PR, progesterone receptor.

solitary pigmented lesions with a similar female predominance. None of the cases showed regional lymphadenopathy or metastasis, and approximately 25% (4/16) were associated with concomitant malignancies, suggesting an overall favorable prognosis. However, unlike axillary EMPD, no cases of bilateral axillary involvement or concurrent genital lesions were observed in PEMPD patients (It should be noted that this retrospective analysis may be subject to reporting bias, as some EMPD cases with pigmented features might not have been classified as PEMPD in the literature. However, further analysis was precluded by insufficient clinical data).

The differential diagnosis for PEMPD includes superficial spreading melanoma (SSM) and pagetoid Bowen's disease (PBD). EMPD tumor cells are typically dispersed throughout the epidermis, while melanoma cells may directly infiltrate from the dermo-epidermal junction into the papillary dermis or scatter within the stratum spinosum. However, when tumor cells infiltrate the dermis, distinguishing PEMPD from SSM is challenging with conventional hematoxylin–eosin staining— particularly when melanin-laden Paget cells coexist with reactively proliferated dendritic melanocytes interspersed among the tumor cells. Therefore, selecting appropriate antibody markers for IHC is crucial. In PEMPD, the neoplastic Paget cells typically demonstrate positive expression for CK7, CK8/18, CEA, CAM5.2, EMA, and GCDFP-15, while being negative for S100, Melan-A, HMB-45, and MITF. Concurrently, melanocytic markers (eg, S100, Melan-A) may highlight interspersed and surrounding reactive and colonized melanocytes associated with the neoplastic proliferations. The histologic criteria that favor PBD over EMPD include the following: detection of dyskeratotic cells, identifiable intercellular bridges, and absence of atypical cells in the stratum corneum. PBD presents immunohistochemical characteristics of squamous cells, usually positive for CK5/6 and p63. However, some cases may also express CK7 and CAM5.2, leading to a potential diagnostic pitfall for EMPD. Therefore, combined detection with GCDFP-15 and PAS staining may be required for differential diagnosis when necessary (Table 2).

CK20 or CDX2 positivity may suggest secondary EMPD; however, primary EMPD in the perianal region can also exhibit CK20/CDX2 expression. Thus, the diagnostic utility of these markers must be interpreted in conjunction with anatomical location, and endoscopy or imaging may be required to exclude underlying internal malignancies. Recent studies have identified TRPS1 as a valuable marker for differentiating MPD/EMPD from histopathological mimics such as squamous cell carcinoma in situ (SCCIS) and melanoma in situ (MIS).[23,24] While MPD and primary EMPD at non-perianal sites consistently express TRPS1, secondary EMPD and MIS uniformly lack TRPS1 expression. Notably, 92% (12/13) of SCCIS cases exhibit TRPS1 positivity, whereas 91% (10/11) of perianal primary EMPD cases are TRPS1-negative. These findings underscore the importance of avoiding overreliance on single immunohistochemical markers in diagnosing pagetoid intraepidermal neoplasms. Regardless of anatomical location, accurate diagnosis requires comprehensive correlation of histomorphological features, multi-marker immunohistochemical profiles, and clinicopathological context.

Dermatoscopy and reflectance confocal microscopy (RCM) serve as valuable adjunctive tools for the diagnosis and differential diagnosis of PEMPD. Dermatoscopic examination reveals regression-like area, irregular dark globules, irregular brown/black dots, gray-blue structures and structureless areas, and white negative pigment network. RCM imaging demonstrates a disarranged honeycomb pattern in the epidermis, with pagetoid spread of hyporeflective large round nucleated cells and hyper-reflective atypical dendritic cells. As the distribution of melanin in PEMPD is similar to that in melanoma, skin imaging cannot exclude melanoma, and immunohistochemical diagnosis is still needed.

Table 2 Immunohistochemical Markers and Special Stains of PBD, SSM, and PEMPD

Disease	Positive Stains	Negative Stains
Pagetoid Bowen's disease (PBD)	CK5/6, HMWCK, P63	CAM5.2 (+/−), CK7 (+/−), GCDFP-15, HMB45, Melan-A, PAS, S-100
Superficial spreading melanoma (SSM)	HMB45, Melan-A, S-100	CK7, CEA, EMA, GCDFP-15, P63
Pigmented extramammary Paget's disease (PEMPD)	CK7, CK8/18, CEA, CAM5.2, EMA, GCDFP-15, PAS	CK5/6, HMB45, HMWCK, Melan-A, S-100, P63 (+/−)

Abbreviations: CEA, carcinoembryonic antigen; CK, cytokeratin; EMA, epithelial membrane antigen; GCDFP-15, gross cystic disease fluid protein-15; HMB-45, human melanoma black-45; HMWCK, high molecular weight cytokeratin; PAS, periodic acid-Schiff.

Currently, three possible mechanisms for pigment production in PEMPD have been proposed: (1) proliferation of reactive melanocytes secondary to tumor growth factor production; (2) transfer of excess melanin from proliferative dendritic melanocytes to neoplastic Paget cells; and (3) an increase in dermal melanophages. However, the molecular-level mechanism remains to be studied. In our case, Paget cells contained abundant intracytoplasmic melanin granules (primary contributor), with scattered dendritic melanocytes interspersed among tumor cells (secondary factor). We hypothesize that this dual mechanism collectively underlies the pigmented clinical appearance of the lesion.

The prognosis of EMPD depends on several factors, among which tumor invasion depth (> 1 mm) is the most critical. Other significant determinants include lymph node metastasis and underlying internal malignancies.[25] It should be emphasized that when tumor cells are confined to intraepithelial extension along hair follicles or sweat gland ducts without breaching the basement membrane, the lesion remains categorized as an intraepidermal process and does not qualify as invasive disease. The treatment options for PEMPD are similar to those for EMPD and include surgery (eg, Mohs micrographic surgery), local immunomodulators (eg, imiquimod), radiotherapy, and photodynamic therapy. Regular physical examinations, including lymph node examination, are recommended every 3–6 months for 3 years and then every 6–12 months for at least 5 years after diagnosis. Suspicious lesions should undergo histopathological or skin imaging examinations, with imaging of local lymph nodes if necessary. For patients with secondary or metastatic PEMPD, regular monitoring and examinations are necessary according to the specific situation.[26]

In this report, we have described two cases of axillary PEMPD, which is an extremely rare occurrence. One of the patients is only 31 years old, indicating that PEMPD can affect young and middle-aged patients. Thus, when encountering pigmented plaques under the axilla in young and middle-aged patients, PEMPD should be considered in the differential diagnosis. Through a literature review, we found that women are more likely to develop PEMPD, with an earlier age of onset than men. Lesions are more common in the vulva, while axillary involvement is relatively rare. Moreover, IHC plays a crucial role in the diagnosis of PEMPD.

Data Sharing Statement

The original contributions presented in the study are included in the article. Further inquiries can be directed to the corresponding author Lixiong Gu.

Ethics Approval and Consent to Participate

Written informed consent for publication of this case report including photography and medical data was obtained and signed by the patient. Institutional ethical approval was not required to publish this case report.

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Disclosure

The authors declare no conflicts of interest regarding the publication of this paper.

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