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DISORDERS OF PIGMENTATION

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Introduction

Almost all pigmentary conditions involving the extra-genital skin and mucosa can affect the vulva and perineum. These often cause stigmatization, social isolation and ostracization. These psychosexual implications hamper interpersonal relations. Vulvar pigmentation is ultraviolet light independent. Post-inflammatory hyperpigmentation is the karma of skin types 4–6, especially on frictional areas such as the axillae, vulva, groins and inner thighs. This is especially common in Asians, Hispanics and African-Americans. Also, inflammatory processes stimulate the activity of the melanocytes to increase the production and distribution of eumelanin. Increase in dermal melanophages renders a bluish hue to the skin, the Tyndall effect. This is seen as intense pigmentation in certain vulvar inflammatory conditions, which may be mistaken as melanomas. In contrast, the vaginal mucosa generally has no melanocytes, and any pigmented area needs to be investigated. Complete or partial loss of pigmentation, resulting in depigmentation or hypopigmentation, is more evident in the skin of color. Important causes of vulvar dyspigmentation are listed in Table 2.1.

TABLE 2.1: Causes of Dyspigmentation in the Vulvar Area

Hypo or Depigmentation in Vulvar Area	Hyperpigmentation in Vulvar Area
Post-inflammatory hypo or depigmentation	Physiologic hyperpigmentation
Vitiligo	Acanthosis nigricans
Lichen sclerosus	Vulvar melanosis
Vitiligoid LS	Post-inflammatory hyperpigmentation
Nevus depigmentosus	Fixed drug eruption
Contact depigmentation	Lentigines
Fordyce's Spots	Nevi
	VIN
	Melanoma
	Vascular lesions which appear deeply pigmented e.g. angiokeratoma, bruises

Hyperpigmentary disorders

Pigmentary vulvar lesions have been reported in 10–15% of the Caucasians population. This number is higher in darker skin types, but exact statistics are unknown. The lesions may be melanocytic or non-melanocytic.

Hyperpigmentation of the vulva often presents as a diagnostic dilemma. Physiological hyperpigmentation is usually symmetrical, asymptomatic and without any textural changes. It occurs in dark-skinned individuals more marked on the labia, introitus and upper inner thighs. Pregnancy, obesity and B12 deficiency enhance pigmentation.

Acanthosis nigricans (AN)

Skin under the influence of insulin excess gets thick, velvety and appears hyperpigmented. This may be seen in all folds including genitocrural folds, vulva and inner thighs (Figures 2.1 and 2.2). The face, neck, axillae and infra-mammary folds are the other sites involved. Vulvar AN maybe associated with obesity and diabetes mellitus. Dermoscopy of AN shows linear crista cutis and sulcus cutis, corresponding to the papillomatosis

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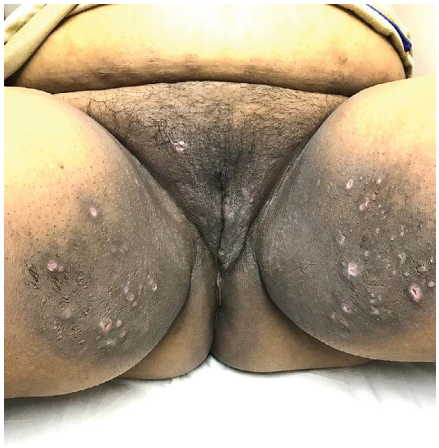

FIGURE 2.1 Thick velvety hyperpigmented patched of acanthosis nigricans on the vulva and inner thighs in a patient of HS.

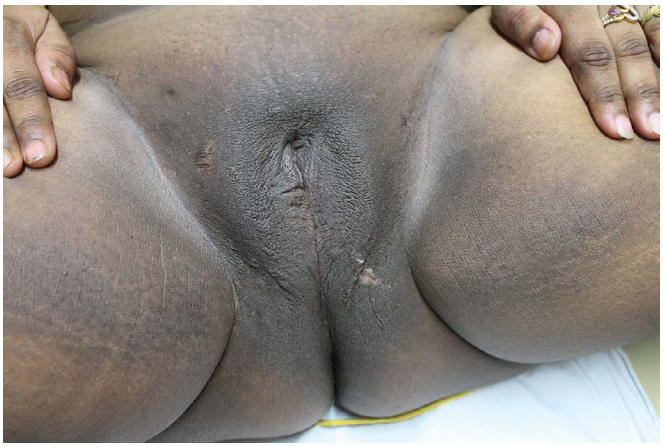

FIGURE 2.2 Selected velvety pigmented plaques of acanthosis nigricans involving the vulva and inner thighs. (Photographs by Dr Nina Madnani.)

and interspersed sulci, with well-defined black or brown dots of pigmentation. Insulin-sensitizing drugs over several months, together with significant weight loss, may bring about a reduction of acanthosis. AN is commonly associated with polycystic ovarian syndrome (PCOS).

Recommended reading

1. Grasinger CC. Vulvar acanthosis nigricans: a marker for insulin resistance in hirsute women. Fertil Steril (1993). PMID: 8458461.

Vulvar melanosis

Melanotic macules are a benign condition commonly seen on the non-keratinized mucosa and sometimes on the keratinized skin of the vulva, in women of reproductive age with a median age of 40–44 years. Also known as idiopathic lenticular mucocutaneous pigmentation. This is usually asymptomatic, but picked up during a self-examination or during a routine gynecological check-up (Figure 2.3). The pigmented area involved is more than 4 mm in diameter, is usually dark brown and occurs mainly on non-keratinized mucosae of the lower vagina, vestibule, perineum, labia majora or minora as single or multiple patches with irregular borders. The lesions are flush with the surrounding skin. Brownish reticular, parallel or honeycomb pattern may be seen on dermoscopy. Histopathology shows basal cell hyperpigmentation, with no increase in melanocyte numbers or epidermal hypermelanosis (Figure 2.4). Malignancy (melanoma and pigmented vulvar intraepithelial neoplasia (VIN)) is the most important differential diagnosis in vulvar melanosis. An adequate and prompt biopsy is required for a correct diagnosis.

Counselling and reassurance about the benign nature of the condition is necessary.

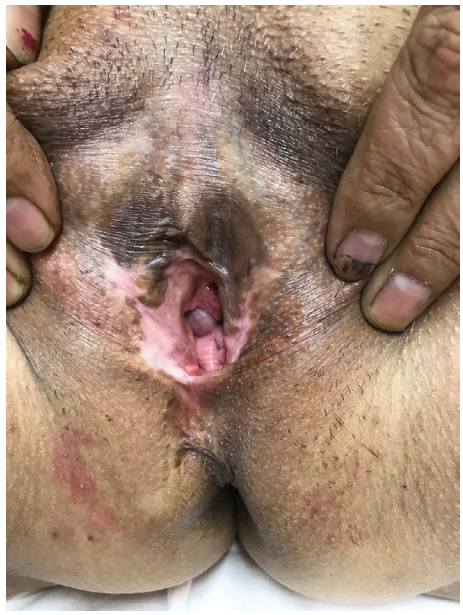

FIGURE 2.3 Asymptomatic patches of vulvar melanosis on either side of the clitoris. (Photograph by Dr Nina Madnani.)

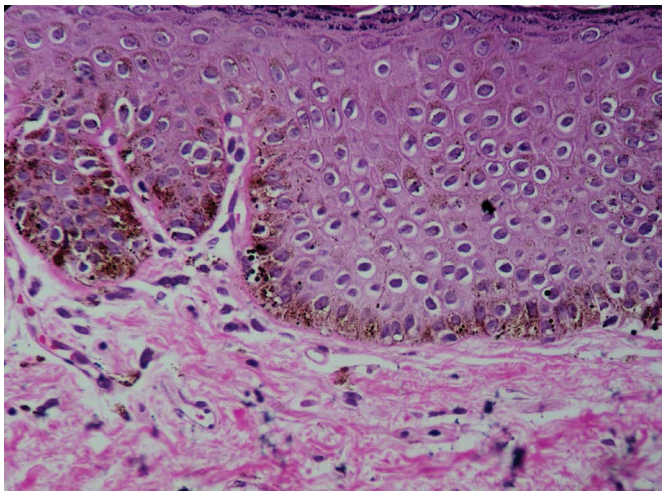

FIGURE 2.4 H & E, 400×, prominent melanin in basal layer with pigmented dendrites in spinous layer. (Photograph by Dr Rajiv Joshi.)

Recommended reading

1. De Giorgi V, et al. JAMA Dermatol (2020). PMID: 32785609.

Post-inflammatory hyperpigmentation

In skin types 4–6, the tendency to intense post-inflammatory hyperpigmentation lasting over months to years is a common phenomenon. Most injuries or dermatoses can elicit this (Figure 2.5). Detailed history and tell-tale signs of inflammation may give clues to the primary inciting condition. Controlling the inciting agents and use of topical medications containing kojic acid, arbutin and botanicals are helpful. Often, the pigmentation fades off over weeks/months/years, once the inflammatory process has settled. Post-inflammatory pigmentation of dermatomycosis is a classic example of this phenomenon (Figures 2.6 and 2.7).

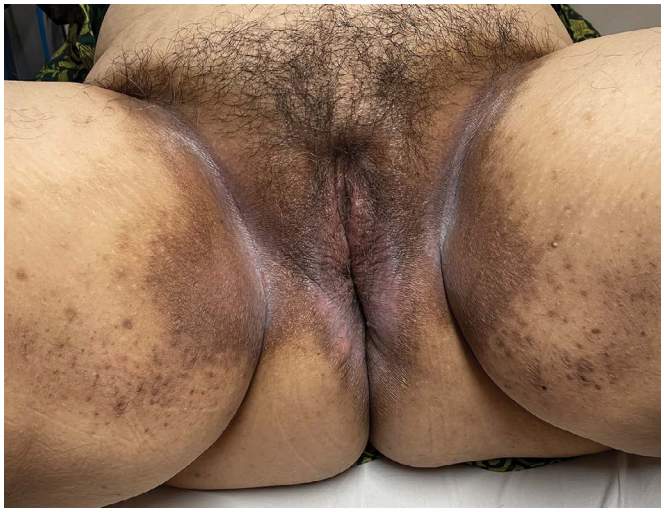

FIGURE 2.5 Many inflammatory disorders can result in hyperpigmentation upon resolution. (Photograph by Dr Nina Madnani.)

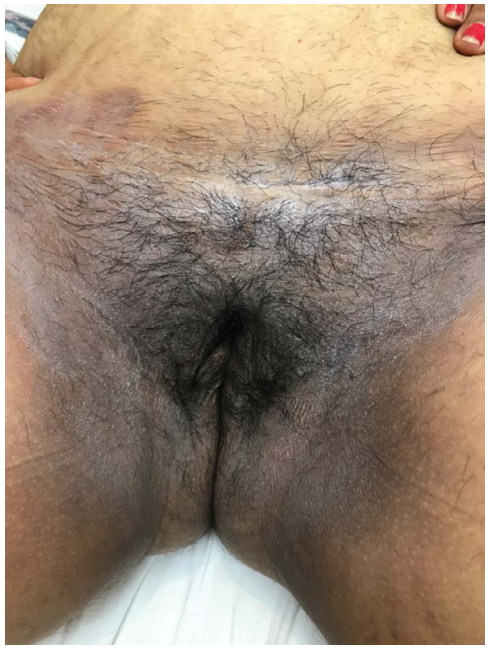

FIGURE 2.6 Deep pigmentation in areas of past dermatophyte infection. (Photograph by Dr Nina Madnani.)

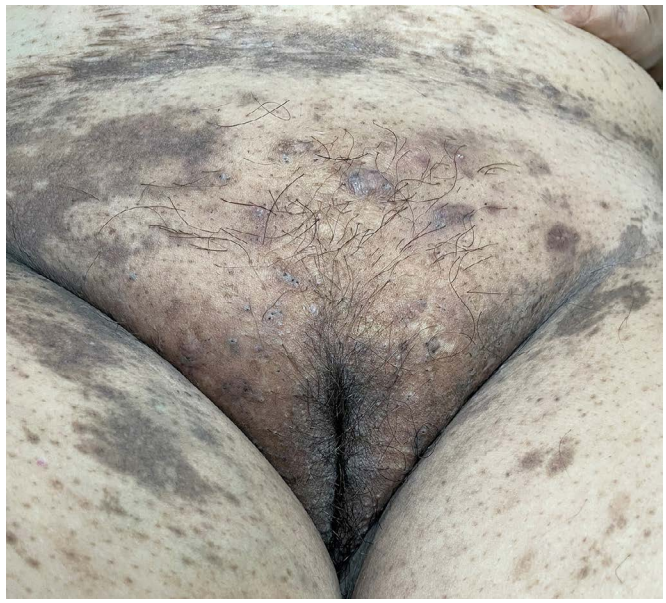

FIGURE 2.7 Grayish black pigment with follicular involvement in a case of lichen planus pigmentosus. (Photograph by Dr Nina Madnani.)

Fixed drug eruption

This is a common, distinct cutaneous allergic reaction resulting from exposure to some medication and characteristically recurring at the same sites on re-exposure.

On keratinized skin, it presents as a single, well-defined, round or oval dusky red patch which may later blister or ulcerate to heal with intense, brown-black pigmentation. Non-pigmented variants have been reported on the mucosa. Fixed drug eruption (FDE) reported on the vulva is usually bilaterally symmetrical and presents as an erosive vulvitis, unresponsive to treatment until the offending drug has been identified and stopped. It is observed most commonly on mucocutaneous junctions. Non-steroidal anti-inflammatory drugs (NSAIDS), anti-epileptics and anti-microbials are commonly implicated in this reaction. Others implicated are griseofulvin, fluconazole, ibuprofen, COX2-inhibitors and metronidazole. Once the culprit drug is discontinued, topical mid-potency corticosteroids are useful. In severe cases, systemic corticosteroids may be administered. The use of depigmenting agents and lasers may hasten the resolution of the post-reaction pigmentation.

Recommended reading

1. Fischer G. J Reprod Med (2007). PMID: 17393766.
2. Drummond C et al. Australas J Dermatol (2009). PMID: 19397565.
3. Abril-Pérez C, et al. Am J Obstet Gynecol (2022). PMID: 34610321.

Nevi

These are common in darker skin types and are often asymptomatic but cause considerable anxiety to the patients. They are uniformly colored, have a regular border and are small in size (Figure 2.8). These mainly present on the labia majora, minora and clitoral hood. Those on the non-keratinized surface need to be kept under observation for any changes.

Dermoscopic findings include globular and uniform reticular pigment pattern (Figure 2.9). A biopsy may be required in equivocal cases, wherein histopathology examination will give a definitive diagnosis (Figure 2.10).

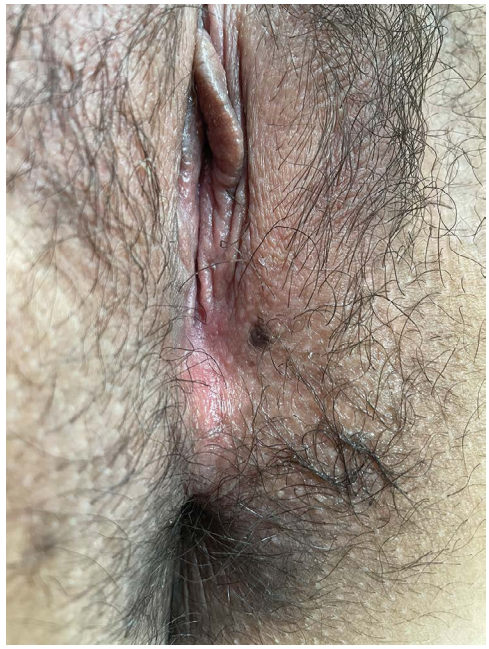

FIGURE 2.8 Well-defined, dark brown, flat papule. (Photograph by Dr Nina Madnani.)

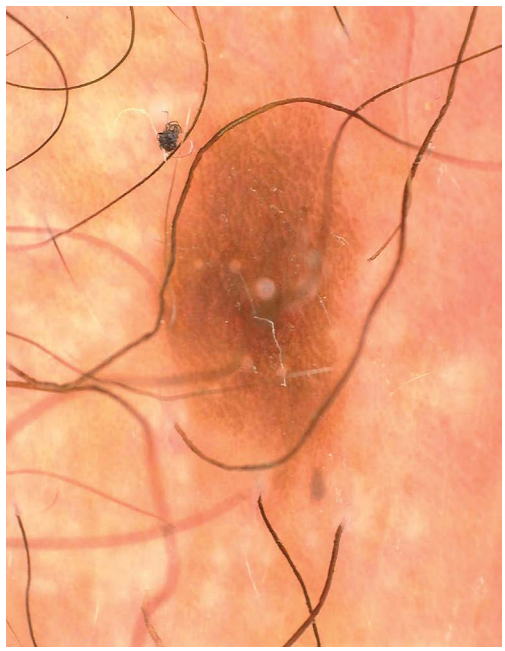

FIGURE 2.9 Dermoscopic image of the nevus. Dinolite Digital Microscope WF-20_polarized 10×. (Photograph by Dr Nina Madnani.)

Recommended reading

1. Allbritton JI. Obstet Gynecol Clin North Am (2017). PMID: 28778635.
2. Venkatesan A. Dermatol Clin (2010). PMID: 20883921.