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Review

Benign "lumps and bumps" of the vulva: A review

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ABSTRACT

Vulvar dermatology represents a challenge for many providers. Given that the vulva is both a gynecologic and dermatologic organ, patients with cutaneous lesions involving the vulva may present to primary care, gynecology, or dermatology. Particularly within dermatology, the vulva remains understudied, which can lead to anxiety among providers regarding appropriate next steps in the diagnosis and management of vulvar lesions. Thus, the purpose of this review is to highlight commonly encountered anatomic variants and benign neoplasms of the vulva, distinguish them from key pathologic mimickers, and provide guidance to practicing dermatologists on what may constitute normal vulvar variations.

Contents

Introduction

The vulva refers to the external female genitalia and includes the mons pubis, labia majora, labia minora, clitoris, vulvar vestibule, urethral meatus, vaginal introitus, and Bartholin's and Skene's vestibular glands (Nguyen and Duong, 2020). As both a gynecologic and dermatologic organ, the vulva lies well within the purview of the dermatologist's practice. That said, vulvar lesions can often elicit an elevated level of anxiety for patients and dermatologists alike—this is likely due to the broad range of etiologies of vulvar lesions, some of which carry significant risk of morbidity, but many of which are benign (Kelekçi et al., 2016). Dermatologists' ability to recognize benign variants of the vulva is critical, not only to reduce patient anxiety, but also to minimize unnecessary workup.

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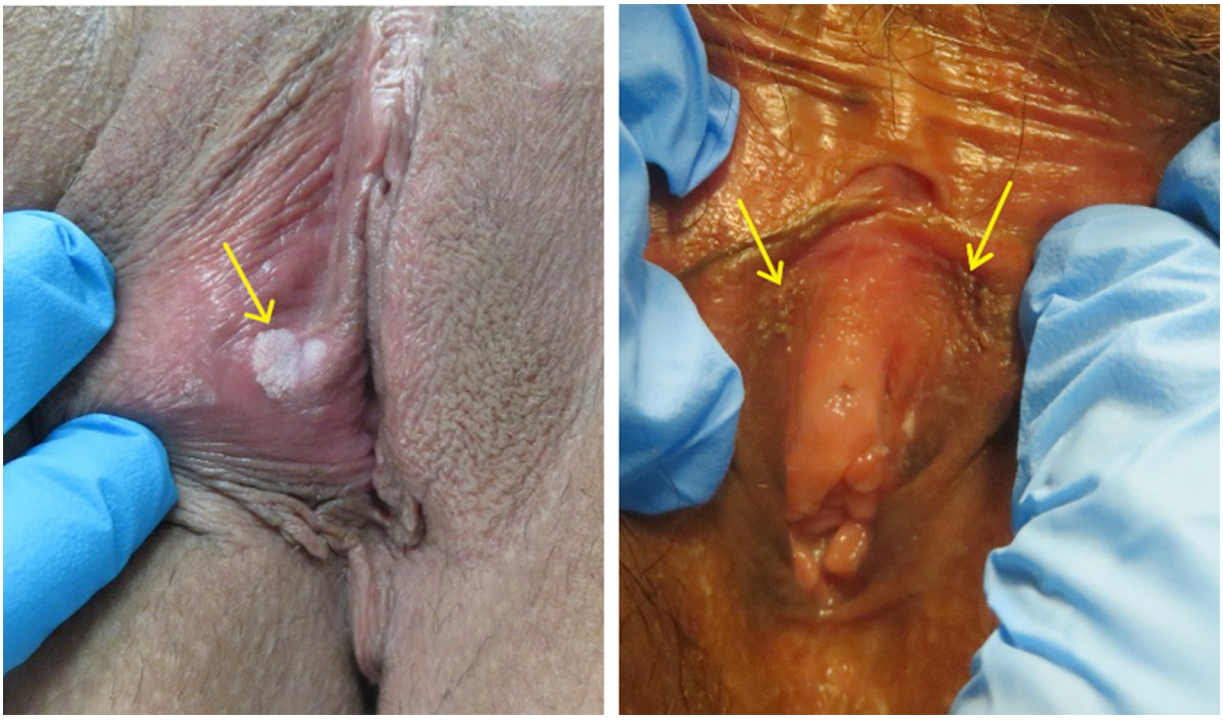

Fig. 1. Condyloma acuminatum (left) versus vestibular papillomatosis (right). Vestibular papillomatosis (right) is symmetrical and has soft, finger-like projections, each with its own insertion point. Condylomas (left) are firmer, with fused bases, and are asymmetrically distributed.

Unfortunately, the current literature on benign neoplasms and anatomic variants of the vulva is both limited in scope and principally found in gynecologic journals not widely read by dermatologists. Thus, the purpose of this review is to highlight some of the most commonly encountered anatomic variants and benign neoplasms of the vulva, distinguish them from key pathologic mimickers, and provide reassurance as to what may constitute normal vulvar variations. These entities include vestibular papillomatosis (VP), variation in vulvar dimension, urethral caruncle (UC), seborrheic keratosis (SK), syringoma, epidermal inclusion cyst (EIC), vestibular gland cyst (VGC), and hidradenoma papilliferum (HP).

Anatomic variants

Vestibular papillomatosis

VP was first described in 1982 by Altmeyer et al. (1981) as "pseudocondylomata of the vulva" and has since been reported under a variety of names, including hirsutoid papillomas of the vulva, vulvar squamous papillomatosis, micropapillomatosis labialis, and squamous vestibular micropapilloma (Altmeyer et al., 1981; Sarifakioglu et al., 2006). The exact prevalence of VP remains unknown, but current estimates suggest that it ranges from 1% to 33% of the population (Welch et al., 1993). Importantly, despite the suggestion of its monikers and its clinical resemblance to human papillomavirus (HPV)-associated condyloma acuminata, VP is considered a benign anatomic variant of the vulva (Fig. 1; Moyal-Barracco et al., 1990).

Clinically, VP is characterized by the presence of multiple frond-like mucosal papillae in a linear and symmetric distribution within the vulvar vestibule and inner labia minora. The presence of grouped papules in the vulva invokes HPV-associated condylomas as part of the differential, but VP can be distinguished from condylomas by two key features. First, whereas the filiform projections of condylomas tend to fuse at their base, the bases of individual projections of VP remain separate and distinct. Second, whereas condylomas tend to be firmer, randomly distributed, and not confined to the vulvar vestibule, VP tends to be softer and symmetrically distributed within the confines of the inner labia minora and vestibule (Muhammed et al., 2019).

Although clinical appearance and workup alone are usually sufficient to make a diagnosis of VP, should histopathological analysis be pursued, prominent fibrovascular cores covered by mature squamous epithelium are characteristic of VP. Epithelial cells often demonstrate clear vacuoles containing glycogen, rather than the true koilocytic changes that are seen in condylomas (Sarifakioglu et al., 2006). Importantly, because VP represents a normal physiologic variant, it does not require treatment beyond patient education and reassurance.

Variations in vulvar dimensions

Although not a specific clinical entity, variation in vulvar dimension and appearance can be challenging for dermatologists to contextualize, especially when lacking a robust background in performing vulvar examinations. Moreover, despite representations of female nudity being a common feature in popular culture, surprisingly few depictions of normal female genitalia and vulvar anatomy exist. More recently, a limited number of observational, cross-sectional studies within the gynecologic literature have sought to better characterize "normal" vulvar variations. Lloyd et al. (2005) described the genital dimensions of 50 healthy premenopausal women and found wide ranges in clitoral size (0.5–3.5 cm), labial length (7.0–12.0 cm for the labia majora, 2.0–10.0 cm for the labia minora), and rugosity (ranging from smooth to marked). Other studies in both pre- and postmenopausal women have demonstrated a similarly broad range of normal vulvar dimensions (Verkauf et al., 1992; Weber et al., 1995).

From the dermatologist's perspective, it is important not only to appreciate these normal variations in vulvar dimensions, but also to contrast them with physical signs of scarring or erosive vulvar pathology. For example, both lichen sclerosus and erosive

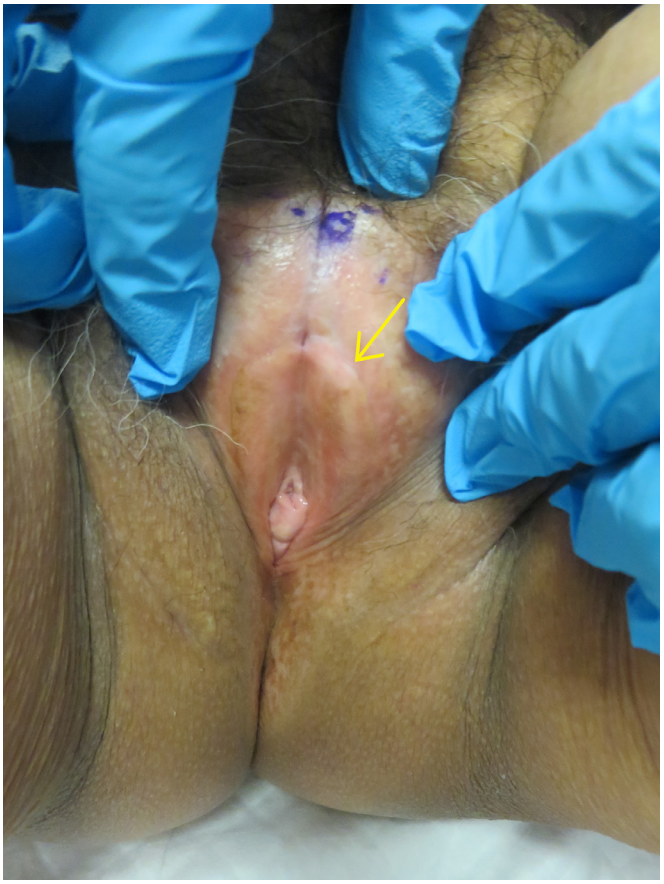

Fig. 2. Scarring from lichen sclerosus causing fusion of the labia minora and labia majora.

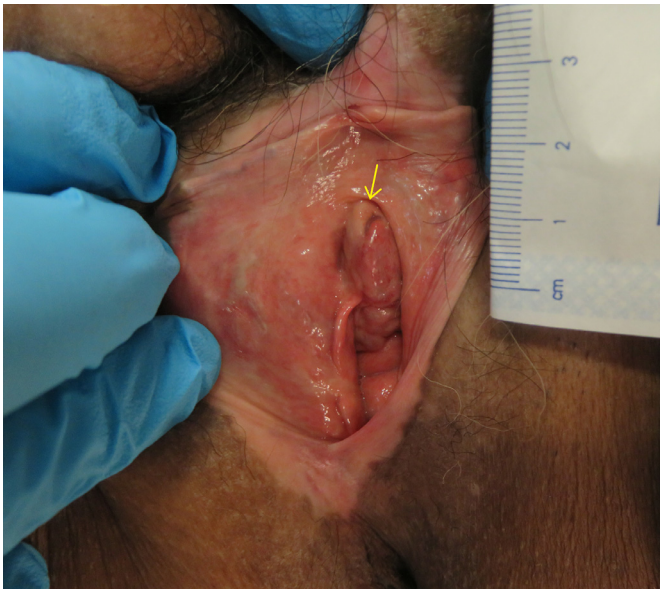

Fig. 3. Urethral caruncle arising from the urethral meatus; notably, this patient has concomitant lichen sclerosus.

lichen planus are inflammatory lichenoid dermatoses of the vulva that, when left untreated, can lead to irreversible scarring, sexual dysfunction, and, in extreme cases, interference with urination (Fruchter et al., 2017). Although these dermatoses typically manifest with overt symptomatology (most commonly, pruritus or pain), in some instances early anatomic changes are the only clue that a scarring process is underway. Notably, although vulvovaginal atrophy in the setting of age-related hypoestrogenism may manifest with shrinkage of the labia minora and vulvar vestibule, overt signs of inflammation (including the presence of erosions, dyspigmentation, fusion of the labia minora with the labia majora, fusion and loss of mobility of the clitoral hood, and/or scarring of the vaginal introitus) should prompt a thorough inquiry into vulvar symptomatology and possible biopsy to evaluate for an inflammatory vulvar process (Fig. 2; Fruchter et al., 2017; Schlosser and Mirowski, 2015).

Benign neoplasms

Urethral caruncle

UCs are the most common benign tumor of the female urethra (Chiba et al. 2015; Ferrier, 1926). Most often arising from the posterior urethral meatus, these exophytic, polypoid masses are typically seen in postmenopausal women, although isolated reports in prepubertal women exist (Burkland, 1952; Conces et al., 2012; Haley et al., 1998). UCs are typically solitary and small in size (Fig. 3), but some patients may present with multiple lesions up to 1 to 2 cm in diameter. Their marked variation in size and morphology can make clinical recognition of UCs quite challenging (Ferrier, 1926; Lobo et al., 2017).

Although their etiology remains incompletely understood, UCs are thought to arise in the setting of hypoestrogenemia and vaginal atrophy. Inflamed urethral mucosa everts and ultimately progresses to localized prolapse of the posterior urethral wall (Haley et al., 1998; Lobo et al., 2017). Growth of the caruncle is thought to occur secondary to chronic irritation and inflammation. Moreover, antiestrogen therapies (e.g., tamoxifen) have been found to accelerate UC formation (Bachmann and Nevadunsky, 2000).

UCs may be asymptomatic or can present with pain, dysuria, bleeding, or other irritative urinary symptoms (Burkland, 1952; Coban and Biyik 2014; Conces et al., 2012). Early excisional biopsy must be considered should features suspicious for malignancy (e.g., changing size, irregular consistency, or failure to respond to topical estrogen) be noted (Chiba et al., 2015; Tunitsky et al., 2012). Histopathologic examination typically reveals a proliferation of transitional and stratified squamous epithelium with vascular connective tissue and an inflammatory infiltrate of lymphocytes and plasma cells (Haley et al., 1998). Benign entities, including pyogenic granuloma, urethral prolapse, varicosities, or periurethral gland abscesses, are common UC mimickers (Table 1; Lobo et al., 2017). Urethral prolapse typically can be distinguished from UC because a prolapse tends to manifest as an annular, circumferential lesion resembling a donut (Lobo et al., 2017). Notably, malignant neoplasms, including urothelial carcinoma, squamous cell carcinoma, melanoma, lymphoma, and sarcomas, can also share features with UC and must be considered in the differential (Conces et al., 2012).

If symptomatic, management of UC can be divided into a conservative treatment approach versus surgical resection. Initial therapy includes topical estrogen or steroidal ointments. If the caruncle does not regress, grows further, or symptoms continue, excisional biopsy is indicated (Lobo et al., 2017).

Seborrheic keratosis

First described by Freudenthal in 1927, SKs are exceedingly common, benign, epidermal tumors whose prevalence increases with age (Kwon et al., 2003; Yeatman et al., 1997). Although a link between HPV and genital SKs has also been suggested, several re-

Table 1
Characterization of benign vulvar lesions

	Clinical appearance	Histopathologic features	Treatment
Cystic lesions			
Bartholin gland cyst	Small cystic swelling and/or palpable mass in the vulvar vestibule at 4 and 8 o'clock positions	Cyst lined by columnar squamous or flattened epithelium	For cysts that do not spontaneously resolve, catheterization or marsupialization can be considered
Canal of Nuck cyst	Painless, reducible, inguinal-labial swelling	Hydrocele lined with columnar, transitional, and stratified squamous epithelium	Refer to gynecology and/or general surgery for surgical excision and closure of persistent canal
Epidermal inclusion cyst	Firm, round, yellow-white papulonodules	Cyst lined with keratinizing stratified squamous epithelium with an intact granular layer filled with laminated keratin	Reassurance; incision and drainage if infected; intralesional steroids if inflamed; excision
Mesonephric cyst	Small cystic swelling and/or palpable mass on the lateral aspects of the vulva	Cyst lined by cuboidal cells	Reassurance; larger cysts may be excised
Milia	Waxy, firm, white to flesh-colored dome-shaped papules 1-3 mm in size	Homogeneous deposition of eosinophilic colloid in the papillary dermis	Reassurance; electrodessication or expression of keratin contents after incision can be pursued for cosmesis
Skene's duct cyst	Small cystic swelling and/or palpable mass adjacent to the urethral meatus, occasionally with drainage	Cyst lined by columnar, transitional, or squamous epithelium	For cysts that do not spontaneously resolve, marsupialization can be considered
Steatocystoma multiplex	Small, flesh-colored to yellowish cystic papules or nodules	Cyst lined by stratified squamous epithelium with sebaceous glands attached to cyst epithelium	Reassurance; incision and drainage if cosmetically unfavorable
Vestibular gland cyst	Translucent soft, smooth, and round cysts	Cyst lined by simple mucous-secreting columnar epithelium	Reassurance
Glandular neoplasms			
Hidradenoma papilliferum	Firm, smooth-surfaced, red, blue, or skin-colored nodule with well-defined capsule, 0.5–2.0 cm	Papillary and glandular hyperplasia	Excision
Poroma	Single, slow-growing, well-circumscribed, pink-to-red papule, nodule, or plaque	Proliferation of small, round, monotonous cuboidal cells (poroid cells)	Excision only indicated if malignancy suspected
Spiradenoma	Slow-growing, flesh-colored nodule, occasionally painful	Multilobulated basophilic dermal nodules surrounded by hyalinized collagen capsule	Excision only indicated if malignancy suspected
Syringocystadenoma Papilliferum	Firm, smooth-surfaced or verrucous red nodule or plaque	Papillary and glandular hyperplasia	Excision
Syringoma	Multiple 1–4 mm, firm, skin-colored to brownish-pink papules in symmetric, bilateral distribution on labia majora	Dermal proliferation of eccrine duct-like tubular structures lined with two layers of cuboidal epithelium	Reassurance; surgical excision, carbon dioxide laser vaporization, cryotherapy, or electrosurgery if symptomatic
Melanocytic neoplams			
Melanocytic nevus	Pink to dark black–brown macules or papules with well-demarcated borders and uniform pigmentation	Groups of benign nevi cells in basal epidermis, dermis, or both	Excision only indicated if malignancy suspected
Lymphovascular proliferations			
Angiokeratoma	Dark-red to purple papules 2–5 mm in diameter	Large dilated blood vessels in the superficial dermis with overlying epidermal hyperkeratosis	Reassurance; electrocautery or pulsed dye laser can be employed for cosmesis
Endometriosis	Tender, reddish-brown papules or nodules that cause cyclical pain in association with menses	Dermal proliferation of endometrial glands lined by pseudostratified columnar epithelium	Referral to gynecology; wide local excision
Lymphangioma	"Frog egg"-like grouped thin-walled vesicles	Dilated lymphatic channels with flat endothelium	Reassurance; recurrence common after surgical excision, laser, or sclerotherapy
Pyogenic granuloma	Glistening, friable, bright-red papule or nodule that bleeds spontaneously or after trauma	Lobular proliferation of capillary size blood vessels	Excision is recommended to rule out malignancy
Keratinocytic neoplasms			
Epidermolytic acanthoma	Single or multiple, variably pigmented (flesh-colored to white to erythematous) papules	Hyperkeratosis, papillomatosis, and focal vacuolar epithelial degeneration	Diagnostic biopsy to rule out condyloma acuminata
Seborrheic keratosis	Firm, "stuck-on" appearing, well-demarcated papules and plaques often with "waxy" or oily texture	Sharply demarcated proliferation of monotonous epidermal keratinocytes	Reassurance
Pilar/sebaceous lesions			
Pilar cyst	Smooth, mobile nodule, variable size; when ruptured, significant associated inflammation can occur	Cyst lined with multilayered epithelium with characteristic absence of the granular layer	Excision; incision and drainage in setting of cyst rupture
Pilomatricoma	Firm, subcutaneous nodules 1–3 cm in size, often with bluish-red hue	Circumscribed dermal nodule with shadow/ghost cells (basophilic cells resembling hair matrix cells with absent nuclei)	Excision only indicated if malignancy suspected
Trichoepithelioma	Flesh-colored or slightly erythematous, smooth and round papules, 1–5 mm in size	Small cords of basaloid cells in fibroblast-rich collagenous stroma	Excision only indicated if malignancy suspected

(*continued on next page*)

Table 1 (*continued*)

	Clinical appearance	Histopathologic features	Treatment
Other solid neoplasms			
Acrochordons	Fleshy, pedunculated, flesh-colored papules	Acanthotic, flattened, or frond-like epithelium overlying papillary-like dermis	Reassurance; snip excision, cryotherapy, or electrodessication can be employed for cosmesis
Leiomyoma	Solitary, flesh-colored to reddish-brown nodule, occasionally painful	Neoplastic proliferation of smooth muscle fibers	Excision
Lipoma	Soft, slow-growing subcutaneous nodule	Circumscribed proliferation of adipocytes	Excision
Schwannoma	Solitary, flesh-colored nodular lesion	Neoplastic proliferation of Schwann cells	Excision
Urethral caruncle	Solitary or multiple polypoid growths arising from posterior urethral mucosa	Proliferation of transitional and stratified squamous epithelium with vascular connective tissue	Excisional biopsy if lesion grows, becomes symptomatic, or fails to respond to conservative management

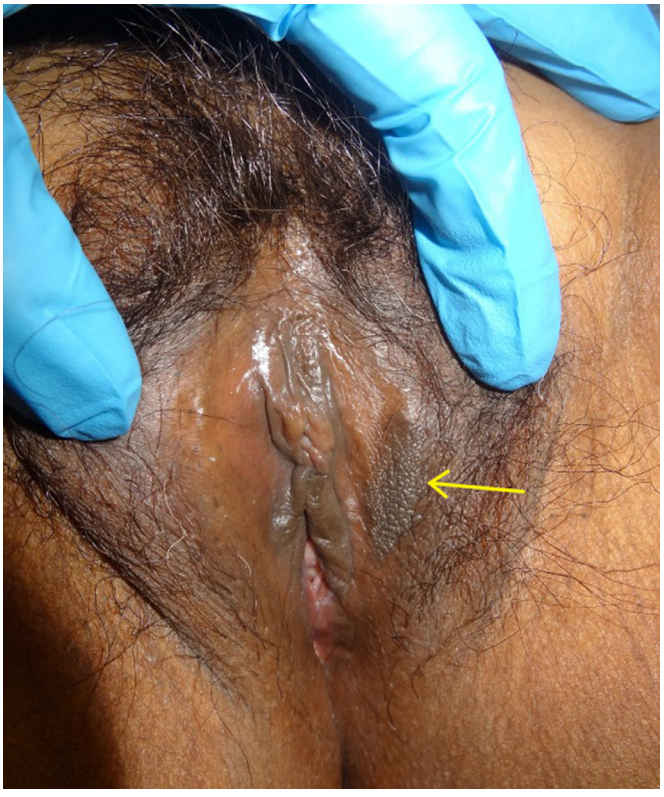

Fig. 4. Seborrheic keratosis.

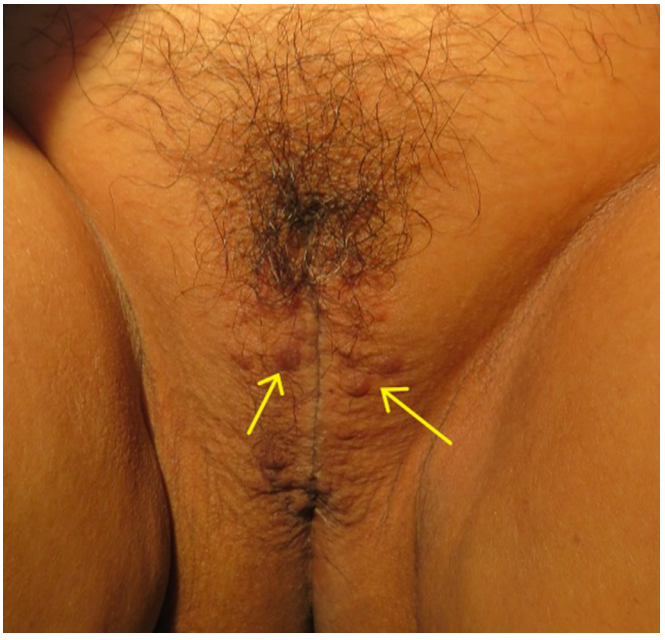

Fig. 5. Syringomas with symmetrical involvement of the labia majora.

cent studies have largely debunked this association (Reutter et al., 2014; Tardio et al., 2012).

SKs may be found on all keratinized skin surfaces, including the vulva (Fig. 4). SKs are characterized by their sharply demarcated, "stuck-on" appearance; color and size can vary considerably and the surface is often uneven and may appear oily or waxy (Hafner and Vogt, 2008). Of note, vulvar SKs may be less keratotic and exhibit less follicular plugging than SKs found on nonintertriginous sites (de Giorgi et al., 2005; Venkatesan, 2010). Although SKs typically can be identified macroscopically, dermoscopic evaluation can reveal characteristic keratotic invaginations, known as "comedone-like openings" and "milia-like cysts," that reinforce the diagnosis. These clinical and dermoscopic findings are echoed by the histologic features of SKs, which include papillomatous acanthotic epithelium consisting of basaloid cells with small regular nuclei, intraepidermal horn pseudocysts, and varying degrees of hyperpigmentation and hyperkeratosis (Heller, 2015).

SKs are benign neoplasms without malignant potential and thus do not require removal or clinical monitoring. However, in certain instances, darkly pigmented SKs may be difficult to distinguish from tumors of melanocytic origin. Dermoscopy serves as a valuable tool in these cases because melanocytic lesions feature pigment networks, streaks, and globules that are not present in SKs (de Giorgi et al., 2005; Hafner and Vogt, 2008). Should diagnosis still be unclear or a suspicious change be noted, biopsy is strongly encouraged because 0.5% of clinically suspected vulvar SKs are histologically proven melanomas (Edwards, 2010; Venkatesan, 2010). Furthermore, multiple or clustered SKs are uncommon on the vulva, particularly in patients without other cutaneous SKs. Such lesions also merit biopsy to rule out pigmented condyloma acuminata, HPV-associated vulvar intraepithelial neoplasia, or other malignancies (Edwards, 2010; Venkatesan, 2010).

Syringoma

Initially described as lymphangiomas by Kaposi and Biesiadeki (1872), syringomas have since been recognized as adnexal in origin—specifically, as benign tumors of the eccrine sweat glands (Mahajan et al., 2012). Although most commonly seen on the face and neck of young women, vulvar syringomas have also been described on the labia majora, typically appearing as multiple 1- to 4-mm, firm, skin-colored to brownish-pink papules in a symmetric, bilateral distribution (Fig. 5; Corazza et al., 2017; Dereli et al., 2007; Hoffman et al., 2020). These lesions are usually asymptomatic and diagnosed incidentally during routine examination; however, pruritus can be a complaint, with some patients ex-

periencing exacerbations during menstruation or warmer months (Gerdsen et al., 2002; Huang et al., 2003). Syringomas often arise during puberty and enlarge during pregnancy and with the use of oral contraceptives (Mahajan et al., 2012). It has therefore been suggested that the growth of syringomas is hormonally influenced, although the literature has remained inconclusive to date (Huang et al., 2003).

Unfortunately, given the rather banal appearance of vulvar syringomas, the differential diagnosis can be broad and includes Fox-Fordyce disease, EICs, steatocystoma multiplex, condyloma acuminata, milia, senile angiomas, lichen simplex chronicus, and lymphangioma circumscriptum (Table 1; Corazza et al., 2017; Mahajan et al., 2012). Thus, a biopsy with microscopic examination is often warranted (Hoffman et al., 2020). Histopathologically, the dermis features eccrine, duct-like, tubular structures lined with two layers of cuboidal epithelium embedded in a fibrous stroma (Corazza et al., 2017; Gerdsen et al., 2002; Mahajan et al., 2012).

If patients endorse significant pruritus, short courses of topical steroids and oral antihistamines may be trialed. Although topical atropine and topical tretinoin have both emerged as treatments for eruptive syringomas on the face and chest, their efficacy in vulvar syringomas remains to be seen (Hoffman et al., 2020; Huang et al., 2003). Definitive removal of the syringomas can be accomplished with surgical excision, carbon dioxide laser vaporization, cryotherapy, or electrosurgery (Huang et al., 2003).

Hidradenoma papilliferum

HP is a benign adenomatous neoplasm of anogenital, mammary-like glands, first described in 1878 by Worth (Baker et al., 2013). HP is most commonly seen in postpubertal women and typically affects the labia majora and labia minora with roughly equal frequency (Fig. 6). HP is rarely found on the clitoris or in the perineal region (Hernández-Angeles et al., 2017; Scurry et al., 2009). HP presents as a firm, solitary, smooth-surfaced, red, blue, or skin-colored nodule with a well-defined capsule, ranging in size from 0.5 to 2.0 cm (Kazakov et al., 2011; Lobo et al., 2017; Scurry et al., 2009). HP is usually asymptomatic but may become ulcerated, leading to pruritus, pain, and/or bleeding (Maldonado, 2014).

Histologically, HP shows both papillary and glandular architecture with pronounced glandular hyperplasia (Baker et al., 2013). There is a complex, "labyrinthine" pattern of branching tubules and acini forming the papillae, lined by cuboidal or columnar epithelial cells with pale eosinophilic cytoplasm and surrounded by a thin myoepithelial layer (Kazakov et al., 2011). Although there is prominent glandular proliferation, the mitotic index is usually low (Lobo et al., 2017). Adjacent normal mammary-like glands are often present (Scurry et al., 2009). Due to the proclivity of HP to ulcerate, clinically differentiating HP from adenocarcinoma may be difficult (Lobo et al., 2017). Furthermore, malignant transformation of HP has been described (Baker et al., 2013). Biopsy is therefore indicated to establish a diagnosis and rule out malignancy (Hernández-Angeles et al., 2017). The differential diagnosis should include mammary-like gland adenocarcinoma, Bartholin's cysts, lipomas, and syringocystadenoma papilliferum (Table 1; Hernández-Angeles et al., 2017; Maldonado, 2014). Treatment of HP is complete surgical excision, and recurrence is uncommon (Maldonado, 2014).

Cystic lesions

Epidermal inclusion cysts

EICs, also referred to as epidermal cysts, epidermoid cysts, epidermoid inclusion cysts, infundibular cysts, and sebaceous cysts,

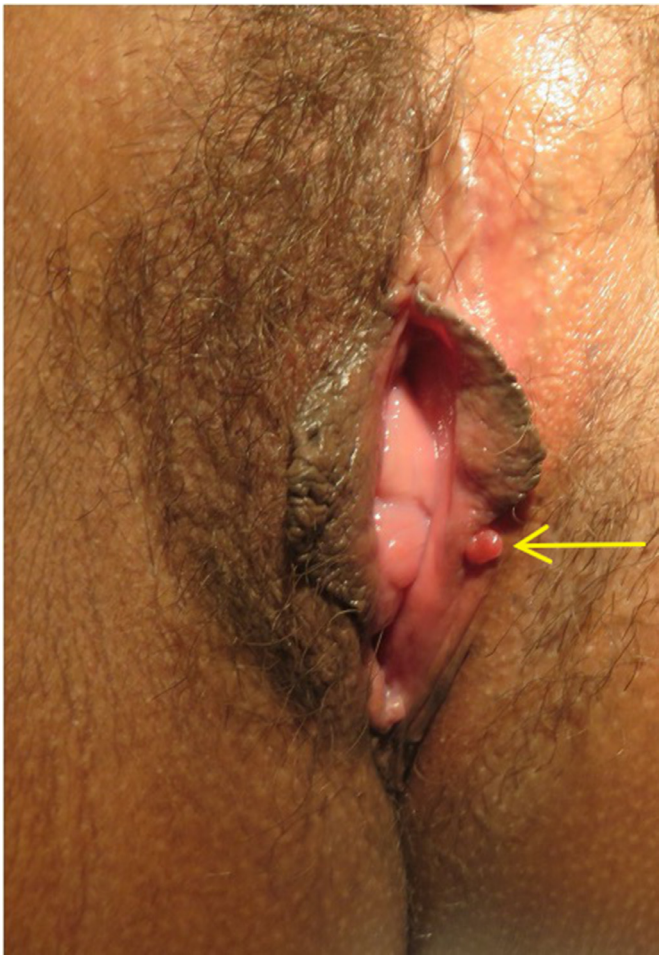

Fig. 6. Hidradenoma papilliferum on the labia minora.

are the most common cutaneous cyst. Of note, the term "sebaceous cyst" is a misnomer because EICs are derived from follicular infundibulum and do not feature sebaceous gland differentiation (Hoang et al., 2019). EICs arise when keratinizing squamous epithelium becomes trapped in the dermis, which can occur spontaneously or after trauma to the pilosebaceous unit (Heller, 2015; Nigam et al., 2017). The development of vulvar EICs is a well-described complication of female genital mutilation (Rouzi, 2010).

EICs typically present as firm, round, yellow-white papulonodules. Vulvar EICs are most often found on the labia majora, can be multicystic, and range in size from a few millimeters to several centimeters in diameter (Fig. 7; Apostolis et al., 2012; Pehlivan et al., 2015; Yang et al., 2012). In rare cases, EICs may also affect the labia minora and clitoris (Pehlivan et al., 2015). The clitoral location is most closely associated with previous female genital mutilation (Heller, 2015). Notably, epidermoid cysts may occur at any age—tiny superficial epidermoid cysts, termed milia, have even been described in neonates (Hoang et al., 2019).

EICs are lined with stratified squamous epithelium, which produces keratin and causes the cyst to fill with caseous debris (Heller, 2015). Histopathologic examination reveals a cyst lined with keratinizing stratified squamous epithelium with an intact granular layer filled with laminated keratin. If the cyst ruptures, the keratin can cause a foreign-body reaction with a dense inflammatory infiltrate featuring multinucleated giant cells and histiocytes (Cuda et al., 2019; Nigam et al., 2017). EICs are generally slow growing and asymptomatic but may become secondarily infected, inflamed, or rupture (Cuda et al., 2019; Rouzi, 2010). EICs are be-

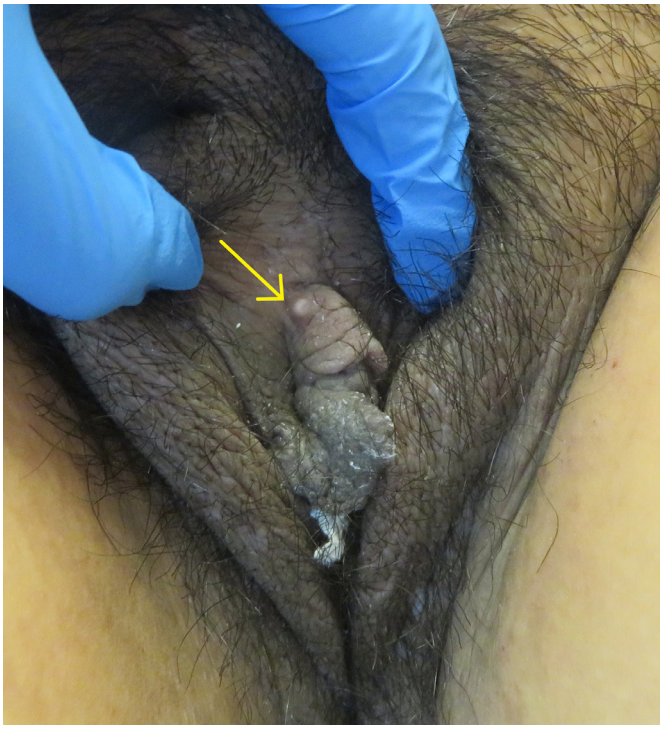

Fig. 7. Epidermal inclusion cyst on the clitoral hood.

nign and do not require removal or treatment. If symptomatic or if the patient requests removal, complete excision of the cyst lining is required to prevent recurrence and should be performed when the cyst is not inflamed (Endrizzi, 2017). Incision and drainage may be required if the cyst becomes infected, purulent, or painful (Cuda et al., 2019). For smaller cysts that become inflamed, intralesional steroids may be considered (Weir and St. Hilaire, 2020).

The differential for vulvar EICs includes Bartholin's gland cysts, lipomas, Skene's duct cysts, VGCs, cysts of the canal of Nuck, syringomas, and endometriomas (Table 1) (Pehlivan et al., 2015; Yang et al., 2012). There have been reports of squamous cell carcinoma, basal cell carcinoma, and other malignancies arising from EICs, including in the vulva (Delacretaz 1977; Sze et al., 2016). Malignancy should be suspected if the EIC is rapidly enlarging, recurring, or not responding to treatment. In these situations, excisional biopsy should be pursued.

Vestibular gland cysts

VGCs, also termed vestibular cysts or mucinous cysts, are benign cysts of the vulva. As the name suggests, VGCs are found within the vestibule of the vulva on the medial labia minora and are of minor vestibular gland origin (Maldonado, 2014). The cysts are soft, smooth, round, and range in size from 2 to 30 mm (Scurry and McGrath, 2012). VGCs can be distinguished from other vulvar cysts by their translucent nature, which is attributable to the clear, liquid mucin they contain (Fig. 8; Karakaya et al., 2018; Maldonado, 2014; Scurry and McGrath, 2012). The lining of these cysts is a simple mucous-secreting columnar epithelium, sometimes with squamous metaplasia and rarely with ciliated epithelium (Anderson, 2017; Maldonado, 2014). Hormonal involvement in the formation of VGCs has been suggested, given that onset is most frequently between puberty and the fourth decade in parous women and those exposed to contraceptives. There are also reports of cysts being strongly estrogen receptor positive (Scurry and McGrath, 2012).

VGCs are generally asymptomatic; thus, reassurance to patients can be provided. However, if there is associated pain, discomfort, or cosmetic distress, surgical excision can be performed (Karakaya et al., 2018; Maldonado, 2014). Although there is evidence of marsupialization of gland cysts in the vulvar vestibule (e.g., Bartholin's and Skene's gland cysts), limited data on this approach in VGCs exist (Campbell et al., 2019). As with EICs, VGCs may be mistaken for Bartholin's gland cysts, Skene's gland cysts, Gartner's duct cysts, and cysts of the canal of Nuck (Table 1; Campbell et al., 2019). Biopsy can be deferred unless features suspicious for malignancy are identified (Lee et al., 2014).

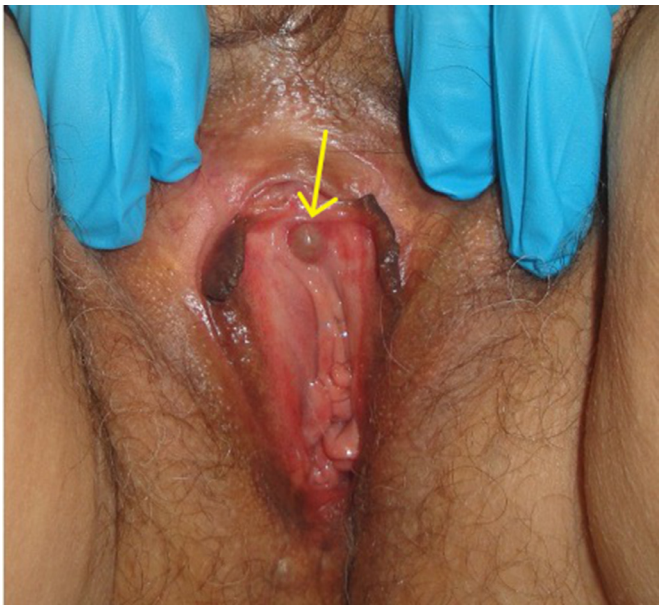

Fig. 8. Vestibular gland cyst in the medial vestibule.

Conclusion

Vulvar dermatology lies at the cross-section of gynecology, dermatology, and women's health. Although traditionally underrecognized and understudied, the diagnosis and management of benign vulvar lesions lies firmly within the purview of the dermatologist's practice. Herein, we highlighted some of the most commonly encountered anatomic variants and benign neoplasms of the vulva of which every dermatologist should be aware. We hope this manuscript serves as a framework for practicing dermatologists to help distinguish benign vulvar lesions from their pathologic mimickers.

Declaration of Competing Interest

None.

Funding

None.

Study approval

None.

Supplementary materials

For patient information on skin cancer in women, please click on Supplemental Material - Patient Page. Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.ijwd.2021.04.007.

References

Altmeyer P, Chilf GN, Holzmann H. Pseudocondylomata of the vulva. Geburtshilfe Frauenheilkd 1981;41(11):783–6.

Anderson SR. Benign vulvovaginal cysts. Diagn Histopathol 2017;23(1):14–18.

Apostolis CA, Von Bargen EC, DiSciullo AJ. Atypical presentation of a vaginal epithelial inclusion cyst. J Minim Invasive Gynecol 2012;19(5):654–7.

Bachmann GA, Nevadunsky NS. Diagnosis and treatment of atrophic vaginitis. Am Fam Physician 2000;61(10):3090–6.

Baker GM, Selim MA, Hoang MP. Vulvar adnexal lesions: A 32-year, single-institution review from Massachusetts General Hospital. Arch Pathol Lab Med 2013;137(9):1237–46.

Burkland CE. Common lesions of the urethra in women. Calif Med 1952;76(2):69–73.

Campbell K, Panza J, Zimmerman C. Symptomatic vulvar mucinous cyst: A case report and review of the literature. Case Rep Womens Health 2019:24.

Chiba M, Toki A, Sugiyama A, Suganuma R, Osawa S, Ishii R, et al. Urethral caruncle in a 9-year-old girl: A case report and review of the literature. J Med Case Rep 2015;9:71.

Coban S, Biyik I. Urethral caruncle: Case report of a rare acute urinary retension cause. Can Urol Assoc J 2014;8(3–4):E270–2.

Conces MR, Williamson SR, Montironi R, Lopez-Beltran A, Scarpelli M, Cheng L. Urethral caruncle: Clinicopathologic features of 41 cases. Hum Pathol 2012;43(9):1400–4.

Corazza M, Borghi A, Minghetti S, Ferron P, Virgili A. Dermoscopy of isolated syringoma of the vulva. J Am Acad Dermatol 2017;76(2S1):S37–9.

Cuda JD, Rangwala S, Taube JM, et al. Benign epithelial tumors, hamartomas, and hyperplasias. In: Kang S, Amagai M, Bruckner AL, Enk AH, Margolis DJ, McMichael AJ, et al, editors. Fitzpatrick's Dermatology. New York, NY: McGraw-Hill Education; 2019.

Delacretaz J. Keratotic basal-cell carcinoma arising from an epidermoid cyst. J Dermatol Surg Oncol 1977;3(3):310–11.

Dereli T, Turk BG, Kazandi AC. Syringomas of the vulva. Int J Gynaecol Obstet 2007;99(1):65–6.

Edwards L. Pigmented vulvar lesions. Dermatol Ther 2010;23(5):449–57.

Endrizzi B. Benign tumors and vascular lesions. In: Soutor C, Hordinsky MK, editors. Clinical Dermatology. New York, NY: McGraw-Hill Education; 2017.

Ferrier PA. Urethral caruncle. Cal West Med 1926;24(4):500–1.

Fruchter R, Melnick L, Pomeranz MK. Lichenoid vulvar disease: A review. Int J Womens Dermatol 2017;3(1):58–64.

Gerdsen R, Wenzel J, Uerlich M, Bieber T, Petrow W. Periodic genital pruritus caused by syringoma of the vulva. Acta Obstet Gynecol Scand 2002;81(4):369–70.

de Giorgi V, Massi D, Salvini C, Mannone F, Carli P. Pigmented seborrheic keratoses of the vulva clinically mimicking a malignant melanoma: A clinical, dermoscopic-pathologic case study. Clin Exp Dermatol 2005;30(1):17–19.

Hafner C, Vogt T. Seborrheic keratosis. J Dtsch Dermatol Ges 2008;6(8):664–77.

Haley JC, Mirowski GW, Hood AF. Benign vulvar tumors. Semin Cutan Med Surg 1998;17(3):196–204.

Heller DS. Benign tumors and tumor-like lesions of the vulva. Clin Obstet Gynecol 2015;58(3):526–35.

Hernández-Angeles C, Nadal A, Castelo-Branco C. Hidradenoma papilliferum of the vulva in a postpartum woman: A case report. J Obstet Gynaecol 2017;37(5):683–4.

Hoang VT, Trinh CT, Nguyen CH, Chansomphou V, Chansomphou V, Tran TTT. Overview of epidermoid cyst. Eur J Radiol Open 2019;6:291–301.

Hoffman BL, Schorge JO, Halvorson LM, Hamid CA, Corton MM, Schaffer JI. Benign disorders of the lower reproductive tract. 4 ed. New York, NY: McGraw-Hill Education; 2020 Williams Gynecology.

Huang YH, Chuang YH, Kuo TT, Yang LC, Hong HS. Vulvar syringoma: A clinicopathologic and immunohistologic study of 18 patients and results of treatment. J Am Acad Dermatol 2003;48(5):735–9.

Karakaya BK, Kansu-Celik H, Ersak B, Ozyer S, Turker M, Evliyaoglu O. An unusually large mucinous cyst of vulva. Gynecol Reprod Endocrinol 2018;02(01).

Kazakov DV, Spagnolo DV, Kacerovska D, Michal M. Lesions of anogenital mammary-like glands. Adv Anat Pathol 2011;18(1):1–28.

Kelekçi KH, Özyurt S, Özkan B, Karaca Ş, Karakuzu A, Bilgin İ. The impact of inflammatory and infectious diseases of vulvar on quality of life. J Menopausal Med 2016;22(3).

Kwon OS, Hwang EJ, Bae JH, Park HE, Lee JC, Youn JI, et al. Seborrheic keratosis in the Korean males: Causative role of sunlight.. Photodermatol Photoimmunol Photomed 2003;19(2):73–80.

Lee MY, Dalpiaz A, Schwamb R, Miao Y, Waltzer W, Khan A. Clinical pathology of Bartholin's glands: A review of the literature. Curr Urol 2014;8(1):22–5.

Lloyd J, Crouch NS, Minto CL, Liao LM, Creighton SM. Female genital appearance: "Normality" unfolds. BJOG 2005;112(5):643–6.

Lobo RA, Gershenson DM, Lentz GM, Valea FA. Comprehensive gynecology. 7th edition. Philadelphia, PA: Elsevier; 2017.

Mahajan R, Bang D, Nagar A, Bilimoria F. Rare sweat gland tumors of vulva: Report of two cases. Indian J Sex Transm AIDS 2012;33(2):124–7.

Maldonado VA. Benign vulvar tumors. Best Pract Res Clin Obstet Gynaecol 2014;28(7):1088–97.

Moyal-Barracco M, Leibowitch M, Orth G. Vestibular papillae of the vulva. Lack of evidence for human papillomavirus etiology. Arch Dermatol 1990;126(12):1594–8.

Muhammed RT, Afra TP, De D. Vestibular papillomatosis: A normal variation commonly misdiagnosed as genital condylomata. Am J Obstet Gynecol 2019;220(4):403.

Nguyen J, Duong H. Anatomy, abdomen and pelvis, female external genitalia. Treasure Island, FL: StatPearls; 2020.

Nigam JS, Bharti JN, Nair V, Gargade CB, Deshpande AH, Dey B, et al. Epidermal cysts: A clinicopathological analysis with emphasis on unusual findings. Int J Trichology 2017;9(3):108–12.

Pehlivan M, Ozbay PO, Temur M, Yilmaz O, Gumus Z, Guzel A. Epidermal cyst in an unusual site: A case report. Int J Surg Case Rep 2015;8C:114–16.

Reutter JC, Geisinger KR, Laudadio J. Vulvar seborrheic keratosis: Is there a relationship to human papillomavirus? J Low Genit Tract Dis 2014;18(2):190–4.

Rouzi AA. Epidermal clitoral inclusion cysts: Not a rare complication of female genital mutilation. Hum Reprod 2010;25(7):1672–4.

Sarifakioglu E, Erdal E, Gunduz C. Vestibular papillomatosis: Case report and literature review. Acta Derm Venereol 2006;86(2):177–8.

Schlosser BJ, Mirowski GW. Lichen sclerosus and lichen planus in women and girls. Clin Obstet Gynecol 2015;58(1):125–42.

Scurry J, McGrath G. Multiple mucinous cysts on the anterior of Hart's lines of the vulva. Pathology (Phila) 2012;44(5):479–80.

Scurry J, van der Putte SCJ, Pyman J, Chetty N, Szabo R. Mammary-like gland adenoma of the vulva: Review of 46 cases. Pathology (Phila) 2009;41(4):372–8.

Sze S, Richmond I, Bickers A, Saha A. Squamous cell carcinoma arising from a vulval epidermal cyst. J Obstet Gynaecol Res 2016;42(11):1623–6.

Tardio JC, Bancalari E, Moreno A, Martin-Fragueiro LM. Genital seborrheic keratoses are human papillomavirus-related lesions. A linear array genotyping test study. APMIS 2012;120(6):477–83.

Tunitsky E, Goldman HB, Ridgeway B. Periurethral mass: A rare and puzzling entity. Obstet Gynecol 2012;120(6):1459–64.

Venkatesan A. Pigmented lesions of the vulva. Dermatol Clin 2010;28(4):795–805.

Verkauf BS, Von Thron J, O'Brien WF. Clitoral size in normal women. Obstet Gynecol 1992;80(1):41–4.

Weber AM, Walters MD, Schover LR, Mitchinson A. Vaginal anatomy and sexual function. Obstet Gynecol 1995;86(6):946–9.

Weir CB, St Hilaire NJ. Epidermal inclusion cyst. Treasure Island, FL: StatPearls; 2020.

Welch JM, Nayagam M, Parry G, Das R, Campbell M, Whatley J, et al. What is vestibular papillomatosis? A study of its prevalence, aetiology and natural history. Br J Obstet Gynaecol 1993;100(10):939–42.

Yang WC, Huang WC, Yang JM, Lee FK. Successful management of a giant primary epidermoid cyst arising in the labia majora. Taiwan J Obstet Gynecol 2012;51(1):112–14.

Yeatman JM, Kilkenny M, Marks R. The prevalence of seborrhoeic keratoses in an Australian population: Does exposure to sunlight play a part in their frequency? Br J Dermatol 1997;137(3):411–14.