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A Review of Lesions of the Posterior Fourchette, Posterior Vestibule (Fossa Navicularis), and Hymen

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Running title: Lesions of the Posterior Fourchette, Posterior Vestibule, and Hymen
Precis:

Lesions of the posterior fourchette, posterior vestibule (fossa navicularis), and hymen are reviewed.
Abstract:

Lesions specific to the posterior fourchette, posterior vestibule (fossa navicularis), and hymen are reviewed. A knowledge of these regional lesions will be helpful if such a patient is encountered.

Key words: Vulvar diseases, vulvar neoplasms, hymen,
**Introduction:**

The posterior fourchette, posterior vestibule (fossa navicularis), and hymen (figure 1) may develop lesions that are unique, in addition to usual vulvar and vaginal lesions. This review discusses specific pathology that occurs in this region (tables 1, 2).

**Embryology, Anatomy & Histology**

The embryology of the female external genitalia is not completely understood. The labioscrotal folds of the indifferent embryo, when differentiating towards female development, fuse posteriorly, forming the posterior labial commissure, which delineates the beginning of the posterior introitus, the fourchette. The vestibule arises from the urogenital sinus, which meets up with the developing vagina, separated by the hymen, which is formed from an invagination of the urovaginal sinus. The hymen ruptures during the perinatal period (1). The vestibule ends at the hymen, which is normally perforated by the vaginal opening, technically the introitus. The term introitus is also used more broadly, encompassing the fourchette and posterior vestibule (fossa navicularis) as well as anterior vestibule. The fourchette is comprised of the joining of the labia minora and the anteriormost portion of the perineum. The fourchette is lined by stratified squamous epithelium with a thin keratin layer, and may contain sebaceous glands. The vestibule begins posteriorly from Hart’s line, which delineates the change from the keratinized squamous epithelium of the fourchette to the nonkeratinized squamous epithelium of the vestibule. The vestibule is thought to be derived from endoderm, unlike the ectodermally derived labia (2). The urethra, vagina, Skene’s (paraurethral) and two Bartholin’s ducts empty into the vestibule.
Urethral, paraurethral and Bartholin’s lesions have been previously reviewed(3,4), and this paper covers the fourchette, the fossa navicularis, the concave portion of the vestibule that resides posteriorly between the fourchette and the outer hymen, and the hymen. The fossa navicularis is lined by nonkeratinizing stratified squamous epithelium, and may contain minor vestibular glands (5), mucin producing glands which are lined by mucinous columnar epithelium, and have transitional epithelial-lined ducts, but may undergo metaplasia. The hymen is lined by glycogen containing non-keratinized stratified squamous epithelium.

Congenital Anomalies

Anomalies of the Fourchette:

An uncommon anomaly involving the fourchette is perineal groove; which is comprised of a red wet mucosal lined groove that extends from the fourchette back to the anus, due to failure of fusion of the median raphe(6).

Anomalies of the Posterior Vestibule:

Even in the absence of diethylstilbestrol(DES) exposure in utero, adenosis, the persistence of glandular elements, may be present in the vestibule, and may appear as raw red mucosa, which may be friable and bleed. Adenosis may also develop secondarily in the vestibule after Stevens-Johnson syndrome(7). Heterotopia, the occurrence of tissue in an incorrect
location, can occur at the introitus, and intestinal (enteric) heterotopia has been described. The patient of Horn et al(8) was an 82 year old woman who presented with intestinal heterotopia of the vulva and introitus, presenting as dyspareunia, in a background of lichen sclerosus. The lesion was described as white and papillary with red areas. Rarely, a congenital fistula between the anus and vulvar vestibule can occur. This lesion, termed “perineal canal” is a rare variant of anorectal malformation consisting of a fistula between the anus and vulvar vestibule(9).

**Hymenal anomalies**

The purpose of the hymen has never been established(10). The hymen may have isolated anomalies, or these may occur in association with other genital tract anomalies. In combination with vaginal agenesis, a hymen may or may not be present. It has been estimated that in the general population, hymenal agenesis has an incidence of one in several hundred to 1000(11), although reliable statistics are not available. In one study, hymenal absence with vaginal agenesis was associated with greater risk of renal agenesis, and increased failure of dilation therapy(11). More commonly encountered by clinicians is imperforate hymen, which is estimated to occur in about 1 in 2000 girls(12). The hymen may also be microperforate, cribriform or septate. Imperforate hymen is usually sporadic, but rare familial cases have been reported(13). Imperforate hymen may present early in childhood, or even in utero life with mucocolpos, secondary to mucus produced under the influence of maternal hormones, or may present after menarche with hematocolpos. Mucocolpos will be seen as bulging of the hymen on examination of the newborn. If not appreciated, the mucus may resorb, and then the patient can
present after menarche with cyclic pain, amenorrhea, and bulging of the hymen with a blue discoloration. A significant pelvic mass comprised of blood may accumulate. Less common symptoms include tenesmus, back pain, urinary obstruction, intestinal obstruction or vascular obstruction(14). Rarely, life-threatening sequelae have been reported in infants with imperforate hymen, including post-obstructive acute renal failure secondary to urethral obstruction by the mass produced by mucocolpos(15), and fetal or neonatal ascites or peritoneal calcification(16). Fetal urinary obstruction by mucocolpos may secondarily result in severe oligohydramnios(17). Nazir et al(18) described congenital obstructions, combining the most common imperforate hymen with other etiologies, mainly transverse vaginal septum, and described symptoms as varying by age. Neonates in this series commonly presented with abdominal mass, sepsis or respiratory distress, adolescents with abdominal pain, voiding dysfunction and back pain, and adults with inability to have intercourse and infertility. Of note, some of both the adolescent and adult groups had endometriosis, which is potentially related, due to retrograde menses(18). Hydrocolpos or mucocolpos may become infected, secondary to urinary tract infections in infancy, hematogenous spread of bacteria in adolescence, or rarely due to iatrogenic introduction of bacteria due to needle puncture of an imperforate hymen(19). Tumor markers are not needed for the diagnosis of imperforate hymen, however clinicians should be aware that patients may show elevated CA125 and CA19-9(20), although the etiology for the false positive results is unclear.
Infections and Inflammations

Vestibulodynia

Our understanding of vestibulodynia is evolving. This challenging condition presents with severe introital tenderness on touch, with entry dyspareunia making intercourse difficult to impossible. Originally thought to be inflammatory or psychogenic, subsequently some recent studies have shown both an increase in nerve fibers as well as degranulated mast cells(21), but these findings are not universally supported, and there is no consensus on exact etiology. Treatment is often multidisciplinary, and may include neuromodulators, biofeedback, sexual counseling, and physical therapy as well as comfort care(22). Refractory cases may come to vestibulectomy.

Vestibular papillomatosis

Regular symmetrical tiny 3-4 mm finger-like projections may be present at the introitus. These are now considered an anatomic variant, and are not related to human papillomavirus or genital warts. Aside from matching the color of the mucosa, and the lack of koilocytosis seen in vestibular papillae, each papilla originates from its own base, in contradistinction to condyloma, which branch out from a common base(23). Good lighting and magnification, which could include vulvoscopy, should assist the clinician in making the distinction and avoiding unnecessary biopies.
**Graft versus Host Disease**

Narrowing or obliteration of the introitus with its attendant difficulties may be a manifestation of graft versus host disease in patients who have undergone bone marrow transplantation(24). The underlying etiology of graft versus host disease is thought to be activated donor T-cells reacting against host histocompatibility antigens(25). Findings include thickening of mucosa and the formation of synechiae, often with ulceration. Therapy has included topical estrogens, topical steroids, topical cyclosporine, dilators and surgery(24,26). It has been suggested that the use of topical immunosuppresants may reactivate human papillomavirus disease of the lower genital tract(26).

**Lichen planus & Lichen sclerosus**

Lichen planus(LP) can sometimes be difficult to distinguish from lichen sclerosus(LS). LS affects the vulvar skin, and hence may be present at the fourchette, where it may cause fissuring. LP characteristically involves the posterior vestibule, where there is usually well-demarcated erosion or glazed erythema, sometimes with white stria. Unlike LS, LP can extend into the vagina, which can assist in making the distinction. The vaginal involvement by LP may appear similar to desquamative inflammatory vaginitis, with a similar profuse discharge. Oral lesions should be looked for as well with LP. Patients are usually mid-life for LP, and present with burning, stinging, and rawness. LS patients are either children, or elderly in most cases, and
more likely to present with pruritis. Although biopsy is not always undertaken, there is a characteristic histology for both conditions, with sawtooth acanthosis with a band-like chronic dermal inflammatory infiltrate for LP, and loss of rete pegs, and dermal homogenization with a variable dermal chronic inflammatory infiltrate in LS. These findings are very helpful to the pathologist when present, although these diagnostic features may not be present. These difficult to manage chronic conditions are usually treated with topical steroids.

**Ulceration**

Ulcers, defined as loss of the full thickness of the epithelium may be of infectious etiology or noninfectious. Most do not have pathognomonic histopathologic features, although biopsy may become necessary. Their occurrence raises the question of a sexually transmitted disease such as herpes, syphilis or chancre. Another rare infectious cause of ulceration in the posterior fourchette includes tuberculosis(27).

Noninfectious causes of ulceration include as a manifestation of an anovaginal fistula(28). Aphthous ulceration at the introitus can be seen in adolescents and young women, sometimes in association with Epstein-Barr virus(29) or cytomegalovirus, but often no specific agent is found(30), and hence the etiology is uncertain. These ulcers are similar to oral canker sores(apthae), and there may be a prior history of oral apthae. The vulvar ulcers may be associated with a viral syndrome, concurrent oral lesions, and occasionally ophthalmologic manifestations, and are extremely painful. Vulvar apthae may be necrotic in appearance. Usually they are a one-time recurrence, but if recurrent, ophthalmologic evaluation should be performed to rule out Behçet’s syndrome. Initial therapy may include prednisone.
Fissures

Fissures are cracks in the epidermis that are not full-thickness. These differ from erosion, a more diffuse partial loss of epithelium. Fissuring of the posterior fourchette may occur in association with such conditions as lichen sclerosus or contact dermatitis(31), or fungal infection. Recurrent idiopathic fissuring of the posterior fourchette is a chronic condition of unknown etiology that presents with dyspareunia, as well as pain on examination or tampon insertion. Often intercourse causes refissuring after early healing has occurred. Medical management has been attempted, but some patients come to perineoplasty(31). Medical management was tailored to specific underlying conditions in this study, but more general measures included avoidance of contact irritants such as spermicides, avoidance of inappropriate treatment with anti-fungals, and utilization of sitz baths and topical steroids(31). Surgical therapy(perineoplasty) is reserved for refractory cases(31). Histology shows hyperkeratosis, parakeratosis, and chronic inflammation(32), all nonspecific findings.

In Michlewitz’s patients(33), hymenal fissures were in the 4 or 8 o’clock positions. Laser ablation has been utilized with some success(33).
Benign Tumors and Tumor-like Lesions

Epidermal cyst

Epidermal inclusion cysts may be associated with prior surgery such as episiotomy, however may arise spontaneously(35).

Mucinous cyst

Mucinous cysts may also arise in the anterior or posterior vestibule, and are thought to be Müllerian in origin(36), but it seems plausible that some may be of minor vestibular gland origin as well. Histologically, mucinous cysts are comprised of a cavity lined by a single layer of mucinous epithelium.

Adenomas

Minor vestibular glands may be the source of adenomas. In one series of patients who had undergone excisions for vulvar pain, there were incidental 1-2 cm adenomas found in several cases, composed of crowded mucous glands similar to minor vestibular glands(37). Multinodular vestibular gland hyperplasia can also occur(38). Rare lesions such as tubular apocrine adenoma(39) have been reported, and may be derived from mammary-like glands. These glands are known to arise in the vulva, including the fourchette, and hence a variety of breast-like neoplasms may arise in them (40).

Mixed tumors:

Rare vaginal neoplasms resembling salivary gland mixed tumors with a propensity for the hymenal ring(41) have been described, and have behaved in a benign fashion after excision.
**Hymenal cysts, polyps, and tags**

Hymenal skin tags and cysts may be seen in the newborn, but hymenal cysts, polyps and tags can occur in women of all ages. Hymenal cysts often resolve spontaneously(42). Borko et al report a large hymenal polyp in a virginal 21-year-old. This is an unusual occurrence of a lesion most commonly seen in infants, and resolving spontaneously in the first few years of life(43). Excision revealed a polyp with benign non-keratinizing squamous epithelium lining a fibrovascular core (43).

**Bartholin’s Gland Masses**

Lesions of the Bartholin’s ducts and glands have been reviewed previously(4). Most masses are cysts or abscesses of the duct. The question of malignancy should always be considered in a mass in a woman over 40 years.

**Malignant Neoplasms:**

The majority of neoplasms affecting the introitus are pre-invasive and invasive squamous lesions, which are not specific to the introitus.

A lesion described as an adenocarcinoma of sweat gland was reported as arising from the posterior fourchette. The lesion was described as resembling intraductal carcinoma of the breast(44), suggesting that it may have arisen from the mammary-like anogenital glands of the region. A lesion described as a colonic-type mucinous adenocarcinoma arising in a villous
adenoma has also been reported as arising in the fourchette(45). The authors questioned the prevailing theory of cloacal remnant origin, suggesting instead entrapment of rectal mucosa in a fourth degree laceration repair. Episiotomies may also contribute to the development of the rare cancers in the region, as in a case of clear cell adenocarcinoma arising in endometriosis in an episiotomy site(46). Although malignant transformation of endometriosis is low, the development of clear cell adenocarcinoma in endometriosis is a well described phenomenon. Metastatic carcinoma may also present at the introitus. Endometrial carcinoma has metastasized to the introitus, and lesions may recur in an episiotomy, such as a case of metastatic cervical carcinoma(47), where the authors postulated implantation during delivery. While most nodules in episiotomies are benign squamous inclusion cysts, mucinous cysts, or endometriosis, malignancy must be considered as well.

Extremely rarely, malignancies may arise from the hymen. Dubé et al(48) describe a villoglandular adenocarcinoma-in-situ of intestinal type arising in the hymen of a 64 year old woman, postulating de novo origin from the squamous epithelium. The authors discounted other theories, such as the neoplasm arising from minor vestibular glands, cloacogenic remnants or adenosis.

**Summary:**

The introitus rarely comes to specific attention unless there is a complaint, usually dyspareunia. A knowledge of specific lesions of the region will be helpful if such a patient is encountered.
References:


