

An Unusual Case of Large Vulvar Soft Fibroma in an Adult Patient and Review of the Literature

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We present a case of large vulvar fibroma that was effectively treated with surgical excision. The present case is among the largest case reported in the literature. A 25-year-old female patient applied to our clinic with a complaint of a growing mass in external genitalia. Patient's complaint began 10 years ago as a small papillomatous lesion in the right labia majora, which increased in size progressively reaching to present size of 20-30 cm in 2-3 years after the first symptom. The mass was prediagnosed as vulvar soft fibroma, and surgical excision was decided with the request and approval of the patient. For histopathologic evaluation of the specimen, H&E and HPV immunohistochemistry staining were applied. Under light microscopy, polypoid lesions covered with acanthotic squamous epithelium were seen in serial sections. Stroma was rich in collagen with homogenous structure and had fibroblastic proliferation in loose-lined, randomized bundles. Thick-walled and ectatic vascular proliferations were observed especially in the central part of the lesion. Chronic inflammatory cell infiltration was present in the subepithelial area. Immunohistochemically, there was no positivity for HPV Biocare / BPV1 in squamous epithelial cells. The histopathological findings were compatible with fibroepithelial polyp. The patient had no postoperative complication, and there was no residue or recurrence at the last assessment on postoperative 3rd month. In order to avoid this unwanted consequence, vulvar fibroma should be excised without delay and also histologically differentiated from other tumors originating from vulva that can be potentially malignant. Soft fibroids are described as pedunculated tumors due to an elongation of their connective tissue, especially in superficial tumors. When localized in the vulva, they occur more often in the labia majora and less frequently in the labia minora, clitoris, vestibule, and posterior commissure. In tumors of long clinical duration, ulcerations with superficial hemorrhage, often due to repeated trauma, are often observed. For starters, the tumor may be asymptomatic but has the potential to grow to enormous sizes. In addition to causing physical signs due to its size and location, the important differential diagnosis includes lipoma, inguinal hernia, vulvovaginal

cysts, vulvar elephantiasis, and fibroepitheloid tumors. Depending on the histological variants, fibroids of the vulva can be classified as pure or mixed. The pure form can be hard or soft. The pure-hard variant presents a fibrous and hard connective tissue, white or pink in color, and lobulated and well differentiated in appearance. Treatment for malignant tumors includes: surgery, radiation therapy, and chemotherapy. The treatment recommended for each case depends on several factors such as the stage of the cancer, the exact subtype or "quality" of the cancer, and general health. Total vulvectomy with bilateral inguinal lymphadenectomy in one piece is the intervention of choice in cases of malignancy, the treatment of benign tumors involves surgical removal: cryo-surgery, electrocoagulation or laser surgery. For our patient, we perform a biopsy resection for aesthetic purposes, knowing that our attitude will be final in the event of a benign tumor, will not delay the treatment if it was a malignant tumor or will adversely affect the quality of the cases taken in. malignant tumor burden. Human papillomavirus (HPV) infections of the vulva are common clinical events. HPV infections are classified as clinical or subclinical, depending on the infectivity of the virus and the response of the affected epithelium. The typical expression is a soft, pinkish-white papillary epithelial lesion. These lesions can occur singly or in clusters which can become confluent. The usual vulvar locations are the foreskin, the vestibule and the perineal body. Perianal and anal loci are also commonly seen. Histologically, condylomas appear as epithelial papillomas with acanthosis and parakeratosis. Some epithelial cells have atypical nuclei and perinuclear halos that are believed to be a manifestation of HPV infection. These cells are called koilocytes. The underlying stroma usually shows a mild inflammatory response. Biopsies of condylomatous lesions resistant to treatment or abnormal appearance should be taken for histological confirmation of the diagnosis. Differentiation between condyloma and vulvar papillomatosis or other vulvar lesions can then be made. Treatment consists of destroying the skin manifestations of HPV infection. Eradication of the virus from the epithelium is not clinically possible.

Control of epithelial viral expression can be accomplished with topical agents such as trichloroacetic acid, bichloroacetic acid, podophyllin or its derivatives, or topical 5-fluorouracil (5-FU). Cryotherapy, laser vaporization, and electroexcision or desiccation are other methods of treatment. Intralesional or systemic administration of interferon is another therapeutic option that usually is reserved for recalcitrant lesions. Recurrent viral cutaneous expressions usually are managed with alternative treatments or combinations of treatments. Verrucous carcinoma of the vulva appears as a large condyloma or a lesion suspicious for invasive carcinoma. The lesion, first described by Buschke and Lowenstein as a giant condyloma,² is associated with the HPV 6 viral subtype.¹ Verrucous carcinoma may involve large areas of the vulva, and it has a pushing rather than an infiltrative border. Characteristically, it is localized to the vulva. Management is best accomplished by wide excision and careful postoperative evaluation. Human papillomavirus (HPV) infections of the vulva are common clinical events. HPV infections are classified as clinical or subclinical, depending on the infectivity of the virus and the response of the affected epithelium. The typical expression is a soft, pinkish-white papillary epithelial lesion. These lesions can occur singly or in clusters which can become confluent. The usual vulvar locations are the foreskin, the vestibule and the perineal body. Perianal and anal loci are also commonly seen. Histologically, condylomas appear as epithelial papillomas with acanthosis and parakeratosis. Some epithelial cells have atypical nuclei and perinuclear halos that are believed to be a manifestation of HPV infection. These cells are called koilocytes. The underlying stroma usually shows a mild inflammatory response. Biopsies of condylomatous lesions resistant to treatment or abnormal appearance should be taken for histological confirmation of the diagnosis. Differentiation between condyloma and vulvar papillomatosis or other vulvar lesions can then be made. Treatment consists of destroying the skin manifestations of HPV infection. Eradication of the virus from the epithelium is not clinically possible. Control of epithelial viral expression can be accomplished with topical agents such as trichloroacetic acid, bichloroacetic acid, podophyllin or its derivatives, or topical 5-fluorouracil (5-FU). Cryotherapy, laser vaporization, and electro-excision or desiccation are other methods of treatment. The intralesional or systemic administration of interferon is another treatment option usually reserved for recalcitrant lesions. Recurrent viral skin expressions

are usually managed with alternative treatments or combinations of treatments. Verrucous carcinoma of the vulva presents as a large condyloma or a lesion suspected of invasive carcinoma.

The lesion, first described by Buschke and Lowenstein as a giant condyloma², is associated with the HPV 6.1 viral subtype. Verrucous carcinoma can involve large areas of the vulva, and it has a pushing rather than an infiltrating border. Characteristically, it is localized to the vulva. Management is best achieved by wide excision and careful postoperative evaluation. Vulvar intraepithelial neoplasia is a hyperplastic squamous lesion with atypia confined to the epithelium. VIN is histologically divided into three categories: VIN I (mild dysplasia), VIN II (moderate dysplasia) and VIN III (severe dysplasia and carcinoma in situ). Lesions in VIN are sufficiently atypical to be considered precancerous. The incidence of progression to an invasive malignant process is relatively low (10 to 15%) and the time to progression can extend over several years. Bowen's disease, VIN III, and carcinoma in situ are clinically synonymous terms. These lesions appear as hyperkeratotic, raised and frequently pigmented epithelial thickenings (Fig. 4). They usually occur in women who are in the sixth decade or older; however, a younger age does not exclude the diagnosis. The biopsy results show full thickness epithelial atypia and mitotic activity (Fig. 5). Affected areas may be asymptomatic or excoriated by scratching. Any thickened, raised, or hyperkeratotic lesion seen on pelvic examination should be biopsied, regardless of associated symptoms. Vulvar lesions can be multifocal; therefore, multiple biopsies are suggested. Improvement of lesions with dyes such as toluidine blue has been recommended to address the multifocal nature of these lesions. Toluidine blue should stain areas of nuclear concentration associated with VIN III; however, most lesions are hyperkeratotic and the keratin surface prevents penetration of the dye into the epithelial nuclei. A more effective approach is to apply a dilute acetic acid solution to the vulva. Areas of expression of VIN and HPV turn whitish after soaking for several minutes.⁴ The vulva can then be examined with a magnifying glass or colposcope. In this way, subtle epithelial changes are more easily identified. Bowen's disease or VIN III should be treated by surgical, laser, or electrical excision.⁴ It is best to remove the lesion with a technique that provides tissue for further histological study. The use of 5-FU or systemic interferon has had marginal adherence and

success. Surgical excision with margin assessment remains the preferred management option. Close follow-up to assess recurrence is suggested. Bowenoid papilloma has a clinical and pathological presentation similar to that of VIN III. It occurs in patients in the second, third and fourth decades of life. It manifests as multiple papillomatous lesions that may involve more than one area of the vulva. These lesions are associated with HPV infections.

The results of the biopsy show epithelial atypia with koilocytotic changes in HPV. Progression to an invasive malignant tumor occurs in rare cases, such as in immunocompromised or immunosuppressed patients. Management is conservative unless precancerous changes are present. Precancerous lesions are treated in a manner similar to VIN.