Preneoplastic Lesions of the Vulva

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The diagnostic effectiveness of colposcopy and cytology in investigation of the vulvar skin or mucosa is modest compared to that in investigation of the cervix. Examination with the naked eye provides a great deal of information. Removal of material for cytological examination is rendered difficult because the lesions may be covered by a keratin layer and because secondary inflammatory alterations are common. For this reason, precisely directed tissue sampling for histological investigation of suspicious lesions or carcinoma is still the most important diagnostic measure to be carried out on the vulva.

Formerly, vulvar dystrophies were regarded as precancers. Today, the different forms of dystrophy (lichen sclerosus, hyperplastic dystrophy, mixed form) are classified separately from the precancerous epithelial atypias in the new nomenclature of vulvar diseases. Today, precancers of the vulva are designated as ‘intraepithelial neoplasia of the vulva (VIN)’, which corresponds to cervical intraepithelial neoplasia. Diagnostic criteria exist for the different degrees of atypia: slight dysplasia (VIN I), moderately severe dysplasia (VIN II), severe dysplasia (VIN III), and carcinoma in situ (VIN III). These criteria are: firstly, a disturbance in the architecture of the squamous epithelium, and secondly, disorders of maturation and atypias of the individual cells. The separation of the individual degrees of atypia from each other is not always simple. In addition, the diagnosis is rendered more complicated by the distinction between basaloid and Bowenoid carcinoma in situ. In terms of differential diagnosis, reactive epithelial alterations have to be discriminated in cases of chronic inflammation. It is not clear whether Bowenoid papulosis is merely a reversible reactive alteration due to human papillomavirus infection or a precancer. The prognostic significance of slight and moderately severe dysplasia of the vulva is not known. The progression from untreated carcinoma in situ to invasive carcinoma occurs very much more rarely than in carcinoma in situ of the cervix. As far
as can be evaluated, the incidence of progression in the vulva is less than 10%. Morphologic findings or determinations of ploidy of the cell nuclei do not allow precise prognostic classification.

Etiologically, there are also indications of the involvement of herpesvirus hominis II and of human papillomavirus (types 16 and 18) in intraepithelial neoplasia of the vulva. Especially in younger patients, a flat condylomatous lesion or condylomata acuminata are observed in some cases in addition to intraepithelial neoplasia. It is so far unknown whether the detection of virus DNA or virus-induced antigens allows a prognostic appraisal.

For the diagnosis in patients with intraepithelial neoplasia of the vulva, two factors are important:

1. In about two-thirds of cases, the lesions occur multicentrically. It is not possible to rule out an invasive carcinoma on the basis of the macroscopic appearance alone. Multiple biopsies are thus indicated.
2. Roughly one quarter of the patients with intraepithelial neoplasia of the vulva also develop cervical epithelial neoplasia, and 10%-40% develop invasive carcinomas in the genital tract or in other localizations. A precise general medical examination is necessary.

Intraepithelial neoplasias of the squamous epithelium of the vulva, which have so far been rare in gynecological practice and mostly occur in old patients, are gaining greater practical significance, since they have been observed more frequently, especially in younger patients, in recent years. Part of this change is due to better medical surveillance. However, as in cervical intraepithelial neoplasia, a further proportion is due to the spread of herpesviruses and papillomaviruses even in developed countries with a high standard of hygiene.

Paget’s disease of the vulva can be subdivided histogenetically into two forms:

1. Infiltration of the epidermis by an invasive or in situ carcinoma of cutaneous appendages (mostly sweat-gland carcinoma, in about 20% of the cases of Paget’s disease).
2. Carcinoma in situ of the epidermis arising from ‘stem cells’ which differentiate in the direction of apocrine glands. It follows from this that the rate of progression of Paget’s disease cannot be appraised precisely. The development of an invasive carcinoma from an in situ lesion is even rarer than in the squamous epithelial precancers of the vulva.