VULVA VAGINA UTERUS
TUMORS OF FEMALE
SEX ORGANS

part 2 + part 2 supplement

HERTIG + GORE

TUMORS OF THE FEMALE SEX ORGANS

Part 2

TUMORS OF THE VULVA, VAGINA AND UTERUS

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and

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ARMED FORCES INSTITUTE OF PATHOLOGY

ATLAS OF TUMOR PATHOLOGY

Section IX—Fascicle 33

TUMORS OF THE FEMALE SEX ORGANS

Part 2

TUMORS OF THE VULVA, VAGINA AND UTERUS

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Hazel Gore

TUMORS OF THE VULVA, VAGINA, AND UTERUS

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TUMORS OF THE VULVA, VAGINA, AND UTERUS

VULVA

INTRODUCTION

The vulva is formed by the labia majora, labia minora, clitoris, mons pubis, and the associated structures of the vestibule including the urethral meatus. Although its component parts are mainly skin and subcutaneous fat with their usual appendages, the vulvar tissue contains mucus secreting and apocrine sweat glands, erectile tissue, wolffian duct remnants, and the insertion of the round ligament with its accompanying pelvic peritoneum. The vulvar tissue is comparable to the sex skin of the higher primates and is similarly subject to influence by the steroid hormones. It may, on occasion, contain accessory breast tissue. Because of this complicated morphologic and functional anatomy, the tumors of the vulva are of diverse origin and of varying degrees of malignancy.

BENIGN TUMORS OF EPITHELIAL ORIGIN

Papilloma

The papilloma is the only widely accepted benign neoplasm of epithelial origin. It is uncommon, usually single but may be multiple, and occurs anywhere on the vulva (fig. 2). Papillomas are usually small but may reach 4 to 5 cm. in diameter. They may occur at any age during adult life.

Papillomas are potentially malignant (3.2 percent for true papillomas, Novak) in contrast to the essential lack of malignant potential in **condyloma acuminatum** (fig. 1). The latter is not a true neoplasm but a hyperplasia. It is probably of viral origin and may arise or grow larger during pregnancy. Most authors (Taussig, 1940; Way, 1951) do not consider condyloma acuminatum a predisposing cause of vulvar carcinoma, whereas others (Charlewood and Shippel) have found cases where the cancer appears to have arisen from this papillary tumor-like lesion.

Simple but wide excision of a papilloma, provided careful examination of the adjacent skin shows no evidence of malignant change, is considered adequate therapy.

Pigmented papilloma may also occur on the vulva and is similar to such lesions elsewhere on the skin. Other terms for this lesion are acanthotic nevus, seborrheic keratosis, and senile wart.

PAPILLARY LESIONS ARISING FROM THE SQUAMOUS EPITHELIUM OF THE VULVA

Figure 1. Condyloma acuminatum of vulva. Note pointed or acuminate masses of keratinized epithelium covering a thick arborescent epidermis of papillary pattern. Although this lesion is not a true neoplasm, its pattern and frequently active growth simulate neoplasia. Such a lesion rarely becomes malignant. × 58. F.H.W.* S-50-3800; A.F.I.P. Acc. No. 218754-307.

Figure 2. A true papilloma of the vulva. Note thickened arborescent squamous epithelium with relative lack of keratinization. × 13. F.H.W. S-51-511; A.F.I.P. Acc. No. 218754-184.

Figure 3. Photomicrograph showing a squamous cell carcinoma of the vulva which may have arisen on the basis of a papilloma such as seen in figure 2. This also shows an arborescent papillary pattern without keratinization. × 125. F.H.W. S-49-4854; A.F.I.P. Acc. No. 298591-3.

*In this and following legends, F.H.W. stands for Free Hospital for Women, Brookline, Mass.

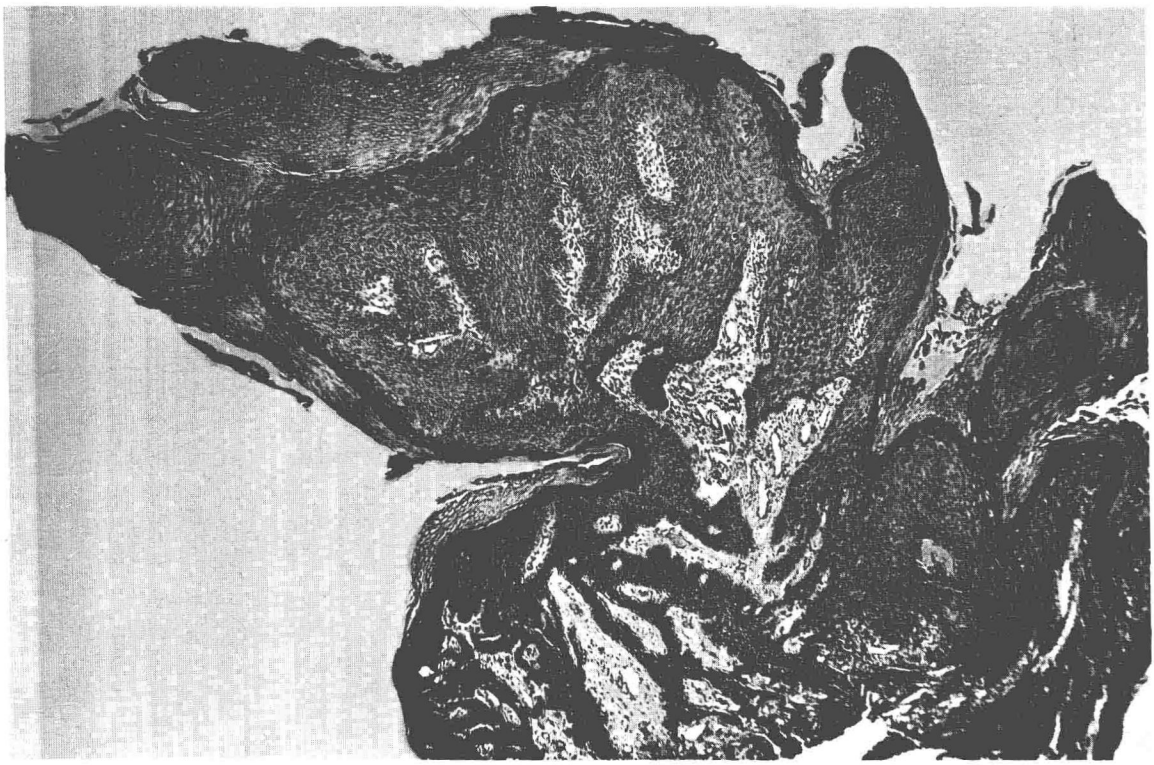

Fig. 1

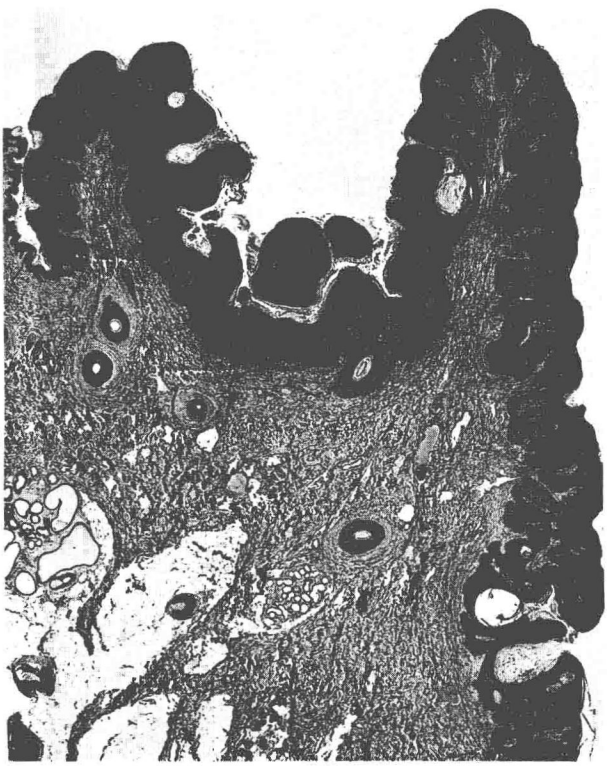

Fig. 2

Fig. 3

MALIGNANT TUMORS OF EPITHELIAL ORIGIN

Carcinoma in Situ

SYNONYMS AND RELATED TERMS: Bowen's disease; erythroplasia (erythroplakia) of Queyrat; extramammary Paget's disease; intraepidermal carcinoma.

All of these lesions occur on the vulva as well as elsewhere in the body. Willis believes that they are all variants of a single condition and states (p. 290): "For the pathologist there is only one entity, intra-epidermal carcinoma, which, like intra-duct carcinoma of the breast, may long remain confined within epithelial boundaries. Also like intra-duct carcinoma, intra-epidermal carcinoma shows many structural variations, but these do not denote distinct forms of tumour nor call for distinctive names."

Despite this point of view, it is well to describe Bowen's disease and extramammary Paget's disease separately. Bowen's disease appears to be a lesion purely of squamous epithelial origin, whereas extramammary Paget's disease appears to involve both squamous epithelium and apocrine sweat glands, sometimes resulting in a sweat gland carcinoma. The rare erythroplasia of Queyrat may well be a focal variant of Bowen's disease. It occurs on the glans and prepuce of the penis, on the vulva, or in the mouth.

Bowen's Disease

Bowen's disease, an epithelial lesion (pl. I–A; figs. 11, 12) described by Bowen in 1912, may appear anywhere on the skin of middle-aged or elderly patients of either sex ". . . as slightly raised reddish-brown papular or plaque-like lesions, with crusted or eroded surfaces. The patches are often multiple in the affected region; they slowly enlarge, and may coalesce to form extensive irregular areas. After a long period, usually many years, invasive squamous-cell carcinoma develops . . ." (Willis, p. 287).

It may be added, however, that removal of the crusts leaves a dull red, smooth, moist surface with focal pearly elevations (Knight). The gross description and the account of the clinical course are applicable to the rare case occurring on the vulva. It appears, however, that invasive carcinoma does not inevitably follow Bowen's disease, since only 2 of 100 cases of this disease on file at the Armed Forces Institute of Pathology developed invasive carcinoma, although some patients were followed for 15 years (Grady). According to Jeffcoate and associates, Bowen's disease of the vulva is apt to become an invasive carcinoma when it spreads to mucosal surfaces such as the vagina.

Knight found that Bowen's disease of the vulva occurs between the ages of 25 and 73 years with symptoms, usually pruritis, of three months' to 11 years' duration (average 4.6 years). He considered that Bowen's disease of the vulva is not as rare as has been thought. He found six cases in the files at the Sloane Hospital, and during this same period there were 12 cases of invasive vulvar carcinoma.

Willis described the microscopic structure (p. 288): "The epidermis is thickened and the interpapillary processes broad and blunt. The cells, especially of the spinous layer, show great irregularity of size and shape, including giant or multinucleated forms. Mitotic figures are plentiful, and many of the nuclei are hyperchromatic or distorted. Large rounded pale or vacuolated cells, resembling those of Paget's disease, are often present. The surface may or may not show excessive keratosis. The dermis shows chronic inflammatory changes and abundant lymphoid and plasma cells."

The treatment is by local vulvectomy, since radiation is ineffective (Jeffcoate et al.). The prognosis in patients treated surgically is potentially good, unless the process has become a truly invasive carcinoma or the lesion is too extensive to be removed completely.

Extramammary Paget's Disease

DEFINITION. A rare dermatosis limited to the skin of the axilla and anogenital regions of either sex, clinically and pathologically resembling Paget's disease of the breast, and characterized microscopically by the presence of large clear "Paget's cells." These cells are probably metastatic from an underlying carcinoma of apocrine sweat gland origin.

If this is a valid definition (Weiner; Stout; Parsons and Lohlein), it becomes obvious that many cases of so-called extramammary Paget's disease of the vulva only simulate this lesion. In reviews of the literature, many cases have been discarded (Weiner; Pinkus and Gould; Woodruff). Weiner accepted only 8 cases of which 4 had proven carcinoma, 1 an early carcinoma, and 3 apparently had no carcinoma. Woodruff found 21 cases in the literature and added 2 of his own but stated only 3 had underlying invasive carcinoma. Two fatal cases have been reported. In Weiner's case there was a typical clinicopathologic syndrome of Paget's disease of the vulva with an underlying apocrine sweat gland carcinoma from which the patient died. At autopsy, metastases were found in the regional, inguinal, pelvic, and periaortic nodes, and in the uterus, right tube and ovary, adrenal, liver, and two ribs near the vertebral column. The fatal case reported by Woodruff and Richardson was found at autopsy to have extended to uterus and tubes and to have metastasized to inguinal and retroperitoneal lymph nodes and to skin and peripancreatic tissue.

AGE INCIDENCE. Woodruff considers that this lesion occurs at a younger age than does vulvar carcinoma, whereas Weiner found an age range of 35 to 84 years with an average of 62.6 years. Symptoms had been present for periods ranging from several months to eight years.

GROSS. The lesion is more commonly found on the labia majora than on the labia minora (Woodruff). It is red, moist, and sharply demarcated with