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Vulval Carcinoma

1. GROSS DESCRIPTION

Specimen

— biopsy/partial/simple/radical vulvectomy/uni-/bilateral inguinal lymphadenectomy/pelvic exenteration.
— size (cm) and weight (g).
— vulval cancer forms 5% of gynaecological malignancies and occurs mainly in women aged 60–75 years. It can present as itch in an area of pallor or redness (leukoplakia/VIN) on a background of atrophic or hypertrophic lichen sclerosus. A nodular, verruciform or ulcerating mass may be present and a diagnostic wedge or punch biopsy taken with the former more likely to establish the presence of any invasive disease and the latter sufficient for abnormalities in a flat epithelium such as VIN. CT and MRI scan can be used to detect and stage inguinal lymphadenopathy, which may also be amenable to FNA cytology. Because of the strong HPV association, concurrent cervicovaginal disease is excluded. Surgical treatment is geared to the patient's age, fitness, tumour site and stage with wide local excision for "early" stage IA disease. Central and lateral stage IB lesions may be treated by partial vulvectomy and bilateral or ipsilateral groin node dissection, respectively. Stage II cancers and above need radical vulvectomy, which includes removal of the perianal skin and bilateral inguinal lymphadenectomy. Pre-operative radiotherapy may be given for tumours with extensive local invasion or involved nodes in an attempt to downstage, facilitate surgery and avoid pelvic exenteration.

Tumour

Site

— anterior/posterior.
— lateral (right/left).
— labia majora/labia minora/clitoris.
— labia majora is the commonest site, then labia minora and clitoris.
— bilateral (25%).

Size

— length × width × depth (cm) or maximum dimension (cm).

Appearance
— polypoid/verrucous/ulcerated/necrotic/satellite lesions/pigmented.
— 50% are ulcerated, 30% exophytic.

Edge
— circumscribed/irregular.

2. HISTOLOGICAL TYPE
Vulval carcinomas show the full range of cutaneous cancers.

Squamous cell carcinoma
— 80–90% of malignant vulval neoplasms.
— keratinizing or non-keratinizing and of two main types.
a. 60% of cases are in older women, not related to HPV and a keratinizing squamous cell carcinoma with adjacent epidermal hyperplasia/hyperkeratosis/differentiated VIN, or,
b. 30% of cases are in younger women, HPV 16/18 positive, of basaloid or warty histology and with adjacent VIN of undifferentiated or classic type (see 8. Other pathology for discussion).

variants:

— basaloid: 28% of cases at a younger age (<60 years) and association with HPV, cervical and vaginal lesions. Nests of basaloid cells with peripheral palisading, central focal keratinization and mitoses.
— warty: association with HPV and koilocytosis. Prognosis intermediate between usual squamous carcinoma and verrucous carcinoma. Distinguish from pseudoepitheliomatous hyperplasia overlying lichen sclerosus, Crohn's disease or granular cell tumour.
— adenoid: pseudoglandular/acantholytic.
— verrucous: exophytic with pushing deep margin of cytologically bland bulbous processes. Local recurrence after incomplete excision or radiotherapy.
— spindle cell: cytokeratin positive sarcomatoid carcinoma.

Basal cell carcinoma
— 20% local recurrence rate and metastases are rare.

Distinguish from basaloid squamous carcinoma, Merkel cell tumour and secondary small cell carcinoma.

Adenocarcinoma
— rare.
— appendage origin/Bartholin's gland/mesonephric duct remnants, or metastatic.

Paget's disease
— 2% of vulval malignancy.
— intraepithelial adenocarcinoma cells probably arising from basal layer multipotential cells differentiating along sweat gland lines. In