Vulvar masses: Imaging features with pathologic correlation

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Learning objectives

Describe the normal imaging features of the vulva
Differentiate and characterize various vulvar masses
Describe the revised International Federation of Gynecology and Obstetrics (FIGO) staging of vulvar cancer

Background

The vulva is located in the anterior urogenital triangle of female perineum. The vulva represents the female external genitalia, composed of the mons pubis, the clitoris, the labia majora, the labia minora and the vestibule. A wide array of diseases affect the female perineum in adults. In the majority of cases, clinical presentation and biopsy findings, if needed, define diagnosis. Imaging techniques are recommended when physical examination is limited, or when malignancy is suspected. Vulvar carcinoma is a rare malignancy, most commonly seen in elderly women. It represents 5-8% of all gynecologic malignancies. Survival depends on the lymph node status.

Findings and procedure details

1. Anatomy of the vulva
2. Diagnostic modalities
3. Diseases involving the vulva

1. ANATOMY OF VULVA

The vulva represents the female external genitalia. It is located in the anterior urogenital triangle of female perineum.

It appears as a triangular soft tissue structure within the perineum, bounded by the symphysis pubis anteriorly, the anal sphincter posteriorly and the ischial tuberosities laterally.

Arterial blood supply is via branches of the external and internal pudendal arteries, with lymphatic drainage into the superficial inguinal nodes, although the deep inguinal nodes
are also involved. In vulvar carcinoma, the largest percentage of sentinel nodes is found in the superficial medial nodes.

The vulva includes the mons pubis, the clitoris, the labia majora and minora, the vestibular bulb, vestibular glands, and the vestibule of the vagina (Fig. 1 on page 10, Fig. 2 on page 11, Fig. 3 on page 11, Fig. 4 on page 12).

The **mons pubis** is composed of adipose tissue that overlies the symphysis pubis and covered with hair. It contains skin appendages with sebaceous and sweat glands.

The **labia majora** form the lateral boundaries of the vulva; they are bilateral folds of skin with underlying fat and appendages running from the mons pubis and merging in the perineum.

The **labia minora** are found just medial to the labia majora. Anteriorly, labia minora divide into two parts. One part passes over the clitoris to form the prepuce, and the other part joins beneath the clitoris to form the frenulum of the clitoris. They contain no hair follicles.

The **clitoris** is ventral to the urethra and vagina. Its body projects into the fat of the mons pubis. It is composed of the body, paired crura and bilateral (vestibular) bulbs.

The **vestibule** is the part of the vulva lying between the two labia minora laterally and extends medially to the hymenal sulci.

The **Bartholin glands** are located in the vestibule on either side. They produce mucus and participate in lubrication during sexual intercourse. **Skene’s glands** are a pair of paraurethral glands. In addition, there are minor vestibular glands located around the vestibule.

By drawing an imaginary line between the two ischial tuberosities, the perineum is divided into two triangles:

- Anterior urogenital triangle, with the pubic symphysis being its apex.
- Posterior anal triangle with its apex at the coccyx.

*The anterior urogenital triangle* is divided into two parts, the superficial perineal pouch and the superficial perineal membrane.

1. The **perineal membrane** fuses superiorly to the fascia of the levator ani muscle and inferiorly to the muscle fibers of the superficial perineal pouch. It contains the deep transverse perinei muscle and the intrinsic muscles of the urethral sphincter mechanism.

2. The **superficial perineal pouch** lies inferior to the perineal membrane and is penetrated by the vagina and urethra and contains the clitoris. The superficial perineal pouch contains the superficial transverse perineal muscles, the ischiocavernosus muscles that
cover the crus of the clitoris, and the bulbospongiosus muscles that flank the vagina and urethra, covering the superficial parts of the vestibular bulbs and Bartholin glands (Fig. 5 on page 12).

The perineal body is a strong fibromuscular tissue located in the midline between the anal canal and the urogenital triangle.

2. **DIAGNOSTIC MODALITIES**

**Ultrasound**

- It is a cheap and available technique.
- Ultrasound has gained acceptance in the evaluation of soft tissue tumors (Fig. 6 on page 13).
- An advantage of ultrasound and fine needle biopsy is the easy evaluation of the inguinal-femoral nodes due to the superficial localization.
- However, it lacks sensitivity for ruling out lymph node involvement.

**CT**

- Performed with iv contrast administration.
- Usually obtained at 2.5 mm axial slice reconstructions with oral, and iv contrast.
- Rectal and vaginal contrast material are optional.
- Field of view should include proximal thighs to cover the entire extent of tumors.

**MRI** protocol includes:

- T1-weighted fast spin-echo imaging: all pelvis and inguinal region are included. They allow excellent differentiation between pelvic structures and surrounding pelvic fat. Axial T1 with fat suppression assists in the differentiation of proteinaceous or hemorrhagic material.
- T2-weighted imaging with small field of view (pelvic coil), axial, sagittal and coronal: These are preferred for anatomical evaluation.
- Diffusion weighted images.
- T1 FS post contrast: Contrast is not essential but can be useful for detection of recurrence.
- Coronal whole abdomen.

The vulva shows low to intermediate signal intensity on T1-weighted imaging and slightly higher signal intensity on T2-weighted imaging.

3. **DISEASES INVOLVING THE VULVA**
A wide variety of lesions occurs on the vulva. Most disorders of the vulva are readily diagnosed on the basis of clinical findings; the mainstay of diagnosis is vulvar biopsy. Imaging methods are indicated when physical examination is limited, particularly for the evaluation of deep located lesions or when malignancy is suspected, for staging. However, vulvar abnormalities can be present as incidental findings at US, CT and MRI, and can easily be overlooked, especially if the examination has been performed for nongynecologic indications.

Benign vulvar disorders include vulvar atrophy, benign tumors, hamartomas and cysts, infectious diseases and nonneoplastic epithelial disorders. Developmental abnormalities of vulva are rare. Vulvar atrophy is related to advanced age.

**Bartholin gland cyst**

- Caused by chronic inflammation or infection of the glands, resulting in ductal obstruction from pus or thick mucus.
- Located on either side of the posterior lateral inferior vagina medial to the labia minora and at or below the level of the pubic symphysis. They are reported to occur in 3% of adult women.
- Usually unilocular, 1-4 cm in size.
- Hyperintense on T2-weighted images with variable appearances on T1-weighted images depending on their proteinaceous or mucinous content. The cysts are well defined, thin walled and show no rim enhancement (Fig. 7 on page 14).

**Skene duct cyst**

- Retention cysts that forms secondarily to inflammatory obstruction of the paraurethral ducts (Skene ducts).
- Located lateral to the external urethral meatus and inferior to the pubic symphysis.
- Round or oval hyperintense lesions on T2-weighted images.
- May be difficult to distinguish between Skene duct cyst and Bartholin gland cysts due to their similar location. Skene duct cysts are centered more anteriorly than Bartholin gland cysts and closer to the external urethral meatus (Fig. 8 on page 14).

**Bartholinitis**

- Common condition, appearing with painful vulvar mass, that arises from secondary infection of a Bartholin cyst.
- Clinical diagnosis, but MRI can be used if there is doubt over the diagnosis or to assess complications.
- Appears as a thick walled cyst mass of variable signal intensity on T1-weighted images, high signal intensity on T2-weighted images, with associated high signal intensity in surrounding tissues due to edema. The
lesion shows irregular rim enhancement on postcontrast images (Fig. 9 on page 15).

Vascular

Varicose veins

• Varicose veins in the vulvar and perivulvar area are seen in 4% of women.
• Most of them are secondary to pregnancy and usually regress spontaneously.
• Vulvar varicose veins are rare in nonpregnant women. When present, they can be seen alone or associated with leg varices or venous malformations, including Klippel-Trenaunay-Weber syndrome. In some cases, vulvar varices are seen as part of the "pelvic congestion syndrome".
• Radiological evaluation of superficial lesions is not typically necessary, except in the case of extensive lesions (Fig. 10 on page 15).

Thrombophlebitis

• Can develop in pre-existing varicosities.
• Color Doppler ultrasonography is helpful in differentiating low flow states such as venous malformations from the high flow seen in arterial malformations, as well as demonstrating expansile intraluminal filling defect in noncompressible vein and the absence of flow in the case of thrombophlebitis (Fig. 11 on page 16).
• MRI provides a large field of view to evaluate the extent of the lesion. Acute thrombus show high signal intensity on T1-WI and T2-WI with progressive enhancement within an expanded vessel with perivascular inflammation.

Fistula

• Abnormal connection between anal canal and skin related to anal gland inflammation and obstruction. May extend to labia, vulva or vagina.
• Can be caused by other conditions such as Crohn's disease, tuberculosis, trauma, pelvic malignancy, or radiation therapy.
• MRI has emerged as the technique of choice for preoperative evaluation of perianal fistula (Fig. 12 on page 17). Postcontrast images are useful to differentiate a pus-filled active perianal fistula from a healing tract undergoing fibrosis.

Hematoma

• Nonobstetric accidental traumatic hematomas are relatively uncommon and are due to blunt injuries, particularly in the straddle position.
• Due to the rich vascular supply to the perineum, it may form a large hematoma when injured.
- Vulvar injuries include lacerations and hematomas (Fig 13).

**American Association for the Surgery of Trauma (AAST) organ injury scale for vulvar trauma**

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<td>II</td>
<td>Laceration, superficial (skin only)</td>
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<td>Laceration, deep into fat or muscle</td>
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<td>Avulsion; skin, fat or muscle</td>
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<tr>
<td>V</td>
<td>Injury into adjacent organs</td>
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</table>

**Fig. 13:** American Association for the Surgery of Trauma (AAST) organ injury scale for vulvar trauma

**References:** Unidad Central de Radiodiagnóstico - Madrid/ES

- Typically isolated to the labia, but extension to the perineum can be seen in as many as 20% of the patients.

- In blunt trauma, when associated vaginal injury is suspected, CT is the imaging technique of choice to evaluate the extent and assessment for assessment of active extravasation.

- Vulvar hematoma can occur as a result of obstetrical delivery, either spontaneously after vaginal delivery or after episiotomy (Fig. 14 on page 18).

**Benign tumors**

Benign tumors of the vulva are relatively uncommon and may show nonspecific findings, and therefore, a biopsy in often needed to exclude malignancy.

**Lipomas**

- Constitute the most common benign soft tissue tumors, but vulvar lipomas are a rare entity.
• Lipomas of the vulva are rare benign tumors that consist of mature fat cells often interspersed with strands of fibrous connective tissue. They arise from the vulvar fatty pads.
• Lipomas usually appears as soft, single or multiple painless, slowly growing subcutaneous masses (Fig. 15 on page 18).

**Vulvar malignancies**

• Rare disease accounting for 4% of all female genital tract cancers.
• Predominantly involves older women: 66% percent of cases occur over the age of 70.
• Over 85% are **squamous cell carcinoma**. Other common histologic types include adenocarcinoma, sarcoma, basal cell cancer and Paget's disease of the vulva, and Bartholin's gland carcinoma.

- **Squamous cell carcinoma:**
  • Slow growing, locally invasive and may extend to the vagina, urethra, anorectum, and rarely to the bladder. Rarely does it extend beyond the pelvis.
  • Nodal spread occurs to regional inguinal and femoral lymph nodes, while metastases to deep pelvic nodes such as the internal or external iliac nodes are considered as distant metastases. Figure 16 and figure 17 summarize the TNM system and 2009 revision to FIGO staging for vulvar cancer.

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**Definition of TNM-FIGO**

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<th>FIGO</th>
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<td>Carcinoma in situ (preinvasive carcinoma)</td>
</tr>
<tr>
<td>T1a</td>
<td>IA</td>
<td>Tumor confined to the vulva or perineum. 2 cm or less in size and with stromal invasion 1 mm or less</td>
</tr>
<tr>
<td>T1b</td>
<td>IB</td>
<td>More than 2 cm in size or more than 1 mm of stromal invasion.</td>
</tr>
<tr>
<td>T2*</td>
<td>II</td>
<td>Tumor of any size with extension to the adjacent perineal structures (lower/distal 1/3 urethra, lower/distal 1/3 vagina, anal involvement)</td>
</tr>
<tr>
<td>T3**</td>
<td>IVA</td>
<td>Tumor of any size with extension to any of the following: upper/proximal 2/3 of urethra, upper/proximal 2/3 of vagina, bladder mucosa, rectal mucosa, or fixed to pelvic bone</td>
</tr>
</tbody>
</table>

*FIGO uses classification T2/T3. This defined as T2 in TNM
**FIGO uses the classification T4. This is defined as T3 in TNM

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<tbody>
<tr>
<td>M0</td>
<td></td>
<td>No distant metastasis</td>
</tr>
<tr>
<td>M1</td>
<td>IVB</td>
<td>Distant metastasis (including pelvic lymph node metastases)</td>
</tr>
</tbody>
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<th>TNM</th>
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<td>N0</td>
<td></td>
<td>No regional lymph node metastasis</td>
</tr>
<tr>
<td>N1</td>
<td></td>
<td>One or two regional lymph nodes with the following features</td>
</tr>
<tr>
<td>N1a</td>
<td>IIA</td>
<td>One lymph node metastasis each 5 mm or less</td>
</tr>
<tr>
<td>N1b</td>
<td>IIA</td>
<td>One lymph node metastasis 5 mm or greater</td>
</tr>
<tr>
<td>N2</td>
<td>IIIB</td>
<td>Regional lymph node metastasis with the following features</td>
</tr>
<tr>
<td>N2a</td>
<td>IIIB</td>
<td>Three or more lymph node metastases each less than 5 mm</td>
</tr>
<tr>
<td>N2b</td>
<td>IIIB</td>
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<td>Extracapsular spread</td>
</tr>
<tr>
<td>N3</td>
<td>IVA</td>
<td>Fixed or ulcerated regional lymph node metastasis</td>
</tr>
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</table>

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**Fig. 16:** Definition of TNM-FIGO
Fig. 17: Vulvar carcinoma Staging FIGO 2009 vs TNM scale

References: Unidad Central de Radiodiagnóstico - Madrid/ES

Prognosis

- Related to the size of the primary tumor, depth of invasion and most importantly lymph node involvement.
- 5-year survival in cases with negative lymph nodes is in excess of 80%. This falls to less than 50% if the inguinal nodes are involved and 10-15% if the iliac or other pelvic nodes are involved.
- While inguinal-femoral lymph node involvement is a significant prognostic factor, clinical palpation is unreliable for determining the presence of lymph node metastases.
- Currently there are not noninvasive imaging technique to exclude the presence of lymph node involvement with a sufficiently high NPV, and surgical evaluation is the standard method to evaluate groin lymph node metastasis. Unfortunately, significant morbidity is common with inguinal-femoral lymphadenopathy (wound breakdown, infection, lymphocele and long term risks of lymphedema and cellulitis).
- Using diagnostic imaging methods, such as CT, PET/CT and MRI, detection and assessment of groin lymph nodes have been developing over the past few years with the ultimate intention of reducing groin node surgery in node-negative patients. MR lymphography and MRI diffusion-weighted imaging may also help in detection of metastatic nodes and are being investigated.

MR imaging
• Useful for evaluation of the extent of more locally advanced cancer to determine the relationship to adjacent structures, and to aid in surgical planning (Fig. 18 on page 20, Fig. 19 on page 21, Fig. 20 on page 21). It can also evaluate lymph node involvement. Using the short/long axis diameter ratio >=0,75%, the sensitivity of MRI is close to 87%, with a specificity of 81% for the detection of metastatic nodes in the groin. Other criteria include irregular contour, cystic changes, and loss of fatty hilium.

*CT imaging*

• Not helpful in local staging of the tumor because of its low soft tissue contrast resolution.
• However, CT can assess the involvement of the bladder or the rectum and detect distant metastases.

**Aggressive angiomyxoma of the vulva**

• Slow-growing mesenchymal tumor.
• Usually in premenopausal women.
• Appears as a soft, cyst-like or polypoid lesion.
• The majority of tumors are greater than 10 cm in size.
• Appears as a solid infiltrative mass that tends to displace rather than invade adjacent structures.
• There is a tendency for local recurrence if not completely excised (Fig. 21 on page 22).

**Metastases**

• Uncommon, most of them occur by contiguous spread (Fig. 22 on page 23).
• Distant metastases occur via lymphatic or hematogenous spread. More than 50% of metastases to vulva have gynecologic primaries, most commonly from uterine cervix, followed by ovary, endometrium and vaginal origin. Non gynecologic primaries are most commonly from gastrointestinal tract, followed by breast, melanoma, lymphoma, bladder and kidney carcinomas.
• MRI characteristics may mimic primary tumors.

Images for this section:
1. Urethra
2. Vagina
3. Rectum
4. Pubis
5. Ischium
6. Pectineus m
7. Obturator externus m
8. Obturator internus m
9. Puborectalis m

**Fig. 1:** Axial T2-weighted MR image of the normal anatomy of the perineum
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1. Urethra
2. Vagina
3. Anal sphincter
4. Crus of the clitoris
5. Body of the clitoris
6. Labia majora
7. Obturator externus m
8. Perineal body
9. Puborectalis m

**Fig. 2:** Axial T2-weighted MR image of the normal anatomy of the perineum
© Unidad Central de Radiodiagnóstico - Madrid/ES
1. Bladder
2. Uterus
3. Iliac bone
4. Crus of the clitoris
5. Body of the clitoris
6. Labia majora
7. Obturator internus m
8. Obturator externus m
9. Femoral head

**Fig. 3:** Coronal T2-weighted MR image of the normal anatomy of the perineum

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1. Bladder
2. Uterus
3. Iliac bone
4. Vagina
5. Anterior recess of ischiorectal fossa
6. Labia majora
7. Obturator internus m
8. Obturator externus m
9. Femoral head
10. Perineal membrane
11. Vestibular bulb/ Bulbospongiosus m
12. Crus of clitoris/ Ischiocavernosus m

**Fig. 4:** Coronal T2-weighted MR image of the normal anatomy of the perineum

© Unidad Central de Radiodiagnóstico - Madrid/ES
1. Vagina
2. Ischiocavernosus m
3. Crus of clitoris
4. Bartholin gland
5. Vestibular bulb
6. Bulbospongiosus m
7. Perineal membrane
8. Pudendal canal
9. Obturator internus m
10. Levator ani m
11. Clitoris
12. Superficial transverse perinei m
13. Mons pubis
14. Urethra
15. Perineal body
16. Anus

**Fig. 5:** Coronal-view (top) and axial-view (bottom) illustration of the muscles of the female perineum

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**Fig. 6:** Transperineal ultrasound

Clinical suspicion of Bartholin cyst
Ultrasound shows solid mass
Doppler US image shows vascularization in and around the heterogeneous soft tissue mass
Diagnosis: Leiomyosarcoma
Fig. 7: Bartholin gland cyst. MR images show a thin walled cystic lesion in the posterolateral aspect of the vulva region (arrow)

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Fig. 8: Location of the Skene ducts and Bartholin glands

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Fig. 9: Infected Bartholin gland cyst. CT images in two different women show rim-enhancing abscesses (arrow)

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Fig. 10: Varicose veins

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Fig. 11: Acute thrombophlebitis in a puerperal woman. Doppler ultrasound shows intraluminal filing defect and absence of flow. MR images show serpentine hyperintense vessels and surrounding hyperemia
**Fig. 12:** Perianal fistulous tract extending to the vulva. Axial contrast-enhanced fat-suppressed T1 weighted images show the fistulous tract (arrow)

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**American Association for the Surgery of Trauma (AAST) organ injury scale for vulvar trauma**

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**Fig. 14:** Large vulvar hematoma after episiotomy. Axial CT shows hyperdense lesion

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Fig. 15: 47 year old woman was evaluated for a slow-growing vulvar mass. Ultrasound revealed an encapsulated subcutaneous mass. Color Doppler imaging (not shown) did not disclose vascularization in the mass. Histological examination confirmed the presence of vulvar lipoma.

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**Table III: Vulvar carcinoma Staging FIGO 2009 vs TNM scale**

<table>
<thead>
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<th>T</th>
<th>N</th>
<th>M</th>
<th>Definition</th>
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<td>IA</td>
<td>T1a</td>
<td>N0</td>
<td>M0</td>
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</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>stromal invasion 1 mm or less. No nodes</td>
</tr>
<tr>
<td>IB</td>
<td>T1b</td>
<td>N0</td>
<td>M0</td>
<td>Confined to the vulva or vulva and perineum, more than 2 cm in size or</td>
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<td></td>
<td></td>
<td></td>
<td>more than 1 mm of stromal invasion. No nodes</td>
</tr>
<tr>
<td>II</td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
<td>Tumor with adjacent spread (1/3 lower urethra, 1/3 lower vagina, anus). No</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>nodes</td>
</tr>
<tr>
<td>IIIA</td>
<td>T1, T2</td>
<td>N1a, N1b</td>
<td>M0</td>
<td>Inguinofemoral nodes. 1 node with ≥5 mm metastasis or up to 2 nodes with</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>&lt;5 mm metastases</td>
</tr>
<tr>
<td>IIIB</td>
<td>T1, T2</td>
<td>N2a, N2b</td>
<td>M0</td>
<td>Inguinofemoral nodes. 2 or more nodes with ≥5 mm metastases or</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>3 or more nodes with &lt;5 mm metastases or up</td>
</tr>
<tr>
<td>IIIC</td>
<td>T1, T2</td>
<td>N2c</td>
<td>M0</td>
<td>Inguinofemoral nodes. Extracapsular spread</td>
</tr>
<tr>
<td>IVA</td>
<td>T1, T2, T3</td>
<td>N3, Any N</td>
<td>M0</td>
<td>Tumor spread to pelvic bone, bladder mucosa, rectal mucosa, 2/3 upper</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>part of the urethra or 2/3 upper part of vagina</td>
</tr>
<tr>
<td>IVB</td>
<td>Any T</td>
<td>Any N</td>
<td>M1</td>
<td>Any distant metastases (including pelvic nodes)</td>
</tr>
</tbody>
</table>

**Fig. 17:** Vulvar carcinoma Staging FIGO 2009 vs TNM scale

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**Fig. 18:** FIGO I. Contrast-enhanced T1-weighted image with fat suppression shows a small lesion in right labia majora, confined to the vulva. No lymph nodes

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**Fig. 19:** FIGO IB. Axial T1 weighted image (A) and coronal contrast enhanced fat-suppressed T1 weighted image (B) show a tumor in left labia majora, without adjacent spread. No lymph nodes

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Fig. 20: FIGO IVA. Coronal (A) and axial (B) T2 weighted images demonstrate a large vulvar carcinoma. Sagital contrast-enhanced T1-weighted image with fat suppression (C) shows mass in vulva infiltrating rectum. Axial T2 WI (D) shows enlarged inguinal lymph nodes

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**Fig. 21:** Aggressive angiomyxoma of the vulva: Coronal (A) and sagittal (B) T2-weighted images show local recurrence of aggressive angiomyxoma

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**Fig. 22:** Rectal carcinoma in a 57 years old woman, with contiguous spread to the vulva. (A) (B) axial T2 weighted images, (C) axial T2 weighted image with fat suppression.

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Conclusion

A wide spectrum of diseases arise within the vulvar area.

Knowledge of the diseases and spatial anatomy allows the radiologist to confidently identify the site of origin and extent of disease with the appropriate selection of imaging modality.

MR imaging is the modality of choice for evaluation of the extent of complex lesion and its relationship to adjacent structures and is helpful to aid in eventual surgical planning.

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