

Introduction

In 2000, approximately 77,500 women will be diagnosed with an invasive gynecologic malignancy and several hundred thousand will be diagnosed with a preinvasive or potentially malignant condition of the reproductive tract. The methods of detection of these women's cancers, as well as their ensuing treatment, are as diverse as the health care systems in which they receive care.

Primary care physicians and specialists, both medical and surgical, are in a position to diagnose and provide some level of care to these patients. Many of these same clinicians will at some point need to refer a patient for additional, specialized care by a gynecologic oncologist. These referral guidelines based on available clinical data were developed to provide various pathways that promote timely, high-quality, and cost-effective care that maximizes patients' opportunity for full recovery. In 1971, the American Board of Obstetrics and Gynecology recognized the importance and necessity of subspecialty training and certification for those physicians involved in the treatment of women with reproductive tract malignancies. After completing a 4-year residency in obstetrics and gynecology, gynecologic oncologists must currently complete an additional 3 years of subspecialty fellowship training in an approved program that encompasses both medical and surgical evaluation and management of the woman with a diagnosis or a suspected diagnosis of gynecologic cancer.

Fellows who complete this training and become subspecialty board certified by the American Board of Obstetrics and Gynecology, Division of Gynecologic Oncology must demonstrate an understanding of:

- The molecular, immunologic, genetic, and environmental aspects of cancer etiology.
 - The role, effect, and benefit of cancer screening.
 - The importance and most beneficial implementation of diagnostic studies.
 - The appropriate utilization of surgery, radiation therapy, and chemotherapy alone or in combination to effect cancer treatment.

Surgical training and subspecialty board certification are based on developing a proficiency in all aspects of surgery, including nonradical and radical pelvic operations, reconstructive procedures, gastrointestinal surgery, urinary tract operations, and retroperitoneal dissection. Special emphasis is placed on the following skills:

- Evaluating operative candidacy.
- Selecting the appropriate procedure or surgical route.
- Minimizing the incidence and managing all perioperative and other treatment complications.

Board certification in gynecologic oncology requires training in the pathologic evaluation of the histologic and microscopic findings that can limit undertreatment and/or overtreatment while optimizing outcomes. In addition, gynecologic oncologists must achieve expertise in directing and administering chemotherapy as well as the management of related toxicities and complications. Finally, to be board certified, the gynecologic oncologist must have the theoretical and technical expertise to recommend adjuvant, therapeutic, or palliative radiation therapy.

It is in the best interest of all parties concerned—patients, caregivers, insurers, and institutions—that women with cancer of the reproductive tract are accurately diagnosed and appropriately managed for the duration of their illness. Appropriate treatment and surveillance can often make the difference between mere survival and a return to good health. Women are entitled to make an informed decision regarding their care for a potentially malignant, premalignant, or malignant condition. While many “pathways” of care may be appropriate, successful management of gynecologic cancer most often correlates with the incorporation of those important aspects of care derived from the growing body of medical evidence and surgical expertise.

Gynecologic oncologists are the physicians most experienced in the nuances of reproductive tract cancer surgery and are experienced in the selection and sequencing of treatment modalities likely to benefit an individual patient while minimizing the hazards associated with undertreatment (failure to control cancer) and overtreatment (avoidable expense and complications). These referral guidelines provide direction so that clinicians can best avail themselves of the support and expertise provided by those specialists dedicated exclusively to the treatment of gynecologic cancer. They are endorsed by the Society of Gynecologic Oncologists whose purpose as stated in their bylaws is (1) to improve the care of patients with gynecologic cancer, (2) to advance knowledge and raise standards of practice in gynecologic oncology within the discipline of obstetrics and gynecology, (3) to encourage research in gynecologic oncology, and (4) to cooperate with other individuals and organizations interested in oncology and related fields.

Endometrial Cancer

Adenocarcinoma of the endometrium is the most common genital cancer in women over 45 years of age in the United States. In the United States approximately 36,100 new cases are diagnosed yearly and 6,300 women die with this disease [1]. The annual mortality to incidence ratio has more than doubled during the past decade. The lifetime incidence rate is about 22 per 100,000. The lifetime risk is approximately 2.4%. The incidence of endometrial cancer in the United States is exceeded only by breast, colorectal, and lung cancers. Adenocarcinoma of the endometrium is considered a disease of postmenopausal women. However, 30% of cases occur prior to menopause, and 5% occur prior to age 40.

The presenting, earliest clinical symptom in over 90% of cases is abnormal or postmenopausal vaginal bleeding. Appropriate evaluation leads to 70% of endometrial cancers being diagnosed while the lesion is clinically confined to the uterus. Delays in diagnosis can occur, related to patient failure to report early symptoms or when symptoms either are unrecognized or are not appropriately evaluated by the health care provider.

Endometrial cancer has been a surgically staged disease since 1988 [2]. Once the diagnosis of adenocarcinoma of the endometrium is established, primary treatment requires surgical removal of the uterus, both tubes and ovaries, and a thorough investigation to establish the presence or absence of extrauterine spread. There is no clinical evidence that the routine use of preoperative radiologic imaging studies contributes to improved survival, although their use increases costs [3–5]. Complete staging includes removal and histologic evaluation of pelvic and periaortic lymph nodes and intra-abdominal cytology. Exact surgical staging will detect the presence of extrauterine disease in 28% of patients who are thought to have disease clinically confined to the uterus [3, 6, 7]. This risk is increased with specific high risk histologic subtypes [8, 9].

Prompt, adequate therapy typically produces excellent results. When cancer is confined to the uterus after comprehensive surgical staging, the cure rate is in excess of 85%, and it is unlikely that the patient will obtain a survival benefit from adjuvant treatment [7, 10–12]. Patients with occult or visible extrauterine disease may benefit from additional therapy and can become long-term survivors [13, 14]. Unfortunately, if extrauterine disease is unrecognized, the chance of cure is dismal. Cancer cure is a more likely result when adequate therapy encompasses all sites of disease.

- Patients with a primary diagnosis of endometrial cancer or with recurrent disease could benefit from pretreatment consultation with or evaluation by a gynecologic oncologist to assist

in determining the most appropriate surgical approach as well as extent of surgery and the potential benefit of adjuvant therapy.

- Removal of regional lymph nodes may provide a therapeutic advantage in all grades and stages of endometrial cancer [15, 16]. Gynecologic oncologists are well trained in the techniques of a complete surgical staging procedure for endometrial cancer. When performed by appropriately trained surgeons, complete surgical staging can be performed without significantly increasing patient morbidity [4, 7, 17, 18]. The incorporation of multiple surgeons may fragment care and can increase costs without adding value to patient care. Individual therapy based on the pathologic results obtained from careful intraoperative surgical staging provides an improved risk/benefit ratio by avoiding overtreatment and undertreatment of individual patients [7, 19–21].

- Gynecologic oncologists are well trained in the translation of histologic and surgical findings into clinical care.

- Inappropriate evaluation of histologic findings can result in suboptimal, potentially morbid, costly treatment decisions. Despite these facts, gynecologic oncologists are only involved in the care of 40% of women with this disease [4]. Their presence is associated with clinically and statistically increased (2.5×) chance of complete staging [4]. Stage for stage survival is much better with surgical staging than with clinical staging [9].

- The physical characteristics of typical women with endometrial cancer render them prone to perioperative complications. The majority have significant existing medical co-morbidity which increases operative risk [7, 22]. Women with a diagnosed uterine cancer and abnormal cervical cytology are more likely to harbor advanced-stage endometrial disease [23, 24].

- There is no justification for the routine use of preoperative radiation. Extensive preoperative testing offers the patient little clinical benefit [3, 5]. Randomized prospective studies have not identified a survival advantage for the addition of adjuvant external radiation in the unstaged (no histologic retroperitoneal evaluation) patient with apparently confined uterine cancer [25]. Despite the absence of documented benefit, recent information suggests that 47% of patients with Stage Ia and 68% of patients with Stage Ib disease receive adjuvant radiation [9]. In completely surgically staged patients, in the absence of proven extrauterine spread or adverse histologic cell type, the *survival* advantage for adjunctive teletherapy has not been proven [2, 7, 10, 12, 26, 27]. When compared to hysterectomy and staging, its use increases health care costs by 30% [27a, 27b] and increases the risk of morbidity [19–21].

- Although low-grade, minimally invasive uterine adenocarcinoma may not require extensive lymphadenectomy, this

decision is best made intraoperatively. Twenty percent of women with presurgical low-grade malignant disease (G₁) will have their histology upgraded with final postoperative evaluation, and 17% of those with G₁ disease will exhibit deeply invasive disease [9, 28]. Thus to “determine” the extent of procedure preoperatively is likely not in the patient’s best interest. Gynecologic oncologists are the best trained physicians to determine the extent of surgical dissection. Every patient with a diagnosed uterine cancer should be offered the opportunity to be operated upon in a situation where staging is immediately available.

- Occult extrauterine spread can easily go unrecognized with incomplete surgical staging. Palpation or excision of enlarged lymph nodes alone is inadequate because only 10% of involved nodes are palpable and the majority of lymph node metastases are ≤ 2 cm [29]. Recognition of occult extrauterine spread allows directed adjuvant therapy and an opportunity for cure [13, 14]. If unrecognized, occult disease portends a dismal prognosis.

- In patients with extensive extrauterine disease, appropriate cytoreductive surgery may improve survival [30, 31]. Gynecologic oncologists are specifically trained to evaluate the benefit and perform this procedure. In this clinical situation the incorporation of multiple surgeons (gynecologist and general surgeon) increases costs without increasing patient benefits. In patients with documented extrauterine disease, the directed addition of radiation or chemotherapy offers significant survival advantage [13, 14].

- If deemed necessary, women diagnosed with an unsuspected endometrial cancer following hysterectomy can be surgically staged with minimal risks [32]. In this clinical situation, the pathologic results better direct additional therapy or allow for no further therapy.

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Pelvic Mass

Surgical evaluation of a pelvic mass is a common indication for a gynecologic operation [1, 2]. In addition to alleviating symptoms attributable to benign ovarian lesions, many of these operations are performed to determine the presence of a malignancy and to complete appropriate surgical treatment of an ovarian cancer if present. Board-certified gynecologic oncologists are currently the best trained subspecialists to complete the operative management of malignant, potentially malignant, or suspected malignant conditions of the female genital tract. Existing clinical data led to the recent NIH consensus panel opinion suggesting that preoperative consultation with a gynecologic oncologist should be offered to all women with a suspected ovarian malignancy [3]. Consultation or referral is clinically important, as an optimal surgical effort exerts a favorable effect on overall response and survival of patients diagnosed with ovarian cancer. Initial incorporation of a gynecologic oncologist into the management schema lessens the need for multiple surgeon involvement or the need for a second cytoreductive operative procedure and should result in cost-effective patient management.

- Transvaginal ultrasound is generally indicated in evaluation of a pelvic mass and is the most efficient, accurate, and least expensive of the imaging modalities [2, 4, 5]. A number of malignant risk indices have been developed, with accuracy related to ultrasonographic experience [6]. Computed tomography or MRI is not routinely indicated in the diagnostic evaluation of adnexal masses, and in general, the use of extensive imaging increases cost without adding value [2, 4]. Young patients with large complex or solid masses should have laboratory evaluation of available tumor markers (CA-125) to detect possible epithelial malignancy and germ cell cancers (hCG, α -fetoprotein, LDH). Perimenopausal and postmenopausal patients with a pelvic mass should have CA-125 testing, although a normal CA-125 does not eliminate the possibility of cancer, particularly early-stage disease [7–10]. Clinical examination by a gynecologic oncologist may lessen the need, cost, and morbidity of additional endoscopic or radiologic evaluation.

- Patients with masses that are clinically suspicious for cancer (see below) should be offered the opportunity of a preoperative consultation with a gynecologic oncologist [3]. Women should receive realistic preoperative explanations of their cancer risk and understand the potential extent of the surgical procedure, including the risks and benefits of a gastrointestinal or genitourinary operation.

- No one benefits when patients undergo inappropriate or incomplete procedures or when patients are not offered appropriate pretreatment referral/consultation. In most in-

stances, initial operation by a gynecologic oncologist should obviate the morbidity and cost of reoperation when an unstaged or less than appropriately cytoreduced malignancy is diagnosed [11].

- While statistical differences exist, most agree that specific clinical situations suggest a higher risk of malignancy and referral or consultation with a gynecologic oncologist may be beneficial to women in the high-risk situations when:

- Evidence of advanced disease is present: pelvic mass with omental caking; presence of effusion, ascites.

- A clinically suspicious pelvic mass [large (>10 cm) complex, fixed, nodular, bilateral] is diagnosed.

- Premenarchal girls require surgical treatment for a pelvic mass.

- Postmenopausal women have suspicious ovarian masses or elevated tumor markers.

- Perimenopausal women have ovarian masses, particularly when associated with elevated CA-125. Elevations between 35 and 65 U/ml are associated with a cancer risk of 50 to 60% [7–9]. A CA-125 >65 U/ml in a 50-year-old or older woman is virtually diagnostic of malignancy with a specificity of 98% [8].

- Young patients have a pelvic mass and elevated tumor markers (CA-125, AFP, hCG).

- Suspicious findings are present on imaging studies. The risk of malignancy in a postmenopausal woman with a unilocular mass without solid components is <1% [12, 13], increasing to 8% in a multilocular mass and 70% in a mass with solid components [5].

- Complex masses with solid components or excrescences or otherwise suspicious for cancer are present.

- Suspicious pelvic masses are found in women with a significant *family or personal history* of ovarian, breast, or other cancers (one or more first-degree relatives).

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Ovarian Cancer

Approximately 23,100 new cases of ovarian cancer are diagnosed annually, and 15,000 deaths are attributable to this disease. Ovarian cancer is therefore the leading cause of death from gynecologic malignancies in the United States and the fifth leading cause of death from cancer in women [1]. Recent reports indicate that only 9% of patients with early-stage ovarian cancer are treated appropriately according to NIH-recommended surgery and chemotherapy [2]. Only 71% of women with Stage III and 51% of those with Stage IV disease receive recommended surgery and chemotherapy [2].

The mainstay for successful treatment of early ovarian cancer, representing approximately 20 to 30% of ovarian malignancies, includes comprehensive surgical staging followed by appropriate adjuvant chemotherapy. Comprehensive surgical evaluation of early-stage disease is completed routinely by gynecologic oncologists (97%), less frequently by gynecologists (52%), and uncommonly by general surgeons (36%) [3, 4]. Retroperitoneal evaluation is completed three times more commonly when a gynecologic oncologist is involved [5]. In patients with advanced disease, efforts at optimal surgical cytoreduction should routinely precede chemotherapy. Gynecologic oncologists are the *only* board-certified subspecialists whose training encompasses all aspects of ovarian cancer treatment, including specific training in surgical staging and cytoreductive techniques, as well as specific surgical procedures for treatment of complications; selection, administration, and management of chemotherapy and its complications; appropriate selection of patients for further operative, medical, radiation, or palliative management. Gynecologic oncologists are well trained to counsel or refer patients for genetic counseling regarding familial/genetic cancer syndromes and individual cancer risks.

Our understanding of the molecular and epidemiologic aspects of both sporadic and familial ovarian cancers has evolved considerably over the past decade. Ovarian cancer has become an important focus of national agencies (NIH, NCI), and patients have been urged to consider participation in ongoing protocols in an effort to improve treatments for this disease.

- Current recommended therapy in patients with epithelial ovarian cancers is stage related. Appropriately staged patients with:

- Stage IA, Grade I tumors require no further treatment.

- Stage I disease may be candidates for an abbreviated chemotherapy treatment schedule. In patients within this subgroup, accurate, complete, comprehensive staging potentially reduces treatment cost and morbidity.

- Stage II–IV disease are typically treated with 6 cycles of systemic chemotherapy following attempts at optimal cytoreduction [6, 7].

- The diagnosis of low malignant potential disease require surgical resection or cytoreduction and are unlikely to benefit from additional chemotherapy.

Acceptance of specific surgical and chemotherapeutic intervention has resulted in improved survival of patient with Stage I, III, and IV ovarian cancer [8].

- A recent NIH Consensus Panel opinion states [9]: “Adequate and complete surgical intervention is mandatory primary therapy for ovarian carcinoma, permitting precise staging, accurate diagnosis, and optimal cytoreduction.” This report also indicates that all women with a suspected ovarian cancer should be offered a preoperative consultation with a gynecologic oncologist.

- Cytoreduction may offer a survival advantage even in those women with Stage IV disease [10]. By virtue of their comprehensive training, gynecologic oncologists are the appropriate health care providers uniquely suited to provide primary longitudinal care for ovarian cancer patients and to head disease management teams involved in the care of these patients. Successful treatment of those with advanced disease requires an appropriate initial operation. These comprehensive staging procedures and attempts at extensive cytoreductive surgery are more likely complete when performed by a fellowship trained gynecologic oncologist [11].

- Patterns of care studies demonstrate improved outcomes and more complete surgical procedures when surgery for ovarian cancer is performed by gynecologic oncologists than when it is performed by other surgical subspecialists [3, 4, 5, 7, 11, 12]. In patients who do not undergo appropriate initial surgical staging, studies indicate that reoperation results in upstaging of 20 to 30% of patients thought to have “early” disease [13, 14]. Successful, cost-effective adjuvant treatment depends on accurate staging. The best survival in women with advanced disease is demonstrated in numerous studies following primary optimal cytoreduction; therefore, initial operation provides the most important opportunity to affect survival [6, 8, 14]. This operation is routinely best performed by a gynecologic oncologist [5]. When performed by others, it is less likely to achieve optimal results and is associated with higher morbidity and increased use of colostomy [11]. In patients diagnosed with an unstaged malignancy, reoperation with its attendant cost and morbidity may be indicated. Reoperation by a gynecologic oncologist allows optimal cytoreduction in 70% of cases where the tumor was previously thought to be unresectable [16]. Gynecologic oncologists represent the subspecialty most likely to completely surgically stage the disease process primarily or after referral.

- Involvement of gynecologic oncologists in cases of women with early-stage disease *may assist in preservation of*

fertility, if appropriate and desired, particularly when a germ cell or borderline tumor is present [15].

- In patients optimally cytoreduced, reoperation is unnecessary. In patients not optimally cytoreduced, interval cytoreduction should be considered [15a]. Secondary cytoreduction may be beneficial in some patients with recurrent disease [15a, 17] even after evidence of initial chemo resistance [18]. Longitudinal follow-up with a gynecologic oncologist should help identify those patients who are candidates for additional surgery, chemotherapy, or investigational therapy.

- Theoretically, consistent longitudinal care with an ongoing doctor–patient relationship during diagnosis, treatment, and surveillance should lessen miscommunication and misinformation, improve patient satisfaction, and lessen redundant unnecessary testing. Gynecologic oncologists can function as the ideal “gatekeepers” for women with ovarian cancer.

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Cervical Cancer

Despite the availability of Pap smear screening, almost 12,800 women in the United States will be diagnosed with invasive cervical cancer each year. Nearly 5000 women die of their disease [1]. The recent results of prospective randomized trials conclude that the appropriate use of surgery, radiation, and chemotherapy, alone or in combination, leads to improved survival rates [2–7].

The proper clinical stage and actual anatomic extent of cervical cancer must be determined, as they dictate correct therapy. Confirmatory biopsy is needed in conjunction with an expert pelvic examination. Additional studies such as cystoscopy, proctosigmoidoscopy, chest X-ray, intravenous pyelogram CT scan, or MRI may also be needed. Many of these costly diagnostic studies may not be necessary in specific clinical situations. Definitive therapy is prescribed based on clinical staging. Although controversial, pretreatment surgical staging may allow alteration in treatment schema or field to improve outcome [7–9a].

Gynecologic oncologists provide surgical expertise for the treatment of primary, advanced, or recurrent cervix cancer. There are no other surgical specialists dedicated to the treatment and cure of invasive cervix cancer; however, only 64% of women with cervix cancer are surgically treated by a gynecologic oncologist [10].

- Women are likely to benefit from pretreatment evaluation by a gynecologic oncologist if they have:

- A suspicious visible growth on the cervix.
- A Pap smear report suggesting invasive carcinoma.
- A biopsy report confirming invasive carcinoma.

- Radical or modified radical hysterectomy and lymph node dissection are potentially curative for women with early-stage cervical cancer [11]. Gynecologic oncologists are the surgical subspecialists best trained to determine the role and extent of surgery.

- In selected cases, fertility-sparing procedures, including cervical conization or radical trachelectomy, may be performed [12]. Management of the pregnant woman diagnosed with cervix cancer can frequently be successfully managed without pregnancy interruption [11].

- In specific clinical situations adjunctive treatment including radiation or chemotherapy following radical surgery improves outcome [5].

- Determination of disease extent is essential to successful treatment. In some situations, pretreatment surgical staging allows a more rational therapeutic plan, although overall sur-

vival benefit and morbidity are debated [8, 9]. A combined approach using chemotherapy and radiation improves outcome in those women with advanced disease [2–4].

- Patients with central pelvic radiorecurrent disease can be cured with exenterative therapy [11]. This procedure can incorporate reconstructive procedures that not only result in cure but also potentially improve quality of life [13].

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Vaginal Cancer

Primary vaginal cancer, a malignant lesion confined to the vagina without involvement of the cervix or vulva, constitutes 1 to 2% of all female genital malignancies. Historically (1950s), vaginal cancer was considered incurable. However, radiation therapy with or without surgery can cure even advanced disease [1]. Unfortunately, treatment may be associated with significant physical and psychosexual morbidity.

The median age at diagnosis is 60 years (range 18 to 95). Squamous cell carcinoma is the most common histologic subtype. Primary adenocarcinoma is rare but has been associated with *in utero* DES exposure [2a]. Vaginal intraepithelial neoplasia (VAIN) is a possible precursor [2]. Human papilloma virus infection has been postulated to play a role in the pathogenesis of vaginal cancer [5]. As many as 30% of patients with primary vaginal cancer have a previous history of *in situ* or invasive cervical cancer.

The majority of women with vaginal cancer present with vaginal bleeding and discharge. Pain, bladder, or rectal symptoms can occur. As many as 27% of patients are asymptomatic and the diagnosis is made by Pap smear and physical examination.

Treatment decisions depend on the size and location of the malignancy. Maintenance or creation of a functioning vagina and preservation of ovarian function in premenopausal women are important factors to consider during treatment planning [3, 4].

- Gynecologic oncologists are the subspecialists specifically trained to care for women with this disease. They have the appropriate surgical training for radical and ultraradical surgical management and are trained and understand the role, benefit, and risks of radiation therapy with or without concomitant chemotherapy.

- Preinvasive vaginal disease can be among the most challenging problems to diagnose and treat. Most gynecologic oncologists have expert training in colposcopy and management of these difficult lesions.

- Primary surgical therapy is typically limited to those women with Stage I disease involving the upper posterior vagina. A radical or modified radical hysterectomy, upper vaginectomy, and pelvic lymphadenectomy are typically indicated for patients with a uterus *in situ*. If a patient has previously undergone a hysterectomy, a radical upper vaginectomy and pelvic lymphadenectomy should be adequate therapy [7].

- Radiation therapy, often combined with chemotherapy, is the initial therapy of choice for most patients. Brachytherapy may be used alone for small lesions and is combined with teletherapy to control or cure larger lesions [8].

- Vulvectomy, inguinal node dissection, with vaginectomy for tumors in the distal vagina, and anterior exenteration for Stage I and II tumors involving the anterior wall have been

described as appropriate therapy. Vaginal reconstruction after pelvic exenteration should always be considered.

- Premenopausal patients who require radiation therapy may benefit from pretreatment laparotomy or laparoscopy to transpose ovaries and resect enlarged lymph nodes.

- Pelvic exenteration represents a curable option for patients with radiorecurrent or centrally recurrent pelvic disease.

- Loss of vaginal function is the most frequent adverse sequela of therapy. Although rare, rectovaginal and vesicovaginal fistulas are the most frequently reported serious complication of therapy. Sexual dysfunction is common. The risk of vaginal stenosis may be decreased by vaginal dilatation. All women should have intensive pre- and posttherapy counseling to assist them in dealing with the lifelong consequences of therapy.

- Women with the following may benefit by pretreatment evaluation by a gynecologic oncologist:

- Women at high risk of vaginal neoplasia with abnormal pap smears (DES-exposed women, immunosuppressed women, history of lower genital tract disease) [5, 6].

- Women with unexplained abnormal vaginal cytology.

- Women with high-grade VAIN lesions (suspicious of invasion) who require vaginectomy.

- All women with invasive vaginal cancer.

- Women treated for vaginal cancer who need long-term follow-up, sexual counseling, management of vaginal stenosis.

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Vulvar Cancer

Vulvar malignancies are rare and account for 3 to 5% of all female genital cancers, with an annual incidence of 1.5 cases per 100,000 women. Squamous cell carcinomas constitute 86% of all cases [1].

Risk factors include tobacco use, human papilloma virus infection, vulvar intraepithelial neoplasia (VIN), and immunosuppression. The average age at diagnosis is 65 years; however, there is a bimodal age incidence that is thought to reflect distinct epidemiologies. Younger women (average age of 45 years) present with HPV-associated and multifocal lesions in a preinvasive background. Solitary vulvar cancers associated with vulvar dystrophies are seen in the older women (average age of 75 years) [2, 3].

Fewer than 1% of women are asymptomatic. The most common initial complaint is a vulvar mass or lump. Other common symptoms include pruritis, pain, burning, bleeding, dysuria, and discharge. Vulvar cancers and intraepithelial neoplasia are frequently misdiagnosed. Delay in diagnosis may be related to patient embarrassment, denial, and reluctance to be examined [3a]. Additional delay may be secondary to the common tendency of health care practitioners to prescribe topical medications to a patient with vulvar complaints without performing a physical examination and appropriate biopsy. Average delay from onset of symptoms to diagnosis approaches 1 year.

Prompt diagnosis allows curative surgical therapy. Survival is correlated with the presence of lymph node involvement. The incidence of lymph vascular spread is directly related to the size of tumor and the depth of invasion.

- Because successful treatment may involve multimodality therapy, gynecologic oncologists are best suited to direct the care of patients with vulvar cancer.

- The standard therapy for tumors localized to the vulva includes radical surgical resection of the primary lesion and inguinal lymphadenectomy [4, 5]. This procedure, related to disease extent, should be individualized and may incorporate wide local excision, hemivulvectomy, or vulvectomy. Inadequate local surgical excision results in high local failure rates [4]. Radical excision with bilateral groin node dissection has been the recommended treatment for larger central vulvar lesions. When vulvar cancers are diagnosed earlier, smaller localized, less radical surgeries with unilateral lymph node dissection can be performed.

- The consequence of inadequate surgical therapy is a recurrent incurable lesion [4]. However, the consequences of curative surgery can be psychologically devastating, as vulvar surgery can result in lifelong anatomic alterations. Sexual dysfunction is common because of loss of the clitoris in some clinical situations, and in general because of alteration in body image [6, 7]. Lower extremity lymphedema can occur, causing difficulty in walking, pain, recurrent infections, and disfigurement. Therefore, all ther-

apy should include pretreatment counseling about sexual and physical function.

- Reconstructive surgery with skin grafts, flaps, and pelvic floor repair is frequently necessary for the adequate surgical management of large vulvar lesions.

- Postoperative groin and pelvic nodal radiation is usually recommended in the presence of inguinal lymph node metastasis [8, 8a].

- Neoadjuvant chemotherapy prior to surgery or radiation has been advocated for Stage IVa lesions [9–11].

- Women with the following diagnoses may benefit from evaluation by a gynecologic oncologist:

- Those with any suspicious vulvar lesion, including non-healing ulcers, areas of chronic pain or pruritis, areas of pigment change, and mass lesions.

- Women with multifocal, complex, and/or recurrent high-grade preinvasive vulvar lesions.

- Women with suspected or diagnosed Paget's disease of the vulva.

- Women with invasive vulvar cancer should be referred for treatment and long-term follow-up to manage the consequences of therapy (sexual dysfunction, urinary dysfunction, lymphedema) and to screen for recurrence and for new lower genital tract lesions.

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Gestational Trophoblastic Neoplasia

Gestational trophoblastic neoplasia (GTN), the term commonly applied to choriocarcinoma and related tumors, represents a spectrum of premalignant or malignant placental aberrations that occurs in 1/600 therapeutic abortions and 1/2000 pregnancies in the United States. The risk of recurrence is 1/76 pregnancies following one occurrence and 1/6.5 pregnancies following a second event [1]. Malignant sequelae are increased with recurrent molar pregnancies.

Although previously a lethal disease, it is considered today the most curable gynecologic cancer [2]. This progress can be attributed to an available marker (hCG), chemo sensitivity, and the incorporation of aggressive multimodality therapy. The risk of mortality is increased with failure to diagnose the disease in a timely manner [3].

- Gynecologic oncologists are specifically trained in all aspects of diagnosis, surgical management, evaluation, treatment, and surveillance of women with GTN. Most authors agree that patients with trophoblastic disease should be managed by those with special expertise [4, 5].

- The clinical diagnosis of molar pregnancy is confirmed by ultrasound and appropriate serum studies. The histologic diagnosis of molar pregnancy or choriocarcinoma is usually confirmed with uterine evacuation. Unlike a routine dilatation and curettage, uterine evacuation of a molar pregnancy can be associated with life-threatening pulmonary, cardiovascular, endocrine, and uterine complications [3a]. Complete evacuation may lessen risks of other sequelae [6].

- Accurate histologic evaluation and surveillance following uterine evacuation of a molar pregnancy minimize the risk of misdiagnosis and increase the opportunity for the early diagnosis of persistent trophoblastic disease (PTD), a potentially lethal process that occurs in 15% of patients following uterine evacuation.

- Once persistent GTN is confirmed, appropriate evaluation allots this process into nonmetastatic or metastatic with a low-risk or high-risk clinical category. FIGO or WHO staging allows appropriate therapy [7]. Failure to do so increases the chance of over- or undertreatment, both of which increase the chance of poor outcome and health care costs.

- Cure is expected in 100% of patients with nonmetastatic trophoblastic disease with preservation of fertility in virtually all patients [6].

- Appropriate chemotherapy and surgery result in excellent survival (approaching 100%), with fertility being maintained in

the majority (80%) of women [6] with persistent gestational trophoblastic disease.

- Care of those with poor-prognosis metastatic disease requires multiagent chemotherapy, often combined with multimodality therapy. Reproductive tract and other surgical procedures may be essential to effect cure. Remission can be expected in as many as 90%. Failure is associated with extensive disease (late diagnosis) and inadequate initial therapy [2].

- Careful surveillance is necessary following treatment, as recurrences are noted in 2.1% (nonmetastatic), 5.4% (metastatic, good prognosis), and 21% (metastatic, poor prognosis) of those treated. Recurrence risk in Stage I is 2.9%, 8.3% in Stage II, 4.2% in Stage III, and 9.1% in Stage IV [7].

- Routine consultation or evaluation by a gynecologic oncologist may benefit women with:

- An ultrasound diagnosis of molar pregnancy.
- A histologic diagnosis of molar pregnancy.
- A diagnosis of persistent trophoblastic disease (low or high risk).
- A diagnosis of choriocarcinoma.
- A diagnosis of placental site trophoblastic tumor.

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