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EDITORIAL

Primary vaginal malignancy - a rare but significant challenge to the gynaecologists

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Vaginal malignancy though not uncommon, most of the cases (84%) it is found to be a secondary deposit from a primary malignant lesion from other area (viz. uterus, ovary) or a direct extension from a lesion in neighboring areas (viz, cervix or vulva). Primary vaginal carcinoma as such is a rare entity constituting 1-2% of all genital malignancies and only 10% of all vaginal malignancies [1]. Recent studies show an increase in incidence of primary vaginal carcinoma in young women in places with high prevalence of HIV infection with persistent high risk HPV types.

It has also been observed that less than 50% of primary vaginal carcinoma is diagnosed at stage I to give a better prognosis after treatment. On the other hand, due to the tendency of multifocal origin, it may co-exist with some other type of genital malignancy (viz. carcinoma cervix or vulval cancer). Studies also suggest that 30% of women suffering from vaginal carcinoma have a positive history of carcinoma cervix treated successfully within previous 5 years [2]. With the gradual increase of life expectancy these types of genital malignancies are increasing in incidence. All these facts demand further attention of the gynaecologist to detect these types of malignant lesions (so far considered rare and unimportant) at the earliest stage while it is still amenable for proper treatment giving rise.

Types

Histologically there can be different types as follows: Squamous Carcinoma- 84-90%; adenocarcinoma (including DES related) - 4-9%; sarcoma (including Botryoids sarcoma & leiomyosarcoma) - 2-3%; melanoma- 1-2%; others (eg. endodermal sinus tumor, transitional cell neoplasm). Malignancy can also arise in a neovagina created by split thickness skin graft by Mc Indoes technique. Squamous Carcinoma of vagina is discovered in 10-15 % cases after finding squamous cancer in other parts of the genital tract, e.g. cervix or vulva.

Risk factors

Human Papilloma Virus (HPV) is probably responsible for the majority of vaginal carcinomas. Of course, intrauterine exposure to Diethyl Stilboesterol (DES) is considered important aetiological factor for clear cell adenocarcinoma in young women (with a risk of 1 in 1000).

Spread

Direct spread can occur to the adjacent structures depending on the site of origin. Blood spread is very uncommon, can occur to distant organs in rare occasions. Lymphatic spread is of paramount importance. It depends on the site of origin. The lesion in upper third of vagina spreads through the lymphatic pathways of cervix, while the lesions in the lower third follow the lymphatic pathways of the vulva (involving in 6%-7% cases the inguinofemoral glands). Lesions of the middle third spread by following both the upper and lower lymphatics.

Diagnosis

Usually in early stage there may not be any symptom. Gradually vaginal bleeding or foul smelling vaginal discharge appears. On examination early lesions are likely to be missed unless suspected or meticulously examined. In most of the cases exophytic lesions or ulcerative lesions may be noticed which spreads very rapidly along the vaginal length and breadth. Exfoliative cytology (for screening purpose) may be positive. In invasive cases biopsy from the affected area is confirmatory. For screening of high risk or suspected cases, vaginoscopy (with the help of colposcope) is ideal. In selected cases staining with Acetic Acid and

Lugol's iodine may be done to demarcate the pathological area. Four quadrant vaginal smears from the walls of the vagina may be helpful to diagnose early dysplastic lesions (upto the accuracy of 88-90%). While examining the vagina, the speculum should be rotated so that the entire canal can be inspected and no lesion is left obscured. Staging is mostly clinical. Of course other tests are necessary for evaluation before embarking on the management proper. Besides routine biomedical assessments, CECT whole abdomen, pelvic MRI (also PET CT) may be helpful to assess the extension of the disease. Cystoscopy, proctoscopy, or colonoscopy may be necessary in selected cases. Chest X - Ray or CT thorax is necessary. Though not accepted universally, Sentinel node detection in vaginal carcinoma is done by using 99mTc labeled monocolloids in primary or recurrent vaginal carcinoma in some advanced centres [3].

Prevention

Attempt to prevent vaginal carcinoma should always be a concern for the Gynaecologist by all the three different ways: Primary Prevention HPV-16 subtype is found to be associated with the development of HSIL and primary vaginal carcinoma [4]. HPV vaccination has proved to be useful in preventing these particular types of lesions. Long term trend analysis shows promising estimates of reduction in HPV associated cases of vaginal cancer in future years among HPV vaccinated communities[5]. Secondary prevention - Routine screening for vaginal cancer after hysterectomy for benign diseases is not recommended. Routine screening is mandatory following hysterectomy for persistent HSIL. In these cases exfoliative cytology and HPV testing may be combined to get good results. Tertiary prevention - LSIL cases with HPV of non high risk serotype may be treated conservatively. HSIL cases needs treatment, as risk of progressing of these lesions to invasive state is significant (2-12%)[6].

Treatment

Treatment modalities are as follows: I) Laser ablation by CO2 laser - ideal for multifocal lesions. II) Surgical excision - mostly preferred for unifocal lesion. It is advantageous for obtaining tissue for histopathological study. III) Topical treatment with Imiquimod or 5- Fluorouracil (5 FU) is not very effective. Used for those who refuse other modes of treatment. Treatment of invasive vaginal cancer: Surgery is useful in stage I cases only. For lesions in upper 1/3 of vagina radical hysterectomy with bilateral pelvic lymphadenectomy and vaginectomy is ideal. In selected cases paraaortic lymphadenectomy is required. For lesions in lower 1/3 of vagina vulvectomy with inguinofemoral lymphadenectomy and vaginectomy is necessary. Lesion involving midline or both side of vagina needs bilateral lymphadenectomy, otherwise ipsilateral only. For lesions involving middle part of vagina, radical hysterectomy with bilateral pelvic lymphadenectomy, vaginectomy and in selected cases inguinofemoral lymphadenectomy is done. Ideally 1cm cancer free margin should be kept while excising. Less than 0.8 cm cancer free space is considered inadequate and necessitates postoperative radiotherapy. Adenocarcinoma, melanoma and sarcoma etc also needs similar surgical treatment depending on the site and extension. Squamous cell carcinoma responds better to radiotherapy. Stage II and above - all lesions are needed radiotherapy as primary treatment. Chemotherapy may be combined with radiotherapy to give better result. For recurrent cases, in selected cases total pelvic exenteration (for central recurrences) or anterior exenteration (for anteriorly extended lesions) may be done. For stage IV lesions with rectal involvement or RVF or with VVF may be treated with primary pelvic exenteration, pelvic and paraaortic lymphadenectomy, low rectal anastomosis, continent urinary diversion and vaginal reconstruction. For radiotherapy, external beam radiotherapy (EBRT) combined with brachytherapy gives better results. In most conditions, chemotherapy is also used simultaneously. In rare conditions multidrug chemotherapy regimen may be used as a primary treatment.

Personal experience

During a period of 36 years (from 1987 to 2023) the author has witnessed only 4 (four) cases of primary vaginal carcinoma. First 2 cases at stage II and stage III were treated by chemoradiation. One case was suffering from uterine prolapse and had a history of using vaginal pessary for many years. 2nd case was detected in the post operative period following a PFR operation. 3rd case was in stage I and treated surgically by radical hysterectomy and bilateral pelvic lymphadenectomy and total vaginectomy. This case was followed upto 1 year while it was uneventful. First 2 cases were lost to follow up. The 4th and recent patient, a 75 years old lady came with watery and blood stained vaginal discharge with a friable cauliflower like growth in posterior vagina involving upper and middle third of vagina. She was treated with radical hysterectomy with bilateral pelvic lymphadenectomy and total vaginectomy. On histology, growth shows poorly differentiated squamous cell carcinoma with no lymph node involvement. As one point of cut margin

is found to be involved by cancer tissue and the growth is poorly differentiated squamous cell carcinoma, postoperative radiotherapy is planned very soon once the patient recovers completely from postoperative trauma.

Conclusion

Primary vaginal carcinoma is a rare malignancy but the incidence is going to increase day by day for obvious reasons. Prognosis is poor (20%) in stage III and IV. But if detected and treated early prognosis is good (92 % in stage I) [7]. It is preventable to a large extent by HPV vaccination. The premalignant conditions like vaginal intraepithelial neoplasia (VAIN) can be detected by methodical examination (screening) with the help of PAP staining, Colposcopy, visual inspection with lugol's iodine etc. Awareness amongst the gynaecologists and paramedical workers as well as the patients will probably save many valuable lives or help good number of women to lead comfortable life even after detection of the disease.

Conflict of interest: None. **Disclaimer:** Nil.

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