Vaginal primary malignant melanoma (VPMM) is a rare and very aggressive tumor [1,2]. It accounts for 0.3-0.8% of all malignant melanomas, 2-5% of female genital tract melanomas and less than 3% of all vaginal malignancies [1-3]. Only 250 cases have been reported in the English literature [1,2].

The estimated incidence of VPMM is 0.026/100,000 women per year [2,3]. There are no significant differences, between various racial or ethnic groups [3,4]. VPMM most commonly occurs in postmenopausal women [5-8]. The mean age at diagnosis is 57 years [5-8].

The precise pathogenesis of VPMM is relative unknown [9]. Probably arises from melanocytes located aberrantly in vaginal epithelium [9,10]. Those melanocytes can be found in the basal layer of vaginal epithelium in 3% of healthy women [11]. It is thought that active junctional changes are the initial stage in malignant melanoma development [12]. However, ultraviolet radiation is not the causal factor in VPMM [4].

Although VPMM may arise anywhere, it is more common in the lower one third (34%) and the anterior (38%) vaginal wall [2,5,6,10,13]. VPMM may be single or multiple, pigmented or nonpigmented [14]. Also, most of VPMMs are polyoid and ulcerated [9,14]. Moreover, nonpigmented VPMMs may have similar appearance with vaginal epithelial tumors [9,14].

The most common symptoms and signs are: vaginal bleeding (80%), vaginal discharge (25%), palpable vaginal mass (15%) and pain (10%) [5,6,8,9,15,16].

The most common histologic cell type of VPMM, is: epithelioid (55%) [7,9,14]. Other less common histologic cell types of VPMM, are: spindled (17%) and mixed (28%) [7,9,14]. FIGO staging system for vaginal cancer is inappropriate for VPMM, as it does not incorporate tumor size and regional lymph node status [2].

Although there are several treatment options for patients with VPMM, an appropriate and effective treatment protocol has not defined yet [7].

Surgery remains the primary treatment of choice in patients with VPMM [2,7]. The spectrum of surgery ranges from conservative (wide local excision) to radical (vaginectomy, pelvic exenteration) [2,7,15]. If wide local excision with clear margins is possible, the role of radical surgery remains unjustified [2,7]. If wide local excision is impossible, pelvic exenteration may be reasonable [2].

Lymph node dissection is not recommended in patients with VPMM, as the rate of lymph node metastasis is low [7]. Moreover the role of elective lymph node sampling in those patients remains controversival [2,7]. Although lymph node dissection has no survival benefits, it leads to significant morbidity [7,17]. Recently, sentinel lymph node biopsy has gained popularity [7,18].

Radiotherapy in patients with VPMM, includes external pelvic radiotherapy and/or brachytherapy. It can be applied as primary treatment for patients who are unable or unwilling to have surgery [2,7,19,20]. It can be applied preoperative, to reduce tumor size and enable a more conservative surgery [2,7,19,20]. Also it can be applied postoperative as adjuvant treatment for patients with tumor size ≥ 3 cm, incomplete tumor resection or pelvic metastases [2,7,8,19,20].

Especially in elderly patients with bad performance status and comorbidities, we can apply high dose rate brachytherapy (HDRB) with Ir [8,21,22]. It is well tolerated and associated with less side effects than external pelvic radiotherapy [8,22].

For many years chemotherapy with dacarbazine (DTIC) is the standard of care in patients with advanced stage cutaneous malignant melanoma (CMM) [23]. However, the role of chemotheraphy in patients with advanced stage VPMM, has not been established [24].

Immunotherapy with interferon (IFN) or interleukin-2 (IL-2) confers survival benefits in patients with VPMM at high risk for recurrence [16,25-27]. However, immunotherapy has significant toxicity [16,25-27]. IFN has been associated with the generation of autoantibodies and the induction of autoimmune disorders [28]. Also, immunotherapy has very low activity against metastatic or recurrent CMM [26,27]. Moreover, the combination of IFN and IL-2 is superior to IL-2 alone [29].

The combination of chemotherapy and immunotherapy (biochemotherapy) in patients with advanced stage CMM, associated with an increased response rate [27]. Although it clearly improves response rates, it has no survival benefits [27]. Moreover, biochemotherapy has significant toxicity [27]. However, the role of biochemotherapy in patients with advanced stage VPMM has not been established [30].

VPMM is a very aggressive tumor and most patients diagnosed at advanced stage [1,16,25,31]. The extensive vascular and lymphatic network of the vaginal mucosa, contribute to early tumor spread and metastasis development [5,7,15].

Despite treatment modality, 5-year survival ranges from 8.4-17.5% [1,5,7,10]. Tumor size (<3 cm) is the most important prognostic factor [5]. Tumor thickness is only a weak predictor of survival [5]. Many patients with VPMM have: local recurrences in the pelvis and distant metastases in the lungs, liver, bones and brain [5,9]. Most of patients with distant metastasis also have a concomitant local recurrence in the pelvis [5].

It is obvious that the prognosis of VPMM is very poor despite treatment modality, as most cases diagnosed at advanced stage [1,8,16,25]. Moreover its prognosis is much more unfavourable, compared with other vaginal malignancies and CMM [9]. Especially in patients with no clear surgical margins and tumor size ≥ 3 cm, needed postoperative adjuvant radiotherapy.

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