352 ADENOCARCINOMA OF GLANDULE BARTHOLIN FOLLOW UP FOR FIFTEEN YEARS: A CASE REPORT

[1]V Krsic*, [2]J Krsic, [1]B Jocic Pivac, [3]J Milojevic. [1]*GAK Narodni Front, Belgrade, Serbia*; [2]*Military Academy of Belgrade, Belgrade, Serbia*; [3]*General Hospital Lazarevac, Obgyn, Belgrade, Serbia*

10.1136/ijgc-2021-ESGO.619

Introduction/Background* Adenocarcinoma gland Bartholin's is a very rare tumor and there is no agreement of optimal treatment for this type of carcinoma.

It accounting 2-7% of all cancers of the vulva and less than 1% of all female genital malignancies. Basic features are expanding locally, slow grow, and gives unexpected distant metastasis.

Methodology We will show our case and how we treated women with adenocarcinoma glandule Bartholin for fifteen years. 60-year old woman was admitted in our hospital because of tumor mass region glandule Bartolini on the left side and woman complained of the elevated tumor marker carcinoembryonic antigen, CEA (16, 2) detected random.

Result(s)* We did local wide removal of the tumor. Hystopathology confirmed that this is a Bartolini's gland adenocarcinoma. The tumor was removed in its entirety with healty edge. CT, MRI of the pelvis were normal and CEA dropped in 1,6. After seven years of primary treatment she had tumor mass on the same place and elevated tumor marker CEA again.

We did hemivulvectomy with lymph node disection on the left side and radiotherapy with TD 50 Gy on the left part of vulva and in 25 session and TD 45 GY in 22 session on regio ingvini. After surgery tumor marker dropped in normal range.

After fifteen years of primary tretment patient had lung metastasis and brain metastasis and she lived for three months

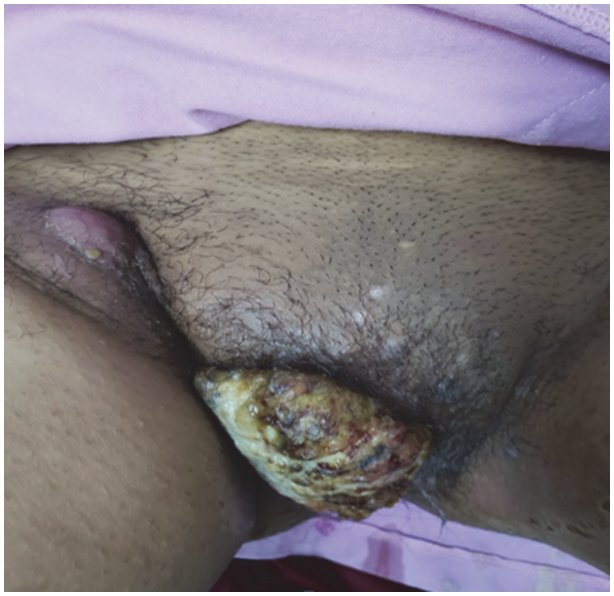
Conclusion* This case which we follwed for fifteen years give us many questions: was the first treatment with local wide excision optimal option, what was the best way to treat this patient and how to predict the way of spread of this malignancies and can we prevent metastasis ?

439 VULVAR CANCER OF RAPID PROGRESSION WITH EVOLUTION TO FAILURE TREATMENT IN PATIENT CARRYING FANCONI ANEMIA

[1]OF Neto*, [2]FM Lafraia, [2]LA Zorzanelli, [2]PEC De Cillo, [2]A Hiromi, [3]FK Tso, [2]MGB Kuster Uyeda. [1]*Escola Paulista de Medicina, Gynecological Oncology Division, São Paulo, Brazil*; [2]*Escola Paulista de Medicina, Gynecological Oncology Division, São Paulo, Brazil*; [3]*Escola Paulista de Medicina, Pathology of the Lower Genital Tract Divison, São Paulo, Brazil*

10.1136/ijgc-2021-ESGO.620

Introduction/Background* Fanconi Anemia is the most frequent cause of bone marrow failure genetically inherited. Patients may have short stature, microphthalmia, skeletal deformities, spots of coffee with milkand cardiac, renal and urinary malformations. One third of the cases are asymptomatic and the presence of pancytopenia is observed as an isolated manifestation.Carriers have 50 times greater predisposition for cancer of the head and neck, esophagus, gastrointestinal tract and vulva. Regarding to head and neck and gynecological squamous cell carcinoma, this incidence is up to 500 times higher than general population, and clinical presentation are more aggressive and disseminated at the time of diagnosis.



Abstract 439 Figure 1

Methodology Case report of a young female with Fanconi anemia who was diagnosed with squamous cell carcinoma of the vulva in advanced stage with unfavorable evolution.

Result(s)* Woman, 21 years old, nullipara, diagnosed with Fanconi anemia since 9 years old, started sexual activity at 18, with only 1 sexual partner since then. Admitted to the Gynecological emergency department with growth lesion 2 months ago, in the right labium majus. Physical examination reveals 8 cm lesion occupying the length of the right labium with a necrotic and friable surface associated with 4 cm ipsilateral inguinal lymph node enlargement. Incisional biopsy showed squamous cell carcinoma, moderately differentiated, with keratinizing foci and basaloid areas, ulcerated. Immunohistochemistry was positivity for p16 and p53 demonstrating effects of HPV infection. Cytology of the lymph node aspirate was positive for metastasis.MRI showed a vulvar lesion restricted to the superficial planes, but the presence of bilateral inguinal lymphondomegaly, at the right side next to common femoral vessels and pectinous muscle, stage IIIC.

Primary chemotherapy was chosen as the initial therapeutic approach to decrease the tumor volume and, subsequently, to allow a vulvar surgical procedure with palliative/hygienic intention. After the first cycle of chemotherapy (platinum-based), the patient had a fast-progressing Stevens Johns Syndrome, culminating in death.

Conclusion* Patients with Fanconi anemia should be screened more severe for gynecological cancer, especially after begining of sexual activity, which should includes Pap-Smear, genitoscopya and research for high-risk HPV. Precursor lesions should be treated vigorously as soon as they are diagnosed.

449 MOLECULAR LANDSCAPE OF VULVAR SQUAMOUS CELL CARICNOMA: REVIEW OF THE LITERATURE

[1]N Carreras Diéguez*, [2]JA Guerrero, [2,3]N Rakislova, [1,3]MT Rodrigo Calvo, [2,3]L Marimon, [2,3]M Ferrando, [1]A Torne, [1]M Del Pino, [2,3]J Ordi. [1]*Hospital Clínic de Barcelona, Gynaecologic Oncology Unit, Barcelona, Spain*; [2]*Hospital Clínic de Barcelona, Pathology, Barcelona, Spain*; [3]*ISGlobal, Barcelona, Spain*

10.1136/ijgc-2021-ESGO.621

Introduction/Background* Molecular landscape and carcinogenesis of vulvar squamous cell carcinoma (VSCC) have been poorly explored and lack a biomarker-driven targeted therapy. In this non-systematic review we aimed to summarize findings of studies exploring molecular landscape of VSCC.
Methodology Key word search was conducted (PubMed, Scopus) in January 2021, using the terms ("vulvar" and "cancer" or "carcinoma") and ("molecular" or "genomic" or "mutation"). Observational studies evaluating molecular alterations in VSCC were deemed eligible. Pre-specified data were extracted from the selected articles, including the number of samples analyzed, DNA sequencing technique, number and frequency of identified mutations, HPV prevalence and prognostic data.
Result(s)* Fourteen studies published between 2005 and 2020 were identified, including a total of 747 VSCC samples. Selected studies were highly heterogeneous in terms of DNA sequencing and HPV testing strategies and included small samples. Twelve studies performed next generation sequencing (NGS), nine of whom used targeted approach, two used whole exome sequencing and one used whole genome sequencing. The two remaining studies used multiplex ligation-dependent probe amplification assay and Sanger sequencing.

The most frequently studies and mutated genes were *TP53* and CDKN2A, followed by *PIK3CA*, *HRAS* and *PTEN* (table 1). Evidence on genomic differences between HPV-associated and -independent VSCC is particularly scarce and variable between studies. Only a single, targeted NGS study showed notorious differences in molecular profiles based on HPV status. Accumulated evidence indicates that in HPV-associated VSCC more frequently involves PI3K/AKT/mTOR pathway, involving *HRAS*, *KRAS*, *PIK3CA*, *KMT2D*, *PTEN* and *FBXW7* mutations. On the other hand, HPV-independent VSCC involve alterations in *TP53*, *CDKN2A*, *CCND1*. The prognostic role of molecular alterations in VSCC was assessed in seven articles, with discordant results. Some articles suggest that *TP53* alterations are associated to worse prognosis in patients with VSCC, particularly when combined with *PIK3CA*, *HRAS* or *CDKN2A*.
Conclusion* Limitations and heterogeneity of available molecular series contribute to a limited view of the molecular landscape of VSCC. Prognostic or therapeutic roles of identified mutations and pathways in VSCC remain to be elucidated. Large-scale, genome or exome-wide studies with robust HPV testing are necessary to expand the knowledge on molecular landscape in VSCC.

Abstract 449 Table 1 Overall frequency of most studied mutations in the included studies

Gene	N	Number of VSCC assessed	Overall frequency	Frequency range	Number of articles
TP53	387	711	54,43%	33-79%	13
PIK3CA	112	711	15,75%	0-34%	13
HRAS	60	677	8,86%	0-28%	11
CDKN2A	156	610	25,57%	6-36%	9
PTEN	26	646	4,02%	0-6%	9

471 VULVAR AND PERINEAL SURGERY : DOES FLAP REPAIR DEFECT STRATEGY ALLOW TO BALANCE FREE MARGIN AND POST-OPERATIVE MORBIDITY?

[1]M Andrault*, [2]N Kerfant, [3]C Rebahi, [3]P Merviel, [2]A Trimaille, [1]PF Dupre. *[1]CHRU Brest France, Breast and Gynaecological Oncology, Brest, France; [2]CHRU Brest, Plastic, Reconstructive and Aesthetic Surgery, Brest, France; [3]CHRU Brest France, Gynaecology, Obstetric and Reproductive Medicine, Brest, France*

10.1136/ijgc-2021-ESGO.622

Introduction/Background* Vulvar and perineal (VP) surgery has to combine surgical efficiency with free margin in an mechanical strained anatomical area with high vulnerability to infectious disease.

The size of excision as well as the anatomical restitution strategy justifies simple or complex associated reconstruction techniques..
Methodology All women who underwent VP surgery from November 2014 to August 2020 in our institution were included.

The cohort was divided into 3 groups : Groupe 0 (G0) : no reconstruction, Groupe 1 (G1) : fasciocutaneous flap, Groupe 3 (G3) pediculed or perforator flap.

Main objective was the margin status specified as follow : Safe Margins (SM) $\geq$ 8mm, Free Margins (FM) < 8mm and Involved Margins (IM).

Secondary objective were tumor and excision size, and postoperative morbidity.

Data was extract from Excel™ database. Quantitative variables were analyzed using the Chi-square test of Pearson.
Result(s)* Twenty-nine consecutive patients (29) were enrolled in the survey : 72,9% invasive disease (main pathological subtype Squamous Cell Carcinoma).

Twelve patients had a radical vulvectomy (41,4%) and 10 superficial vulvectomy (34,5%).

In group 1, V-Y flap was the most used flap (50%), Lotus Flap in group 2 (58,3%). The others flaps were DIEP, Gracilis, Taylor and rotative flap.

Twelve patients achieved SM (41%), 9 patients SM (31%) without any significant difference between the 3 groups (p= 0.68).

Among the 8 patients with IM (27%), 6 presented with Paget disease, no patient presented with invasive disease and IM in Group 2.

Median size of tumor was similar between the groups (2,5cm vs. 2,8cm, p = 0,76, but excision size seems to be superior in the group 2 (9,5cm vs 6.6cm, p = 0,09).

Clavien Dindo Grade 3 complications occurred in 11 cases (37,9%) and grade 2 in 5 cases (17,2%) without significant difference between G1 and G2.

Main complication was wound dehiscence (48,3%) and 3 patients had partial flap necrosis.
Conclusion* Vulvar and perineal surgery should include the use of a wide range of surgical repair options from local flap to complex reconstruction techniques in order to offer the best compromise between quality of the margins and postoperative morbidity.