Primary vulvar mucinous adenocarcinoma, a case report.

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INTRODUCTION:

Primary vulvar cancer is a rare gynaecologic malignancy, seen in about 2–3 per 100,000 women. The majority of cases (about 90%) are squamous cell carcinomas. Primary vulvar adenocarcinoma is much less common, with a very limited number of cases reported worldwide (as shown in the table below). It may originate from Bartholin's gland, sweat glands, Skene's gland, minor vestibular glands, aberrant mammary tissue or endometriotic implants.

CASE REPORT:

This is a report of a 60 years old female, G6P6, menopausal since 13 years and had her last delivery 30 years back. She was presented by a big right-sided labial soft tissue swelling, firm in consistency. She reported a rapid growing course during the last 3 months with development of skin ulcers recently. On examination, the mass was about 7x4 centimetres, at the centre of the right labius majus, firm cystic in consistency, not tender, and freely mobile above the under lying bone. The mass was attached to the overlying skin which showed 2 areas of ulceration (each about 2-3 cm in diameter) at the medial aspect of the swelling. Few small mobile superficial inguinal regional lymph nodes were felt (mostly reactive ones). Vagina and cervix were free. Imaging was performed (ultrasound pelvis), and no further pathologic findings were detected. The case was subjected to examination under anaesthesia and excisional biopsy.

Gross pathologic examination of the mass revealed a skin covered greyish-white lobulated mass. Histo-pathologically, the tumorous growth was divided into multiple compartments by thick fibrous tissue septa.

Each showed pools of mucin with free floating malignant epithelial cells forming glands, thin cords, cribriform islands and solid nests.

The final diagnosis was grade-2 vulvar adenocarcinoma. Lympho-vascular space was not invaded by the malignant growth. Resection margins were free of malignancy with narrow safety margin (1-3 mm).

Immuno-histochemical (IHC) staining for P63 was -ve. The tumour cells were also –ve for ER, PR, CA125, and CA19-9, thus excluding endometrial, ovarian, or gastro-intestinal origin of the disease. Thereafter, extensive metastatic work up was performed (serum tumour markers, chest, abdominal and pelvic CT scan, endometrial sampling, cervical PAP smear & colonoscopy). No other primary site for malignancy had been identified. So, the diagnosis was finally made as primary mucinous adenocarcinoma of the vulva, stage 1A.

The patient received adjuvant chemotherapy. She was disease free along the time of follow up; (11 months since November 2017 till the present time).

Author	Age (In years)	Treatment Modality
Tiltman et al. (1978)	50	Modified radical vulvectomy and LND.
Kennedy et al. (1993)	54	Radical vulvectomy and LND.
	63	Wide local excision.
Ghamande SA, et al. (1995)	80	Radical vulvectomy and bilateral inguino-femoral LND.
	67	Radical vulvectomy and bilateral inguino-femoral LND.
Rahilly MA et al. (1995)	69	Extended right hemi-vulvectomy
Graf AH et al. (1998)	75	Radical vulvectomy, bilateral inguino-femoral LND and postoperative radiotherapy
Willen et al. (1999)	57	Wide local excision.
Zaidi et al. (2001) ¹	43	Modified radical vulvectomy and LND.
Rodriguez et al. (2001)	64	Wide local excision.
Dube et al. (2004)	64	Wide local excision.
Cormio et al. (2012)	59	Radical vulvectomy and LND.
	42	Radical vulvectomy and LND.
Sui et al (2016)	43	Wide Local excision and postoperative chemotherapy
Lee et al (2017)	64	Wide Local excision.
Present case (2018)	60	Local excision and postoperative chemotherapy.

LND =
Lymph Node Dissection

References:

- ❑ Sui Y, et al. Primary mucinous adenocarcinoma of the vulva: a case report and review of the literature. Mol Clin Oncol. 2016;4:545–548.
- ❑ Lee IH, et al. Primary mucinous adenocarcinoma of the vulva, intestinal type. Obstet Gynecol Sci. 2017;60(4): 369–373.