

Recurrent Malignant Rhabdoid Tumor of the Vulva on Mons Pubis with Reconstruction of Profunda Artery Perforator Flap (PAP Flap) and Gracilis Muscle Flap (TUG) and Review of the Literature

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ABSTRACT

Extrarenal malignant rhabdoid tumor is a rare tumor with poor prognosis, only 16 vulvar cases have been reported so far in English literature. The “rhabdoid cells” features are composed by a diffuse proliferation of rounded or polygonal cells with eccentric nuclei, glassy eosinophilic cytoplasm containing hyaline-like inclusion bodies, arranged in nests or sheets, and prominent nucleoli.

The transverse upper gracilis (TUG) muscle flap combined with profunda artery perforator (PAP) flap has been described for breast and perineal reconstruction in defects that requires a large and substantial coverage.

Keywords: Transverse upper gracilis, Extrarenal malignant rhabdoid tumor, Hemi vulvar rhabdoid tumor

INTRODUCTION

Extrarenal malignant rhabdoid tumor is a rare tumor with poor prognosis, 16 vulvar cases have been reported so far in English literature [1].

The malignant rhabdoid tumor was described as a rhabdomyosarcomatoid variant of Wilms’s tumor [2]. The rhabdoid terminology was adopted due to the similarity of tumor cells with epithelioid sarcoma and differentiation between each other is difficult. The most common site has been reported such as hemivulva, the labia majora, mons pubis and combined labia majora and mons pubis [1,3].

The “rhabdoid cells” features are composed by a diffuse proliferation of rounded or polygonal cells with eccentric nuclei, glassy eosinophilic cytoplasm containing hyaline-like inclusion bodies, arranged in nests or sheets, and prominent nucleoli. These features are present in renal cell carcinoma and soft tissue sarcoma such as leiomyosarcomas, synovial sarcomas and extra skeletal myxoid chondrosarcomas [4].

The recurrence is common in a short period of time such as 2 months to 8 months.

We report a case the 15th case of right hemi vulvar rhabdoid tumor with reconstruction with profunda artery perforator flap (PAP flap) and three years later recurrence on mons

pubis with reconstruction with PAP flap combined with gracilis muscle.

CASE REPORT

44 years old woman whom 3 years before was operated for right malignant vulvar rhabdoid tumor with radical hemi vulvectomy and bilateral inguinal femoral lymph node resection. The initial lesion was a single nodular lesion, indurated, located in right labia majora with 3 cm diameter and irregular borders. The wide defect could not be closed primary and we decided to do a profunda artery perforator pedicle flap to cover the wound debulking and z-plasty procedures were performed months later after

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reconstruction. Three years later developed a recurrence of the tumor in hypogastric 3 cm length, nodular-like, bluish, indurated. The tumor was removed with 3.5 cm of margin around the lesion including the muscular fascia. The sample was sending to frozen pathology and reported sarcoma with rhabdoid feature with margins free.

The reconstruction plan was a pedicle Profunda artery perforator (PAP) flap with gracilis pedicle muscle flap (**Figures 1-3**) and mesh to cover the wound and drain was placed on the wound. The donor site was closed primary and drain was placed on the wound.

The microscopic outcomes in the initial tumor and the recurrence were compatible with malignant extrarenal rhabdoid tumor with prominent nucleoli, cytoplasm with hyaline-like inclusions and hyaline fibrosis bodies (**Figure 4a**), xanthogranulomatous inflammation with histiocyte and lymphocyte infiltration and high mitotic activity. INI-1 staining was not contributory. The histiocytes were positive to CD68 staining (**Figure 4b**). The lymph node section in the first surgery were negative for malignancy.



Figure 1. Gracilis Flap combined with PAP flap.

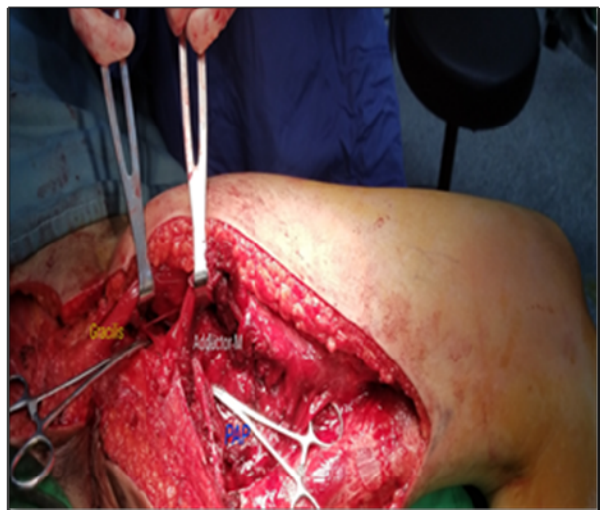


Figure 2. Dissection of the flap, on the top we observe the Gracilis muscle, and the pedicle of the Profunda Artery Perforator that runs through the Adductor Magnus.



Figure 3. Final inset of the flap.

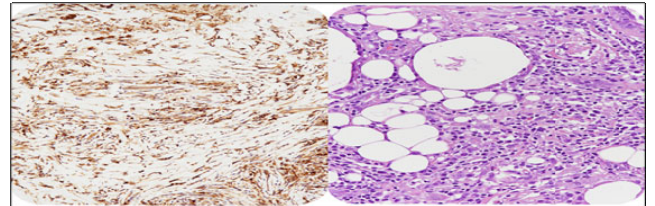


Figure 4. Focal stromal hyaline body fibrotic, with lymphocyte infiltration, and hemosiderin deposition.

DISCUSSION

The malignant rhabdoid tumor initially was described as a rhabdomyosarcoma variant of Wilms's tumor, and were described as extrarenal tumor described in soft tissue, urogenital tract, bowel, skin prostate, liver and central nervous system, then was recognized as an independent clinico-pathological entity [5,6]. The rhabdoid cells presence in a sample leads the diagnosis and the aggressivity of the tumor behavior [7]; most of the reported cases has been described as a painless firm-hard circumscribed mass of 2 to 10 cm diameter most of them with primary closure of the wound and inguinal lymphatic resection. Only one case was reported with reconstruction by pedicle rectus abdominis flap due to the large defect including the inguinal lymph node area resection (**Table 1**) [3]. The tumor usually express vitamin and cytokeratin, and do not retain the nuclear expression of INI-1 protein. The surgery is the first line of management with small margins usually 2 cm, there is no consensus regarding the margins neither the adjuvants therapy, and chemotherapy and radiotherapy has not shown be effective in controlling the recurrence, that is why we recommend extend the margins and closure the wound with pedicle flaps. The histopathological tumor has to include the exclusion of other sarcomas, melanomas and carcinomas due to has S100 marker and PAS marker positive in some cases [13]. The CD68 staining is a marker of histiocytes and histiocytic tumors.

The transverse upper gracilis (TUG) muscle flap combined with profunda artery perforator (PAP) flap has been described for breast and perineal reconstruction in defects that requires a large and substantial coverage [18]. The anatomical territories in the posteromedial area between the TUG-PAP gives the combination of two different major

angiosome nourishment the flap, the medial circumflex femoral artery territory (TUG) and profunda femoral artery territory (PAP) [19]. Usually, the TUG flap gives its main pedicle of the muscle at 6 to 8 cm along the muscle, and PAP offers 2 to 5 perforators along the posterior medial thigh and its proximity offers a perfect match with both flaps in defects that requires muscle and skin coverage.

Table 1. Reconstruction by pedicle rectus abdominis flap due to the large defect including the inguinal lymph node area resection.

Author, year	Age (years)	Site	Treatment	Lymphatic Resection	Reconstruction
Perrone [6]	19	R Labia Majora	Surgery	yes	Primary closure
	30	R Labia Majora		no	
	31	L vulva		yes	
Matias [8]	49	L Labia Majora	Surgery / Chemotherapy	No	Primary closure
Lupi [9]	39	L Labia Majora	Surgery / chemotherapy / radiotherapy	Yes (in secondary surgery)	Primary closure
Igarashi [10]	39	L Labia Majora	Surgery / Chemotherapy	No	Not reported
Sert [11]	44	R Labia Majora	Surgery / chemotherapy / radiotherapy	Yes	Primary closure
Brand [5]	40	Mons's pubis	Surgery	Yes	Primary closure
Haidopoulos [12] *	-	Clitoris	-	-	Primary closure
Tzillinis [14]	63	L Labia Majora	Surgery / radiotherapy	Yes	Primary closure
Argenta [15]	35	R hemivulva	Surgery / radiotherapy	Yes	Primary closure
Chen [16]	23	L Labia Majora	Surgery / chemotherapy	Yes (in secondary recurrence)	Primary closure
Narendra [3]	50	R Labia Majora / mons pubis	Surgery / radiotherapy	yes	Pedicle rectus abdominis flap
Arafah [17]	25	Mons's pubis extended inguinal area	Surgery	No	Primary closure
Venugopal [13]	17	Clitoris	Surgery	No	Primary closure
Dolanbay [1]	51	L Labia Majora / mons pubis	Surgery	No	Primary closure
Current case	41	R Labia Majora / recurrence: hypogastric	Surgery	Yes	R PAP mons pubis: L TUG-PAP

R: Right; L: Left; *: only abstract available

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