Adenoid cystic carcinoma (ACC) originating from Bartholin's gland is a rare carcinoma of the female reproductive tract, accounting for less than 1% of all female genital malignancies and 0.1-7% of all vulvar carcinomas [1]. Histologically, adenoid cystic carcinoma comprises of only 15% of the many diverse Bartholin gland tumors. Ever since the first documentation by Klob in 1864, only approximately 350 cases have been reported[2]. Due to the low number of incidences, knowledge of its management and clinical outcome is limited. Here, we present a patient case with the ACC of Bartholin gland that was treated and recovered well after the surgery.

A 53-year-old multiparous , postmenopausal woman came to our clinical outpatient department with a pain-causing palpable nodular mass at the left vulvar area which has been growing for 2 years. Vulvar examination revealed an elevated firm nodular mass approximately 2.5x2.5 cm in size at the localization of Bartholin's gland. Pelvic and rectal examination showed no abnormalities. Microscopically, a tumor biopsy revealed a cribriform pattern of tubules and gland-like elements filled with homogenous basophilic mucin (Fig.1A and 1B). These characteristic features formed the basis for the diagnosis of ACC of the Bartholin's gland. Subsequent cystoscopy, sigmoidoscopy and chest x-ray were negative. Additional abdomino-pelvic MRI and whole body bone scintigraphy were used to evaluate local and distant metastases. There was no abnormal inguinal lymph nodes except for some minor local infiltration

in the left vulva area measuring 3x3cm (Figure 2). The patient underwent radical hemivulvectomy and bilateral inguinal-femoral lymphadenectomy. A minimum of 2-cm margin of the normal-appearing skin or mucosa around the tumor was removed. Pathological review was consistent with previous biopsy result. All margins of the resection were free of lymph node metastasis. The patient was discharged a month after the surgery without any complication and remained free of tumor for 3 years since the operation.

The original criteria for diagnosis of Bartholin's gland tumor was published in 1887 by Honan. The strict criteria did not correlate well with findings of the advanced tumors[3]. Thus, in 1972 Chamlian and Taylor from the Armed Forces Institute of Pathology reestablished the diagnostic criteria for Bartholin gland carcinoma[4]; histologically, areas of transition from normal to neoplasm must be found, tumor is compatible with Bartholin gland origin and no evidence of other primary tumor is identified.

Adenoid cystic carcinoma of Bartholin's gland is a rare vulva malignancy and often been disregarded as a cyst or inflammation. Its tendency for local perineural invasion is perhaps the cause for the initial infection-like itch and pain sensations. These ambiguous symptoms lead to the delay of diagnosis and treatment. Since the average age of patients is 49 years old with a range from 25 to 80, the possibility of Bartholin's

gland cancer should be investigated whenever a lesion is nearby [1].

Due to the lack of large series study, no consensus regarding the optimal treatment of ACC has been established. In most cases, two types of surgical procedures were performed: simple excision and radical vulvectomy with or without lymph node dissection. Literature reviews suggest a higher recurrence rate in patients undergone simple excisions than in those undergone radical vulvectomy patients, 69% to 43% respectively [5]. It is a common belief that the most important aspect of treatment is to obtain a tumor-free surgical margins to prevent the recurrence. In contrast, Yang et al. found similar recurrence rate in both groups of patients with positive and negative margins, 52.9% and 52.1% respectively [5] . This implied that the status of margin might not be as important as previously thought. As for the patients with positive margins and local recurrences, postoperative adjuvant radiotherapy has shown to be effective in controlling the disease[6].

Whether to perform unilateral or bilateral inguinal-femoral lymphadenectomy is still controversial in the treatment of ACC. However, the main determinant of survival of patients is the status of the inguinal-femoral lymph nodes[7]. Leuchter et al reported a 5-year survival of 52%, 36%, and 18% with zero, one or multiple positive lymph nodes[8]. With these results in mind and the fact that contralateral inguinal node involvement increases with lesion size, some surgeons prefer bilateral

dissection[9].

Bone and lung are the most common sites of distant metastasis for ACC of the Bartholin's gland. Liver, kidney, and brain metastasis are also found but occurred less frequently[10]. Information concerning the use and effectiveness of chemotherapy in the treatment is limited. Several chemotherapeutic treatments have been developed for this disease including various combination of adriamycin, dactinomycin, cyclophosphamide, and methotrexate[5]. Although some reports of the chemotherapy treatments are encouraging, the number of cases is too few for meaningful conclusion to be drawn.

ACC of Bartholin's gland is a rare vulva malignancy with an aggressive and unpredictable biologic behavior. Due to the small collection of cases, there is no consensus on a standard treatment. Similar to the treatment of patients with vulvar cancer, present study suggests that an early diagnosis with the combination of radical vulvectomy and bilateral inguinal-femoral lymph nodes dissection optimize patient chances of survival [7,8].

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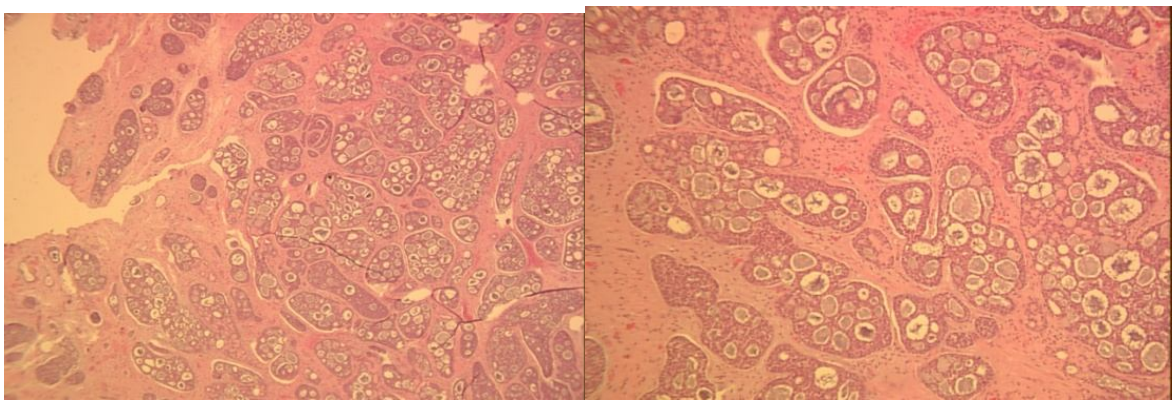
Figure 1 Microscopic features of perineal biopsy showing the tumor has a cribriform

pattern with the lumens containing basophilic mucin (hematoxylin and eosin, A:100x, B:400x).

Figure 2. Abdomino-pelvic MRI shows a soft tissue mass infiltrated into the left vulva area (arrow)



A B

Figure 1

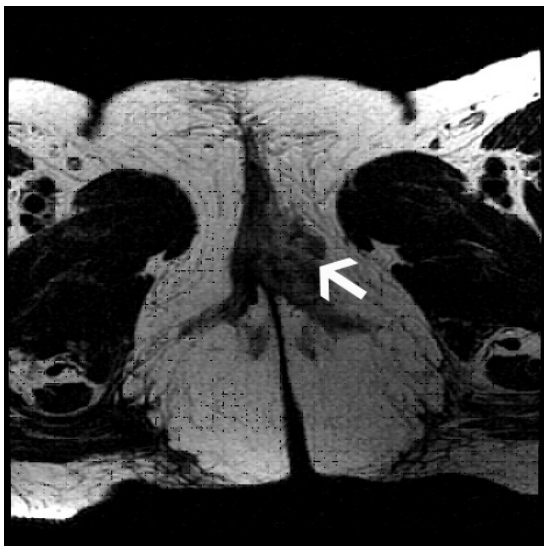


Figure 2