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Case Report

Primary Non-Hodgkin's Lymphoma of the Female Urethra Presenting as Carunculous of Urethra: A Very Rare Case Report

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Abstract

Background : Female urethral carcinoma occurred < 1% of all female malignancies. With a small prevalence of Malignant Urethral Lymphoma, there is lack of data about quality of the therapy.
Case Report : A 41-year-old woman came to Division of Urology, Department of Surgery Dr. Kariadi Hospital Semarang with chief complaint a recurrence mass on her urethra. Her general condition was fine, and has a normal vital signs. On the physical examination of urogenital region, there was mass on the orifice of urethra external that can easily bleed. She is already performed excision of urethral caruncle withimmunohistochemistry examination and found CD-20 positive which indicated a diffuse B-cell Lymphoma. Then she had 6 cycle chemotherapy procedures in Division of Hematooncology, Internal Medicine Department. Until now, she routinely check-up to urology and internal medicine departments to monitor her disease progression.
Conclusion : Female urethral carcinoma is very rare, although surgery alone has been established as effective for treating low-stage urethral cancer, neither surgery nor radiation therapy alone provide good results. Delay in diagnose and treatment can decrease patient survival.

Keywords : non-Hodgkin's lymphoma, immunohistochemistry, Urethral carcinoma, urethral caruncle, chemotherapy

INTRODUCTION

Primary urethral carcinoma is uncommon.[1,2] World event are more prevalent among men than woman.[1] It is more common in people aged over 65 years.[3] Female urethral carcinoma occurred <1% of all female malignancies.[4] Approximately one-third of non-Hodgkin's lymphoma (NHL) arise from sites other than lymph nodes, spleen or the bone marrow.[5] Extra nodal lymphoma can originate from almost every organ, including the urethra.[4]

Immune system abnormalities and infectious agent have been consistently associated with increased risk of Non-Hodgkin's lymphoma (NHL) by approximately 100 fold.[6] Another factor that inconsistent causing NHL include immunosuppressive drugs, exposure of hair dyes, and exposure to x-gamma radiation6. Multistep accumulation of genetic aberrations induces NHL.[6] Recurrent translocations, often as an initial step in transformation of malignancy.[7] Consensus regarding optimal treatment modalities for high-stage urethral cancers not yet established, but surgical procedure has been established as first line therapy for treating low-stage urethral cancer, but multimodality therapy has been shown to be superior to any single modality therapy.[1] With a small prevalence of Malignant Urethral Lymphoma, there is lack of data about quality of the therapy. We want to share our experience in facing this case, we hope our experience will be a reference to other patients with the same disease.

CASE PRESENTATION

A 41-year-old woman came to Urology Department of Dr. Kariadi Hospital Semarang with his family with chief complaint, there were a recurrence mass on her urethra. The mass looked as bigger as corn seed, didn't grow bigger, easy to bleed, and wasn't painful. There wasn`t any complaint in micturition, and changes in the menstrual cycle. The patient never had severe illness before, no allergic history, and no history of routine medication, and no history of changing sexual partners, no family history of cancer, and no history of hormonal contraception. He work as a government employee and had sufficient socio-economic condition.

The mass first appeared in June 2018, she felt a mass that easily bleed after she finished urinating, then she went to the obstetrician. By an obstetrician, she was advice to go to an urologist, and she was diagnosed by urethra caruncle and was planned to get a surgical procedure to remove it. Four days later, she got an operation and the mass sent to pathology anatomy department and the result was a carunculous granuloma with atypical cell.

Two months after the surgical procedure, the mass was reappeared with the same complaint. She came back to urologist to consult and was scheduled to get another surgical procedure to remove the mass. After that, the mass sent to pathology anatomy department and the result was a malignant tumor suspect Non-Hodgkin Lymphoma or Undifferentiated Carcinoma. Because lack of the facilities, she was referred to Dr. Kariadi Hospital Semarang to get a further treatment.

In the Dr. Kariadi General Hospital Semarang, she planned to undergo immunohistochemistry (IHC) examination. IHC examination found CD-20 positive which indicated a diffuse B-cell Lymphoma. Then she had chemotherapy procedure with Division of Hematooncology, Internal Medicine Department.

Physical examination revealed a 41-year-old woman with body weight 70kg, height 165cm, general appearance looks well, and there was no changes in physical activities. Her Glasgow Coma Scale was E4M6V5 = 15, with blood pressure 100/70 mmHg, pulse rate 77x/min (regular, volume and tone were enough), respiration rate 18x/min (regular, deep of breath normal, no retraction), temperature 36,5°C, and pain score 0 Visual Analogue Scale (VAS). There was no anemic on both conjunctivas, isochoric pupil with diameter 3mm/3mm and positive reflex pupil. Chest examination revealed symmetrical chest expansion, normal breath sound without wheezing or rhonchi, normal heart sound, neither murmur nor gallop. No abnormality found in the abdominal examination. On the urogenital examination, there was mass on the orifice of urethra external that can easily bleed. There is no enlargement on the regional lymph node. Motoric and sensory status in both lower limbs were normal, no cold acral and Capillary Refill Time< 2". And there wasn't any edema on the lower limb. Laboratory findings showed Hemoglobin 13.0 gr%, Hematocrit 39,1%, Leucocyte 5.700 mmc, thrombocyte 314.000 mmc, blood glucose 124 mg/dL, Urea 20 mg/dL, Creatinine 0.8 mg/ dL, Sodium 140 mmol/mL, Potassium 3,8 mmol/ L, Chloride 97 mml/ dL, PPT 10.8 (controlled : 11.0), APTT 33.0 (controlled : 33.2).

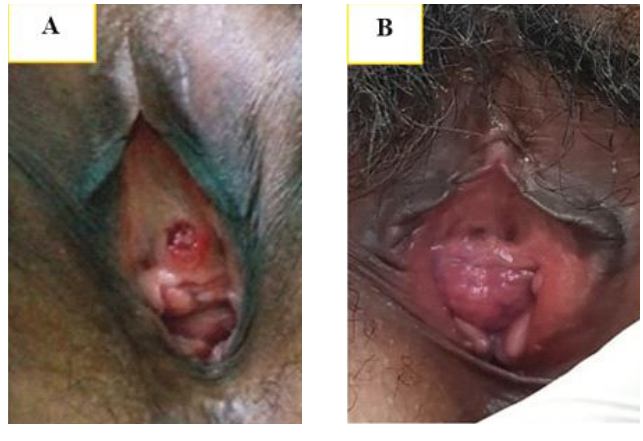

Figure 1. A. Before excision; B. After excision

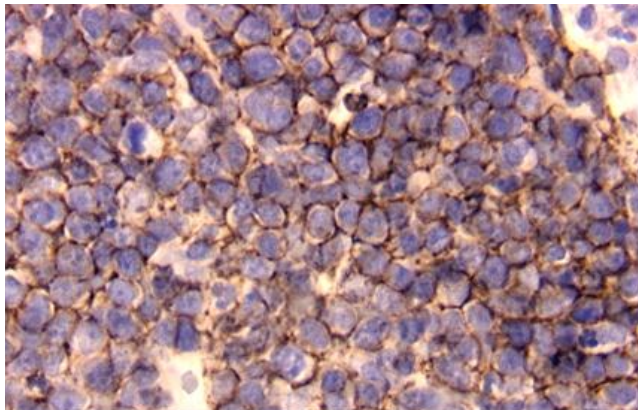

Figure 2.

On January 2019, she got 6 cycle of chemotherapy procedures from Division of Hematooncology, Internal Medicine Department. The protocol for the chemotherapy was Rituximab 650mg, Vincristine 2mg, doxorubicin 70mg, Endoxan 1300mg.

Imunohistochemistry

Until now, she routinely check-up to urology and internal medicine departments to monitor her disease progression.

DISCUSSION

Our patient, first came to the urology department at Kariadi Hospital with a diagnosis of carancule of the urethra, carcinoma of urethra appeared as a benign and stemmed lesion and appeared as a polyp that appeared from the posterior lip of the urethral meatus.[8,9] Carancule urethra is the most common lesion in a woman's urethra and occurs mainly in postmenopausal women.[9,10] Female urethral carcinoma occurred <1% of all female malignancies.[4] Generally, subcategories of this cancer are based on histology and include transitional cell carcinoma, squamous cell carcinoma, adenocarcinoma, and "other" histologic types.[1]

Approximately one-third of non-Hodgkin lymphomas (NHL) arise from sites other than lymph nodes, spleen or the bone marrow.[1] They may also arise from sites normally devoid of lymphocytes. In the population-based cancer registries of the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) Program, referring to the period 1978–1995 approximately 30% of all lymphomas were extra nodal and almost half of all extra nodal NHL cases reported had diffuse large B-cell lymphoma (DLBCL) histology.[5] Extra nodal lymphoma can originate from almost every organ, including the urethra.[5]

Like other malignancies, the etiology of NHL is still difficult to determine. Although immune system abnormalities and infectious agents in theory contribute the most, in this case we cannot link the risk factors that contribute to the emergence of malignancy in these patients. Biopsies in these cases are widely accepted as the gold standard for the diagnosis of malignant lymphoma, and also provide adequate ingredients for immunochemistry.[3]

Because of the paucity of urethral cancer cases and its aggressive nature, the majority of case reports and series published are from single tertiary centers with small sample sizes. Thus, it has been difficult to come to a consensus regarding optimal treatment modalities for high-stage urethral cancers. Although surgery alone has been established as effective for treating low-stage urethral cancer, neither surgery nor radiation therapy alone provide good results. However, multimodality therapy has been shown to be superior to any single modality therapy.[1]

According to EAU-guideline, treatment of localized urethral carcinoma in females is urethrectomy and urethra-sparing surgery (level of evidence 3).[11] In this case, she got excision urethral caruncle surgery to maintain integrity, function of the lower urinary tractand also to know what the cell type of the tumor.[12] After we know the tumor is B-cell type Lymphoma, we decide to joint management with Division of Hematoocology, Internal Medicine Department. They suggest giving a chemotherapy procedure with regimen that consists of Rituximab 650mg, Vincristine 2mg, doxorubicin 70mg, Endoxan 1300mg. The regimen is in accordance with the guidelines of the National Comprehensive Cancer Network (NCCN) (level of evidence 1).[13] According to systematical review too, CHOP 21 regimen combined with Rituximab which used by our patient more superior than another chemotherapy, and also have 5 years survival rate 78%.[14,15]

CONCLUSION

A Non-Hodgkin B-cell type Lymphoma on female urethra is a very rare case. Until now, there is still no

consensus in handling this case. The therapy we used in this case, according to a combination of therapeutic guidelines for urethral carcinoma and guidelines for Non-Hodgkin Lymphoma therapy.

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