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Case Report

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PRIMARY ADENOCARCINOMA OF THE FEMALE URETHRA: A CASE REPORT

A. Kbirou*, A. Ettanji, M. Rkik, Y. Fadel, M. Modeste, H. Lachhab, Y. Daghdagh, Saleh, M. Dakir, A. Debbagh, R. Aboutaib

Department of Urology, IBN ROCHD University Hospital, Morocco.
Mohamed VI Centre of Cancer Treatments, The University Hospital Casablanca, Morocco.

***Corresponding Author: Adil Kbirou**

Department of Urology, IBN ROCHD University Hospital, Morocco.

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ABSTRACT

Primary adenocarcinoma of the urethra is rarely reported.Vague symptoms in the early stages delay the diagnosis in most patients. Surgery, chemotherapy and radiotherapy are used as treatment modalities for these tumors, either alone or in combination. We report a case of a 53-year-old female Magnetic resonance imaging and Cystourethroscopy revealed urethral tumor. Pathology report was consistent with primary adenocarcinoma of the urethra.

KEYWORDS: Female, Urethra, Adenocarcinoma.

INTRODUCTION

Primary malignancies of the female urethra are rare, accounting for less than 1% of genitourinary malignancies. Definitive clinical diagnosis of Primary Adenocarcinoma is difficult and must be differentiated from other genitourinary tumors. we report a case of a 53 year old female diagnosed with PAFU who underwent chemotherapy, initially the evolution was favorable then the patient presented a obstructive acute renal failureat 8 months of follow-up.

CASE REPORT

A 17-year-old patient, without antecedents, with 2-year history of symptomatic urinary obstruction was presented to emergency with gross terminal hematuria. Abdominal examination revealed a non-tender, palpable mass in the hypogastric region. Inspection and bimanual palpation rectovaginal examination was normal (fig.1) Physical examination revelated no cervical, axillary or inguinal lymphadenopathy. Abdomen and pelvic CT scan with contraste descripted a suspicious lesion under vesical, locally advanced with abdominal and pelvic lymphadenopathy and secondary hepatic lesions(Fig.2) An MRI of the pelvis objectived a invasive large urethral mass 6.5× 6.1 cm with heterogeneous enhancement in the bladder neck and urethra concerning for a urethral neoplasm, Bone metastasis of L3 and pelvic lymphadenopathy.(Fig. 3)

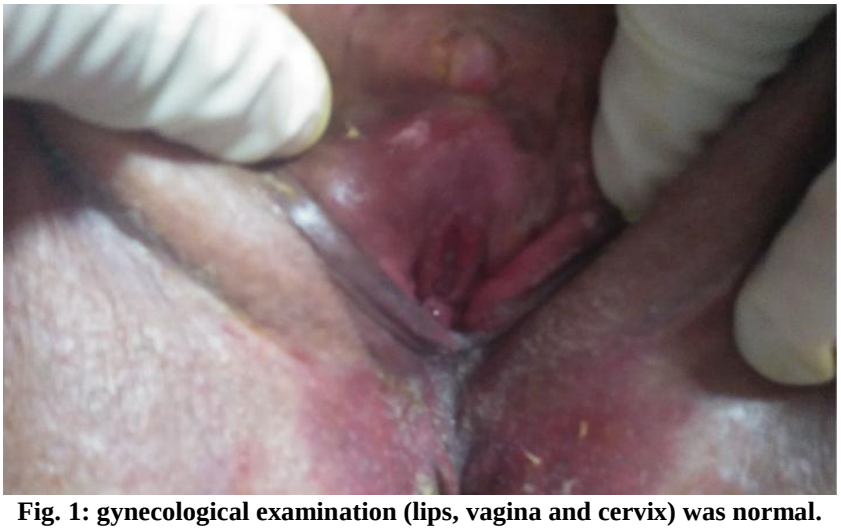

Fig. 1: gynecological examination (lips, vagina and cervix) was normal.

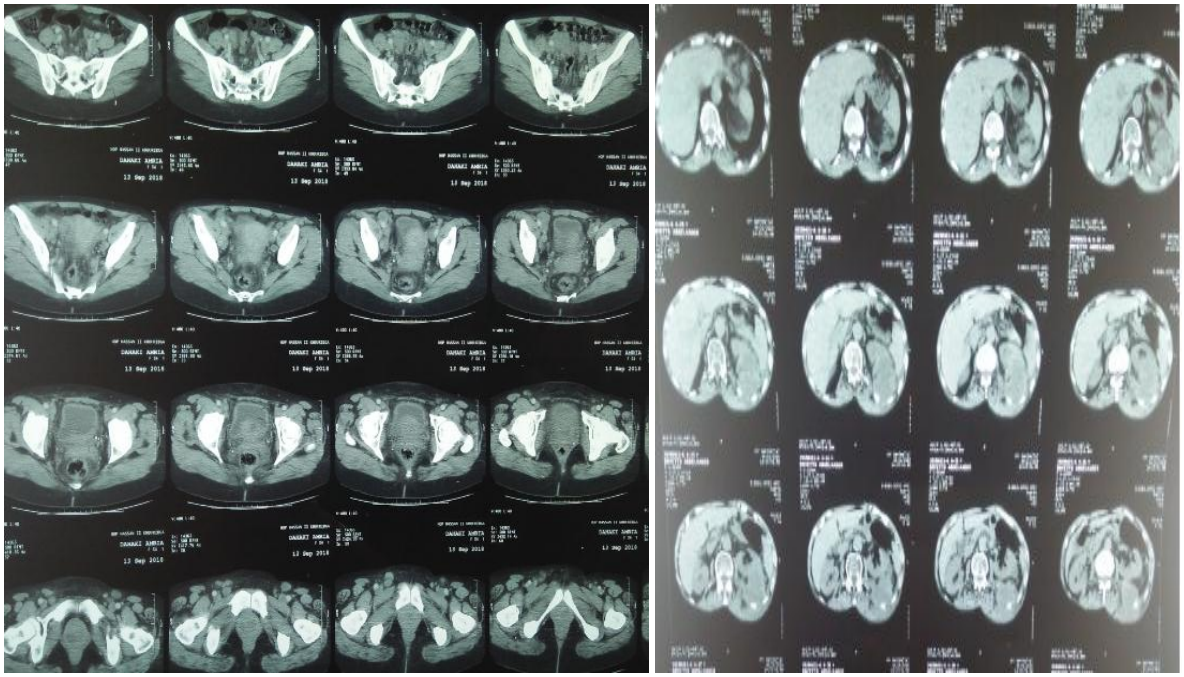

Fig.2 : CT scan demonstrated a large urethral mass with abdominal and pelvic lymphadenopathy and secondary hepatic lesions.

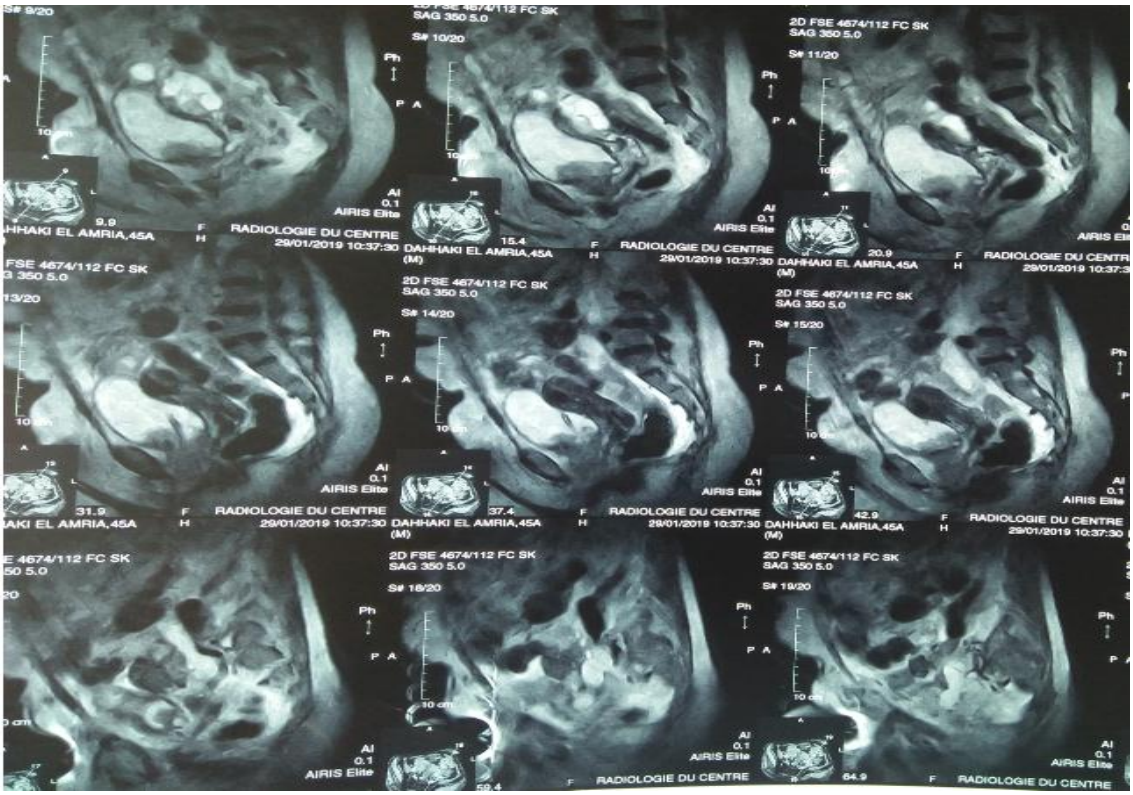

Fig. 1. MRI pelvis showing a invasive urethral mass 6.5 × 6 cm with heterogeneous enhancement in bladder neck and urethra concerning for urethral neoplasm.

The patient benefited of a urethrocystoscopy showing a large invasive mass almost obstructing the entire lumen with bleeding then a biopsy. The pathology report showed moderately differentiated and invasive adenocarcinoma of the urethra. The patient received a systemic chemotherapy without surgery (locally advanced stage), initially the evolution was favorable then the patient presented a obstructive acute renal failureat 8 months of follow-up.

DISCUSSION

Female urethral adenocarcinoma(FUA) is rare and accounts for approximately 0.02% of all female cancers

and corresponds to 0.003% of all malignant cancers occurring in the female urogenital tract.[1] The clinical data reported that the incidence of this disease increases with age, with highest rates in women aged 65 years or older for all histologic types.[2]

They are mostly squamous or urothelial in nature (70% are squamous cell and 20% are transitional cell, respectively). Approximately 10 % of malignant cancers of female urethra have been reported as adenocarcinoma derived from Skene's paraurethral ducts and glands as our case.The two primary histologic subtypes of adenocarcinoma of the urethra are as follows: Columnar/mucinous ("intestinal") and clear cell type.[7] There is a striking anatomical distribution of these tumors based on their histology: distal urethral carcinomas are predominantly squamous cell type whereas proximal urethral carcinomas are more often urothelial and adenocarcinoma type.[2] The aetiology of the FUA is unknown.[10] Although smoke, exposure to aromatic amines, arsenic ingestion, and analgesic abuse are associated with transitional-cell carcinoma of the bladder and urethra, no such correlations has been demonstrated with urethra adenocarcinomas.[7,9] The risk factors for developing this cancer include: urethral stricture, chronic irritation from intermittent catheterization or recurrent urinary tract infections, urethral diverticulum, sexually transmitted disease with human papilloma virus, and prior radiationtherapy, our patient had no risk factors and without antecedents.

Most patients with urethral adenocarcinoma present with obstructive symptoms, dysuria, urethral bleeding, urinary frequency, and often a palpable urethral mass or induration. The signs and symptoms of FUA vary and are neither diagnostic nor pathognomonic.[1.2]

Tumors spread generally by local extension and may ulcerate to the skin and vulvar region as the tumor progresses.[3] Proximal lesions may extend posteriorly into the vagina or proximally into the bladder. Lymphatic spread is uncommon in the early stages, but clinically, palpable nodes may be present in up to one-third of patients at presentation and half of patients with advanced and proximal tumors. Hematogenous spread may occur in the lung, liver, bone, and brain, in order of frequency.[1] The evaluation of females with suspected urethral adenocarcinoma includes cystourethroscopy, physical examination under anesthesia, CT of the abdomen and pelvis and chest radiography. MRI has been used to evaluate pelvic lesions and is beneficial in the determination of local extension. MRI has been reported to be accurate in local staging of urethral tumors in 90% of patients. Transvaginal ultrasonography may also provide clues for diagnosis.[1,5,9] However, the literature shows that an ultrasonographic study has lower specificity when compared to Magnetic Resonance Imaging (MRI) in the diagnosis of urethral cancer and determination of its local extension.[1.2] Cystourethroscopy and biopsy are integrant to confirm the diagnosis, establish the histology and grade, determine the local extent of cancer, the precise location within the urethra, and whether a concomitant bladder cancer may be present and eliminate metastatic lesion originating from the intestinal tract or ovaries.[1]

The prognosis is determined largely by the clinical stage and the location of the lesions. Tumors in the distal urethra tend to have a better outcome.[1] There is no consensus regarding the treatments of FUA due to few reported cases. Among the treatment options in early-stage disease the surgery is the most commonly used: partial resection with preservation of the urethra. Treatment of advanced-stage in women affected by urethral cancers has historically been a wide surgical excision.[5,13] Multimodal treatments with surgery, radiation, and systemic chemotherapy could be beneficial for selected patients with advanced tumors.[11,13] Some authors suggest adjuvant polychemotherapy for locally advanced FUA ,in our case the surgery was impossible whose patient was receiving chemotherapy

CONCLUSION

Primary Female urethra adenocarcinoma (PFUA) is a rare aggressive associated with a relatively poor prognosis. These tumors are usually large masses, which typically spread through the lymphatic system. Patients present with vague symptoms similar to urinary tract infections. Cystourethroscopy permits visualization of the urethral tumor and allows biopsies to be performed to remove samples for histologic examination. The role of imagingin the diagnosis of FUA continues to evolve and strive more accurate methods of differentiating between benign and malignant cancers. Multimodal therapy including surgery, chemotherapy, and radiotherapy is required in the modern management of FUA, although the specific role and combination of each treatment is less clearly determined.

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