Hematotrachelometra with Acute Salpingitis in a 46, XY Disorder of Sexual Development(DSD): A Case Report

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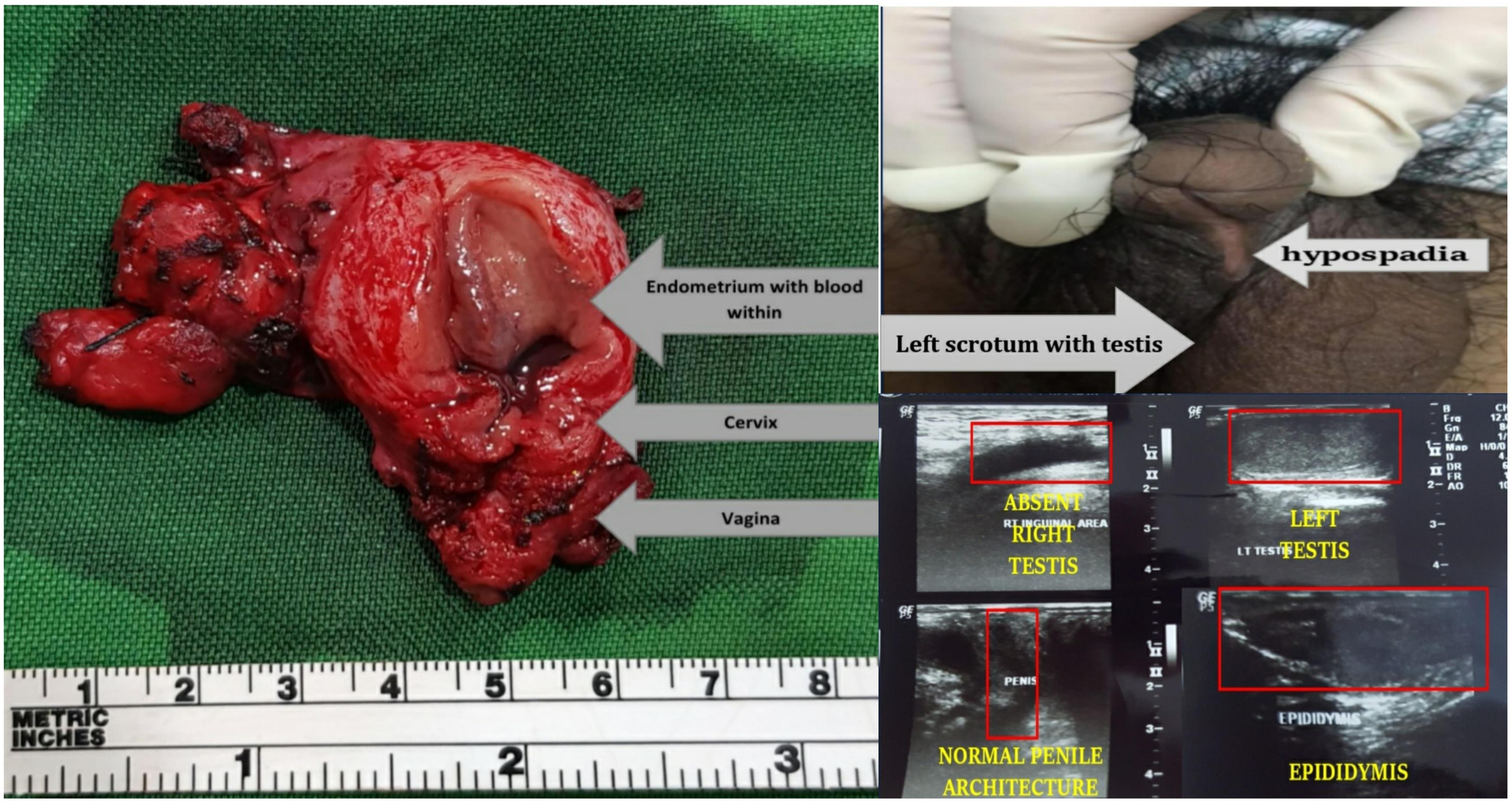
INTRODUCTION

46,XY Disorders of Sexual Development (DSD) is characterized by an atypical external genitalia due to incomplete intrauterine masculinization with or without the presence of mullerian structures.[1]



Gross specime of uterus, cervix and vagina(LEFT); Male External Genitalia (TOP RIGHT); SCROTAL ULTRASOUND (BOTTOM RIGHT)

RESULTS AND DISCUSSION

We present a case with 46,XY DSD in a 23 year old, male, initially presenting with dysuria, hematuria, abdominal pain, and history of urinary tract infection. Patient had hypospadias, normal left scrotal sac with testis but an empty right scrotum. An exploratory laparotomy with biopsy of right adnexal mass with purulent discharge showed salpingitis. Hence, referral to a tertiary center was made and a patient-centered multidisciplinary team composed of gyne-oncologist, reproductive endocrinologist, urologist, psychologist and psychiatrist managed the patient. Work-up done showed pelvic masses on transrectal and transabdominal sonogram: two tubulocystic masses - a 7.7 x 5.0 x 9.0 cm with incomplete septations and medium low level echo fluid posterior and more to the left of the urinary bladder, and a 6.7 x 2.6 x 3.0 cm, superior and more to the right of the previous mass. Plain CT scan showed a 9.5 x 5.2 x 6.6 cm thick walled multilocular cystic mass posterior to the urinary bladder. Impression was tubo-ovarian abscess versus complicated seminal vesicle or prostatic cyst. Elevated follicle stimulating hormone, with normal estradiol and testosterone indicated testicular dysfunction while elevated CA-125 supported infection or malignancy. Surgery and histopathology confirmed uterus with proliferative phase endometrium causing hematotrachelometra and salpingitis. Karyotyping result was 46, XY. Patient suffered mild depression postoperatively, but psychiatric counseling helped patient to adjust to his health concerns. Gender orientation remained male congruent with his external genitalia.

CONCLUSION

This paper reports a rare case of pelvic infection due to salpingitis from hematotrachelometra in a 46,XY Partial gonadal dysgenesis (PGD) which necessitates complete removal of mullerian structure, continuous surveillance of scrotal testis for malignancy and providing psychosocial support in patients with DSD.

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