True Hermaphrodism Presenting as Pelvic Abscess

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INTRODUCTION

True hermaphroditism is a rare form of ambiguous genitalia characterized by simultaneous presence of both normal male and female gonadal tissues in a same patient.[1,2] Although uncommon, true hermaphroditism has been reported in more than 400 individuals, to date.[3] We report a hermaphrodite patient presenting with a pelvic abscess.

CASE REPORT

A 29-year-old patient presented with fever, abdominal pain, and pyuria since 8 days earlier. There was no history of episodic hematuria. The patient was reared as a male and appeared to be a phenotypically male with short stature and male pattern of body hair. Local examination showed suprapubic fullness with distal penile hypospadias and an empty left scrotal sac. Routine investigations showed moderate anemia and a total leukocyte count of 9×10^9/L. Urine examination revealed innumerable pus cells, and culture showed presence of *Escherichia coli*. Abdominal ultrasonography showed a 13 × 7-cm, thick-walled encysted lesion in the midline pelvic area, displacing posterior wall of the bladder anteriorly and extending to the left iliac fossa. Computed tomography revealed a large loculated fluid collection with thick walls and septations in the retrovesical plane including left lateral pelvic wall, which was highly suggestive of a large abscess. The patient underwent exploratory laparotomy in order to drain the retrovesical pus, which revealed a unicornuate uterus ending in a blind vaginal pouch along with left fallopian tube and ovary. The vaginal pouch was filled with pus and had a fistulous connection with the bladder anteriorly. Hysterectomy with left salpingo-oophorectomy was performed along with ligation and excision of the fistulous tract between the bladder and the vaginal pouch and excision of the vaginal pouch.

Postoperative hormonal assay was carried out which showed a raised serum estradiol (66.5 pg/mL) and *luteinizing* hormone (13.2 mIU/mL) with a progesterone (0.9 ng/mL), testosterone (541.4 ng/dL), and follicle-stimulating *hormone* (15.49 mIU/mL) within the reference range. Chromosomal analysis using GTC banding revealed normal male karyotype of 46XY in all cells analyzed. The operative specimen was sent for histopathological examination and revealed uterous endometrium and ovarian and vaginal tissue with diffused inflammatory cell infiltration.

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DISCUSSION

There are 4 major groups of ambiguous genitalia: female pseudohermaphroditism, male pseudohermaphroditism, mixed gonadal dysgenesis, and true pseudohermaphroditism. In true pseudohermaphroditism, 4 types of gonadal distribution are seen. The most common is the presence of bilateral ovotestes. Other variants include ovary on one side and ovotestis on the other side, testis on one side and ovotestis on the opposite side, and presence of testis on one side and ovary on the other side, which is the rarest form.[2,4,5] The most common cytogenetic pattern of these patients is genotypic female (46XX). All forms of mosaicism are next in frequency and the rarest are genotypic males (46XY). Gonadal tumors occur with an incidence of 4% among those with 46XX karyotype and up to 10% in those with 46XY and 46 XX/XY mosaicism.[6]

Combination of 46XY genotype with the presence of the right testis and the left ovary makes this case the rarest of rare cases of true hermaphroditism. To our best knowledge, this is the first case of 46XY true male hermaphroditism with a functioning testis on one side and a contralateral functioning ovary presenting as a pelvic abscess.

CONFLICT OF INTEREST

None declared.

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