A RARE PRESENTATION OF TRUE HERMAPHRODITISM AS HERNIA UTERI INGUINALE IN AN ADULT

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ABSTRACT

BACKGROUND

True hermaphroditism is a rare disorder of sexual differentiation characterised by presence of both ovarian and testicular tissue regardless of their karyotype. Most of these cases result from 46XX karyotypes, and fewer involving 46 XY or 46 XX/46 XY mosaicism.¹ We are reporting a case of true hermaphroditism presented as left-sided inguinal hernia in an adult male patient.

KEYWORDS

Hermaphroditism, Hernia, Disorder of Sexual Differentiation, Ovotestis, Karyotyping, Dysgerminoma, Gonadoblastoma, Chordae, Hypospadias and Cryptorchidism.

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BACKGROUND

True hermaphroditism results in the presence of both testis and ovarian tissue also called ovotestis, and occur in less than 10% of disorders of sexual differentiation cases.¹ About 90% of cases, patients have 46 XX karyotype. Rarely, 46 XY/46 XX mosaicism and 46 XY karyotype.¹

In this condition, Testosterone and Mullerian inhibitory substance (MIS) are either normal or low. Karyotyping and bar bodies from buccal mucosa can be used for the diagnosis; however, for final diagnosis, histological documentation of both types of gonadal epithelium is a must.²

Among the ovotestis, 46% were in ovarian position, 26 % in labioscrotal fold, 24% in inguinal canal, 20% ovotestis bilaterally.² We are reporting a case of true hermaphroditism presented as left inguinal hernia.

CASE REPORT

A male patient aged 45 years, known case of primary infertility presented with history of left inguinal swelling of 4 years' duration, active married life of 15 years. Patient gave history of undescended testis on the right side since birth. Clinically, he was diagnosed to have left-sided indirect partially reducible inguinal hernia with right undescended testis.

General physical examination revealed features of male phenotype in the form of hair over the face, axilla, external genitalia appear normal male with pubic hair. Right hemiscrotum was empty and right testis or testis like structure was not felt in the inguinal canal.

Seminal analysis revealed evidence of azoospermia. USG abdomen and scrotum reported as left inguinal hernia with undescended right testis, no intra-abdominal pathology found. With the above diagnosis, patient was planned for

Lichtenstein's hernioplasty under spinal anaesthesia and found complete indirect sac on left side with uterus like structure along with fallopian tube and ovaries found.

This structure was excised totally and sac was closed, Lichtenstein's mesh plasty done.

One more structure looking like small atrophied testis placed in left hemiscrotum.

HPE of specimen confirmed that they are uterus and ovaries.

**Image A: 1. Ovaries/Testis.
2. Fallopian Tube.**

**Image B: 1. Ovary/Testis. 2. Fallopian Tube.
3. Fundus of Uterus. 4. Cervix.**

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**Image C: 1. Fundus of Uterine Cavity.
2. Body of Uterine Cavity. 3. Cervical Canal.**

DISCUSSION

The term “hermaphrodite” is derived from Greek mythological God “Hermaphroditus”, son of Hermes and Aphrodite, whose body after being merged with nymph Salmacis assumed a more perfect form with both male and female attributes.^{3,4}

The exact aetiology of hermaphroditism remains enigmatic. Clinical presentation is an abnormal external genitalia ranging from normal male to normal female. In many of these cases, such distinction may not be present and hernia, chordae, hypospadias and cryptorchidism might be found.⁵ Our case presented as left-sided indirect inguinal hernia with right undescended testis.

Clinical and pathological documentation of the gonads is a must for diagnosis.

In true hermaphrodites, gonads are always asymmetrical. On the basis of this, they are classified as, 1. Bilateral: Testicular and ovarian tissue identified on both sides (50%). 2. Lateral: Testis and contralateral ovary (30%). 3. Unilateral:

Ovotestis on one side and testis or ovary on other side (20%).⁶ Our case was unilateral type.

The choice of rearing hermaphrodite as male or female sex is governed by phallus size.⁶

True hermaphroditism is rarely associated with gonadal tumours (4.6%),⁴ mixed gonadal dysgenesis, predisposes to gonadal malignancy due to presence of dysgenic cells. Malignancies like dysgerminoma and gonadoblastoma have been reported in true hermaphroditism.

CONCLUSION

Although hernia uteri inguinale is a very rare disorder in phenotypic male, a preoperative diagnosis would be helpful in surgical planning. Surgeons should be aware of the possibility of presence of the uterus or another organ in the hernial sac, and sliding components should be replaced carefully into the abdomen to prevent damage.

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