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DOI: 10.4103/ijawhs.ijawhs_68_25

Bilateral recurrent inguinal hernia complicated by disorders of sex development and Sertoli–Leydig cell tumors: A rare case report and review of the literature

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Abstract

Inguinal hernia associated with disorders of sex development (DSD) is uncommon, and an occurrence further complicated by Sertoli–Leydig cell tumors (SLCTs) is exceedingly rare. A woman underwent surgery for recurrent inguinal hernia. Intraoperatively, the hernia sac contents were identified as adnexal tissue. Postoperative pathological examination revealed a 46,XY DSD with SLCTs. Although rare, hernia surgeons should maintain awareness of these conditions.

Keywords:

DSD, recurrent inguinal hernia, SLCTs

Introduction

Inguinal hernia is a common surgical condition characterized by the protrusion of intra-abdominal organs through a congenital or acquired defect in the inguinal region, resulting in an external abdominal bulge.[1] The incidence rate is 27%–43% in men and 3%–6% in women. The hernia contents are usually the omentum or small intestine.[2,3]

Disorders of sex development (DSD) are congenital conditions marked by atypical chromosomal, gonadal, or phenotypic sex and are sometimes associated with gonadal tumors and infertility.[4,5] In some patients with DSD, reproductive tissues, for example, fallopian tubes, have been found within hernia sacs.[6]

The presence of tumors within a hernia sac is very rare. In 2014, Fagouri *et al*. reported a case of DSD, in which the testes were removed from the inguinal canal, and pathological examination revealed a Sertoli–Leydig cell tumors (SLCTs).[7] SLCTs are rare ovarian sex cord-stromal neoplasms, accounting for less than 0.5% of primary ovarian tumors. It is characterized by uncontrolled proliferation of naturally occurring testicular structures (Sertoli and Leydig cells) of varying degrees of differentiation in ovary.[8,9]

Here, we report a patient with bilateral recurrent inguinal hernia (RIH) complicated by DSD and SLCTs. By emphasizing the significance of abnormal preoperative findings, prompt intraoperative consultation, and thorough pathological examination, we aim to provide ideas into the diagnosis of unusual hernia sac contents.

Case History

The patient was a 67-year-old phenotypic female (socially living as a woman) with a

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Submitted: 14-Jul-2025
Revised: 28-Sep-2025
Accepted: 14-Oct-2025
Published: 18-Apr-2026

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How to cite this article: Lin Y, Huang M, Jin B, Ye Z. Bilateral recurrent inguinal hernia complicated by disorders of sex development and Sertoli–Leydig cell tumors: A rare case report and review of the literature. Int J Abdom Wall Hernia Surg 2026;9:43-7.

body mass index of 21.9 kg/m². Eighteen months earlier, she had undergone a laparoscopic bilateral inguinal hernia repair at a local hospital for reducible masses in both groins.

Approximately 3 months before this visit, a mass reappeared in her right inguinal region; 1 months later, a similar mass developed on the left side. Recently, she began to experience mild discomfort during activities, which finally prompted her to seek evaluation. She denied any urinary complaints, as well as any nausea, vomiting, abdominal distension, or changes in bowel habits.

Her surgical history includes eardrum surgery on the right and left sides 30 and 20 years ago, respectively, as well as a right wrist surgery 4 years ago. Notably, she had never menstruated and has never been pregnant, and she did not seek medical evaluation for these issues. Aside from the above, her medical history was unremarkable.

On examination, there was a firm, mobile mass approximately 2 cm × 2 cm in the left inguinal region and a similar 4 cm × 4 cm mass on the right. Both were irreducible, non-tender, and painless, with no overlying skin changes.

Preoperative ultrasound confirmed the presence of bilateral inguinal hernias. The left inguinal canal was approximately 10 mm in diameter and the hernia was partially reducible, whereas the right canal was approximately 8 mm and its herniated content could not be significantly reduced. Abdominal computed tomography (CT) demonstrated bilateral widening of the inguinal canals with the soft tissue density masses and no uterus was visualized.

Notably, before her earlier surgery, transvaginal ultrasound revealed absence of the uterus and poorly visualized ovaries. Perineal ultrasound demonstrated bilateral hypoechoic groin masses, suggestive of hernia. Pelvic CT likewise confirmed bilateral inguinal hernias and showed no uterus.

The patient was diagnosed with bilateral RIH and underwent tension-free hernia repair. Hernia sac contents potentially appeared to be gynecologic in nature. A gynecologic consultation during surgery suggested that the left-sided mass was likely adnexa [Figure 1A] and the right-sided mass was likely the uterus [Figure 1B]. Considering the patient's age and surgical findings, with consent from the patient's family, bilateral masses were excised and submitted for pathological examination. Meshes were placed on both sides and drainage tubes were placed bilaterally.

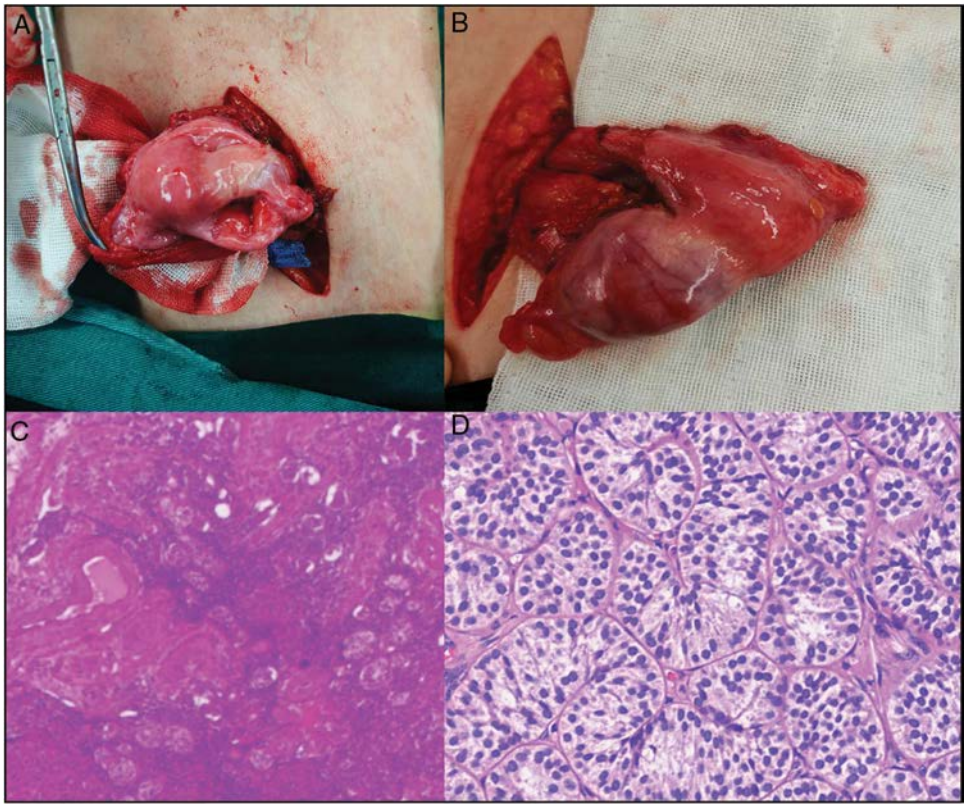


Figure 1: (A) Contents of the left hernia. (B) Contents of the right hernia. (C) Left gonadal tissue (H&E, 50×) showing features suggestive of a disorder of sexual development. (D) Right gonadal tissue (H&E, 200×) demonstrating tumor cells arranged in well-formed nests and lobules, with occasional tubular/acinar-like structures. The cells are polygonal to columnar with moderately abundant pale eosinophilic cytoplasm, and mitotic figures are rare.

Postoperative hormone levels were assessed, and the results were as follows: hCG-β: 1.4 IU/L; FSH: 76.44 IU/L; LH: 30.01 IU/L; P4: 1.08 nmol/L; PRL: 139.63 mIU/L; E2: <55 pmol/L; T: 0.64 nmol/L; CA-125: 6.5 U/mL; AFP: 2.2 ng/mL; CA19-9: 5.5 U/mL; and CEA: 0.8 ng/mL.

Pathological examination of the left-sided mass revealed features consistent with a DSD [Figure 1C]. Examination of the right-sided mass demonstrated ovarian-like stroma with stromal (Leydig cell) hyperplasia and scattered vas deferens-like structures [Figure 1D]. Foci also contained smooth muscle, fallopian tube-like elements, and paramesonephric duct cysts. In addition, a 2.8-cm grayish-yellow mass was identified, composed of proliferating supporting cell-derived structures, consistent with a well-differentiated SLCTs arising in the context of a disorder of sex development.

Immunohistochemistry showed positivity for sex cord-stromal markers, including inhibin-α, calretinin, SF-1, and WT1. The right-sided lesion additionally expressed Melan-A, β-catenin, OCT3/4 (weak), CD99, CK7, PR, and focal EMA, with a low proliferative index (Ki-67 ≈ 5%). The left-sided lesion expressed ER, PR, inhibin-*α*, calretinin, SF-1, and desmin. Negative markers included CD34, CK7 (left side), Uroplakin II, S100P, GATA3, SALL4, PAX8, and PAX2. Overall, the profile supported a well-differentiated SLCTs in the setting of a DSD. Chromosomal karyotyping revealed a 46,XY genotype.

Bilateral drainage tubes were removed on the 9th day. She discharged on the 11th day and was advised to follow-up with gynecology for ongoing monitoring. During the 3-month follow-up after the operation, the patient did not report any obvious discomfort symptoms.

Discussion

DSDs can be grouped into chromosomal DSD, 46,XY DSD, and 46,XX DSD.[10] The 2006 Consensus Statement highlights an emerging perspective: psycho-social care provided by mental health staff with expertise in DSD should be an integral part of management to promote positive adaptation.[5]

In patients with DSD complicated with inguinal hernia, reproductive system-related tissues can occasionally be found within the hernia sac. In male patients, hernia contents such as fallopian tubes and the uterus is most commonly associated with persistent Müllerian duct syndrome.[6] In "female" patients with complete androgen insensitivity syndrome (CAIS), the contents of the hernia can be the testicles. The karyotype of these patients is 46,XY, but the phenotype is female.[11]

In addition, hernia sacs containing adnexal structures have been reported in phenotypic females without DSD. Among female children, particularly premature infants or those with congenital anomalies, hernia sacs may contain the uterus or ovaries. This is often due to the failure of the canal of nuck to close before or at birth, allowing abdominal organs to protrude through the inguinal canal.[3] Although cases of adult women with similar hernia contents have been reported, they are less common.[2]

The coexistence of DSD and SLCTs is very rare and has occasionally been observed in patients with CAIS.[7] SLCTs is rare and account for less than 0.5% of all ovarian cancers. The majority are unilateral and confined to the ovary.[12] Approximately 75% of SLCTs cases occur in women between 20 and 30 years of age.[8] According to the World Health Organization's suggested classification, SLCTs can be categorized into four histological types: well-differentiated, intermediate-differentiated, poorly-differentiated, and retiform.[13]

RIH in a patient with both DSD and an SLCTs is exceedingly rare. This report aims to summarize the clinical characteristics and highlight diagnostic considerations associated with such cases.

First, surgeons should remain vigilant when ancillary test results reveal abnormalities—particularly when these findings are inconsistent with the patient's medical history. For instance, in the present case, imaging indicated absence of structures such as the uterus, despite no prior history of hysterectomy. Such discrepancies should not be disregarded merely because they fall outside the primary focus of the treating department. On the contrary, clinicians should investigate the underlying causes and seek multidisciplinary consultation when necessary to ensure diagnostic accuracy. Furthermore, when adnexal tissues are encountered within a hernia sac, heightened suspicion is warranted. Although these may represent normal anatomical structures, the possibility of malignancy must be considered—especially in individuals with atypical gynecological or reproductive histories. In female patients, the decision to reduce or resect such tissues should be guided by their vascular status. Particular caution is advised when tissues exhibit abnormal morphology, as neoplastic lesions may impact both prognosis and long-term quality of life.

In patients considering future childbearing, genetic testing is a valuable tool for assessing hereditary disease risk. For example, in 46,XX testicular DSD, and *SRY*-positive cases typically arise *de novo* due to aberrant chromosomal translocation, rather than being inherited. In contrast, pathogenic variants in NR5A1 are inherited in an autosomal dominant manner.[14] Genetic testing plays a crucial role in assessing disease

risk and informing clinical management. Most SLCTs are associated with mutations in the *DICER1* gene. For individuals carrying such mutations, regular imaging and long-term surveillance are recommended due to the elevated risk of developing additional *DICER1*-related neoplasms.[15]

A careful review of her history revealed that at her initial visit 18 months earlier, she had no prior abdominal surgeries. However, the preoperative workup documented an absent uterus, a finding that was insufficiently investigated. Likewise, during the current admission, abnormal imaging results (absence of a uterus and indeterminate hernia contents) were initially overlooked. Fortunately, the surgeon identified an unusual feature within the hernia contents and submitted the tissue for pathological examination. The right gonad showed tumor cells arranged in nests, lobules, and tubular/acinar-like structures, with rare mitotic figures, consistent with a well-differentiated SLCTs. Immunohistochemistry demonstrated positivity for sex cord-stromal markers (inhibin-α, calretinin, SF-1, and WT1), with additional expression of Melan-A, β-catenin, OCT3/4 (weak), CD99, CK7, and PR, thereby confirming sex cord-stromal differentiation. The Ki-67 index was low, supporting the diagnosis of a well-differentiated neoplasm. Chromosomal karyotyping revealed a 46,XY genotype. Collectively, these findings established the diagnosis of a well-differentiated SLCTs arising in the context of a 46,XY disorder of sex development.

Conclusion

Bilateral RIH complicated by DSD and SLCTs is exceptionally rare. This case underscores the importance of three key practices: recognizing and pursuing unexplained imaging abnormalities, analyzing intraoperative findings and initiating timely multidisciplinary consultation when indicated, and performing thorough pathological evaluation of unexpected tissues. We propose that any patient presenting with an unexplained absence of reproductive organ on imaging, especially with no history of surgical removal, should be evaluated for an underlying DSD. Definitive diagnosis relies on pathological evaluation and, when appropriate, chromosomal karyotyping. For masses of indeterminate origin, timely specialist consultation and complete excision of the tissue for pathological analysis are recommended. Accurate diagnosis followed by appropriate treatment is vital to optimize patient outcomes. It is imperative that surgeons remain cognizant of this rare but significant clinical entity. A limitation of this case is that the patient's abnormal clinical signs were not adequately recognized before surgery, and confirmatory genetic testing was not performed.

Author contributions

We certify that we have participated sufficiently in the intellectual content, conception and design of this work or the analysis and interpretation of the data (when applicable), as well as the writing of the manuscript, to take public responsibility for it and have agreed to have our name listed as a contributor.

Ethical policy and institutional review board statement

This report received IRB approval, after obtaining patient consent, from the Ethics Committee of the First Affiliated Hospital of Ningbo University (2025-R114RS) on June 16, 2025. The procedures followed were in accordance with the Helsinki Declaration of 1975, as revised in 2013.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. The patient has given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Data availability statement

Data sharing is not applicable to this article as no datasets were generated and/or analyzed during the current study.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Acknowledgments

The authors would like to acknowledge the patient for allowing this case to be published.

Abbreviations

SLCTs	Sertoli–Leydig cell tumors
DSD	Disorders of sex development
CAIS	Complete androgen insensitivity syndrome
PMDS	Persistent Müllerian duct syndrome

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