Case report

Bell clapper deformity in female?

Jayalaxmi Shripati Aihole

Dept of Paediatric Surgery, IGICH Bangalore, Karnataka, India

ARTICLE INFO

Keywords:
Infantile hernia
Ovarian torsion
Bell clapper
Oophorectomy

ABSTRACT

Introduction: Indirect inguinal hernia is one of the most common congenital anomaly common in males, but not uncommon in females. In females, inguinal hernia mostly indirect, containing ovary presenting as a palpable labial mass is common, resulting in strangulation, torsion, and infertility if left untreated.

Importance: The bell clapper deformity (BCD) is an important anatomical risk factor for intra vaginal testicular torsion, its presence in females is hardly been speculated.

Case presentation: A 6 months female baby presenting with tender labial mass, its management has been discussed here.

Clinical discussion: The bell clapper deformity, in which the tunica vaginalis inserts high on the spermatic cord, leaving the testis free to rotate and get torted, however its presence in females has never been explored.

Conclusion: Bell clapper deformity's existence in females yet to be speculated before exploration.

1. Introduction

Paediatric inguinal hernia have a incidence ranging from 0.8% to 4% with a male to female ratio of 3:1 to 10:1 worldwide. The incidence of an incarcerated ovary with or without fallopian tubes in a female inguinal ovarian hernia via canal of Nuck is between 15 and 43%. The bell clapper deformity, congenital anatomical abnormality present in 12% of males, is an important predisposing factor in testicular torsion in which the tunica vaginalis inserts high on the spermatic cord, leaving the testis free to rotate, however its presence in females has never been described.

2. Case summary

A 6 months old female infant was brought to us with history of painful swelling at the left labia with excessive cry since 3 days, not associated with fever, bowel or bladder disturbances and without any history of alleged trauma (Fig. 1A). Baby was born by full term normal vaginal delivery with birth weight of 2.75 kg to a consanguineously married couple. Mother noticed a small nodular non tender swelling at left labial region after 7 days of life. Since it was not associated with pain or any other symptoms, parents deferred to seek medical attention in view of COVID 19 pandemic.

At 6 months of age, there was sudden onset of pain and redness over the swelling in the left labia associated with excessive cry; baby was brought to the emergency room. Sonography revealed hypoechoic mass in the left labial region suspecting ovarian hernia. On examination there was a tender, nodular swelling of 4 cm × 2 cm in the left labial region associated labial redness and edema. Baby's haematological, hormonal assay and karyotyping were all normal. Immediate inguinal exploration was conducted under general anaesthesia by left inguinal skin crease incision. The inguinal canal was normal without any contents, however there was thick hernia sac passing through superficial inguinal ring into the left labial region containing a pedicled structure (Fig. 1B, C, D, Fig. 2E). Thick hernia sac was opened and torted necrotic gangrenous ovary of around 4 clockwise turns (>720°) with fallopian tubes acting as pedicle around which ovary torted in thick hernia sac, hence excised (oophorectomy) and histopathological evaluation revealed, whole ovarian parenchyma was replaced with hemorrhagic necrosis with inflammation (Fig. 2F, G). The hernial sac showed epithelium with blood vessels along with inflammation (Fig. 2H). Baby is doing well on follow up.

3. Discussion

The developing ovaries as they descend from their original position from level of the aorta below the renal arteries into the pelvis as the fetus grows, carries with them a tongue or diverticulum of parietal peritoneum (saccus vaginalis or canal of Nuck). The gubernaculum ovarii, a fibrous band from the lower pole of the ovary ends close to the external inguinal ring on each side, and becomes the ovarian suspensory ligament cranially and the round ligament caudally, extending up to labia majora. A saccus vaginalis extending into the inguinal canal (canal of
Nuck), normally gets obliterated, but however may remain patent after birth and provide a canal for potential indirect inguinal hernia [1–4].

The inguinal canal in females normally transmits the round ligament of the uterus and the ilioinguinal nerve to the labia majora, a vein and an artery from the uterus that forms a cruciate anastomosis with the labial arteries. Incarceration of the bowel and female adnexa may occur especially in infants with inguinal hernia, due to a relatively short and oblique direction of inguinal canal and presence of a diverticulum or

Fig. 1. A—Clinical appearance of left labial swelling  
B—Intra operative picture of left direct hernia sac with gonad in it.  
C—Intra operative picture of opened thick left direct hernial sac; blue arrow pointing at infantile uterus  
D—Left ovarian torsion with necrosis and gangrene; blue arrow pointing at infantile uterus. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Fig. 2. E—line diagram of the pathological anatomy of bell clapper deformity.  
F and G—Histopathological picture of left ovarian parenchyma replaced by hemorrhagic and necrotic areas with few congested vessels.  
H—direct hernial sac with its lining epithelium, vessels and fibro collagenous tissue along with dema and inflammatory infiltrates in its wall.
canal of Nuck, with the round ligament [1–4].

In BCD, the tunica vaginalis covers the entire testicle and inserts high on the spermatic cord, allowing the testis to swing and rotate freely within the processus vaginalis, condition similar to the clapper inside a bell presenting as acute scrotum and acute abdomen in males. Similarly in females, if the ovarian vascular pedicle is narrow at the internal inguinal ring and the size of the ovary is relatively bigger, results inadequate fixation of the ovary and hence tending toward torsion easily [3–5].

A tender labial mass should raise the suspicion of an incarcerated viscus with ovary in female infants. Unlike inguinal hernias involving testes, strangulation by surrounding loops of intestine within the hernia sac is not thought to be the common mechanism of ovarian necrosis [3,4].

It has been suggested that herniotomy decreases the risk of torsion to the other ovary by returning it to a normally-fixed intra-abdominal position [3–5].

Differential diagnosis for female inguinal hernia though rare includes hydrocele, lymphadenitis, abscess, soft tissue tumours, metastatic tumours, epidermoid cysts, cystic lymphangioma and femoral hernia [3–5].

Author is reporting here a 6 months old baby having a left labial mass since 7 days of life without any symptoms, presenting suddenly with acute tender left labial mass since 3 days, when explored urgently, found a direct hernia via superficial inguinal ring containing torted ovary with the fallopian tube acting as pedicle within a thick saccus vaginalis, hence resulting left tubo ovarian necrosis and gangrene with normal inguinal canal.

Apart from the direct hernia occurring through superficial inguinal ring forming a thick hernial sac having no weakness of the posterior wall of inguinal canal, directing to the labial region containing a bigger ovary, hence predisposing to its torsion (BCD), in itself a rare embryological variant to be reported. In view of its rarity of embryology, surgical anatomy and mechanism of occurrence, similar to bell clapper deformity in males, this case has been reported(Fig. 2E).

4. Conclusion

Bell clapper deformity and its consequences has been well established in males, however its presence in females has yet to be explored to avoid catastrophic consequences especially in chronic female inguinal hernias.

The work has been reported in line with the SCARE 2020 Criteria [6].

Provenance and peer review

Not commissioned, externally peer-reviewed.

Sources of funding

None.

Ethical approval

Yes.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

None.

Guarantor

Dr Jayalaxmi Shripati Aihole.

CRediT authorship contribution statement

Dr Jayalaxmi Shripati Aihole.

Declaration of competing interest

None.

Acknowledgement

Author would like to thank all her paediatric surgical colleagues and all the staffs of IGICH, Bangalore, Karnataka, India.

References

[1] J.S. Aihole, The demographic profile and the management of infantile inguinal hernia: a 3-year’s review, Afr. J. Urol. 26 (2020) 28.
[2] J.S. Aihole, N.M. Babu, D. Javaregowda, Santankrishna R, JadharV, Acute scrotum: a rare presentation, Int. J. Case Rep. Images 6 (2015) 267–271.
[3] J.S. Aihole, N.M. Babu, D. Haldar, R. Sahadev, Acute abdomen caused by torsion of an undescended testis: a rare presentation and its diagnostic dilemma, Paediatr. Urol. Case Rep. 4 (2017) 369–372.
[4] S.J. Boley, D. Cahn, T. Lauer, G. Weinberg, S. Kleinhaus, The irreducible ovary: a true emergency, J. Pediatr. Surg. 26 (1991) 1035–1038.
[5] V. Dogra, deformity B-clapper. Bell-Clapper Deformity, AJR Am. J. Roentgenol. 180 (2003) 1176–1177.
[6] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 Guideline: Updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.