CASE REPORT

Unilateral tubo-ovarian agenesis with contralateral adnexal torsion in a premenarchal girl

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SUMMARY

Unilateral adnexal agenesis is a rare entity. Most of these cases are reported in reproductive age group. A case of a 10-year-old premenarchal girl is reported who presented with acute abdominal pain and vomiting. Both Ultrasonography (USG) and MRI suggested right ovarian mass with torsion. Laparoscopic evaluation revealed an enlarged right ovary with torsion and contralateral agenesis of uterine tube and left ovary. Detortion of the ovary and an ovarian fixation to the ovarian pedicle was done. This is the first reported case of a premenarchal girl presenting with an acute abdomen due to adnexal torsion along with contralateral tubo-ovarian agenesis.

BACKGROUND

Congenital unilateral absence of fallopian tube with ipsilateral ovarian agenesis is a rare occurrence with a suggested incidence of 1:11240.¹ The proposed aetiologies for ovarian agenesis or hypoplasia in young girls is a developmental defect or an early torsion of the ovarian pedicle, either during fetal or early neonatal life, resulting in ischaemia and atresia of the relevant adnexa.² This is the first case reported where a premenarchal girl presented with acute abdominal pain due to ovarian torsion along with contralateral tubo-ovarian agenesis.

Figure 1 Sagittal T2 weighted image of pelvis showing enlarged right ovary with peripherally displaced follicle (arrow).

Figure 2 Axial T2 weighted image of pelvis showing enlarged right ovary with peripherally displaced follicles (small arrow) and thickened tortuous pedicle (big arrow).

CASE PRESENTATION

A 10-year-old girl presented to the emergency department with an acute abdominal pain, localised to the right iliac fossa with one episode of vomiting. There was no bladder, bowel complaint or history of fever. She had not attained menarche. Her height was 127 cm and weight was 30 kg (body mass index (BMI): 18.6 kg/m²). Thus, according to BMI, her growth corresponded to 85th centile of WHO growth chart for girls. Her pulse rate was 80/min and blood pressure was 92/60 mm Hg. Her abdominal examination revealed tenderness and rigidity in right iliac fossa. Her breasts, axillary and pubic hairs corresponded to Tanner stage 1. The examination of the external genitalia was unremarkable. Rectal examination was not conducted as the patient did not allow the rectal examination.

Figure 3 Laparoscopic image of absent right fallopian tube and ? streak ovary in the ovarian fossa.

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Figure 4 Twisted and enlarged left adnexa.

INVESTIGATIONS

Her blood investigations were within normal limits with haemoglobin of 12.6 g% and total leucocyte count of 7600 cells/mm³. Her Thyroid Stimulating Hormone (TSH) was 2.3 mIU/L. Her abdominal ultrasound revealed a well-defined right adnexal hypoechoic lesion with multiple fine internal septae and altered signal intensity lesion of approximately 6.7×5.3×3.8 cm in size. Uterus was normal and left ovary could not be appreciated. MRI of abdomen revealed enlarged right ovary with peripherally displaced follicles and thick tortuous pedicle suggesting ovarian torsion (figures 1 and 2). Bilateral urinary tract and the kidneys were normal in size, shape and location. Her karyotype was normal.

Differential diagnosis

Twisted ovary, acute appendicitis and intestinal intussusception.

TREATMENT

Laparoscopic examination was conducted and revealed a normal uterus with an absent left uterine tube and an empty left ovarian fossa.

There was whitish area in the ovarian fossa giving suspicion of a streak ovary (figure 3). The right ovary was enlarged at a size of 8×6 cm size, with a torsion present at the base involving the tube also (two twists as shown in figure 4), no cysts or masses were found. The right ovary was healthy looking (not gangrenous) so detorsion of the pedicle was performed. As repeated torsion was occurring following detorsion, ovarian fixation was done (figure 5).

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful, and the patient was discharged on day 5 postop in clinically stable condition. Her 6-month follow-up proved clinical and radiological improvement

Figure 5 Ovary was fixed to ovarian ligament.

Learning points

- Possibility of ovarian torsion should be considered in a premenarchal girl presenting with abdominal pain and vomiting.
- High index of suspicion and early intervention by laparoscopy can preserve the ovarian function.
- Oophoropexy is a good option to prevent the recurrent ovarian torsion in the premenarchal girls.
- MRI is a good modality for diagnosis of ovarian torsion in these premenarchal girls.¹³

as the patient denied recurrence of symptoms and ultrasound examination of the abdomen revealed regression of the right ovary's size to normal parameters.

DISCUSSION

The unilateral adnexal agenesis without uterine anomaly is a rare condition. Most of the cases of unilateral ovarian agenesis have been reported in association with uterine or urinary tract abnormality.^{3–6} Chen has reported 25 cases of unilateral adnexal agenesis from literature, but all the cases have been reported in a cohort of females of reproductive age.⁷

The agenesis of adnexa in the presence of Mullerian anomalies supports the theory of embryological defects in the development of genital ridge. Other plausible theory for adnexal agenesis postulated by few authors is inadequate blood supply to the caudal portion of Mullerian duct leading to malfunctioning of the autocrine and paracrine signalling.⁸ The present case could be due to a developmental defect of the urogenital ridge which can explain the affected ipsilateral uterine tube.

The adnexal agenesis, along with torsion of the contralateral ovary and fallopian tube in the present case, is difficult to explain. Paediatric ovarian torsion is a rare entity, and the estimated prevalence between 1–20 years of age is 0.005% compared with 2.5%–7.5% in the adult population.⁹ It has been reported that most of the premenarchal ovarian torsion cases present with abdominal pain and vomiting similar to this case.¹⁰ In a study that analysed 16 cases of premenarchal ovarian torsion cases, it has been found that most of the cases had normal-size ovary (69.9%).¹⁰ This is in contrary to our case where the ovary was enlarged in the absence of any cysts which could be due to the compensated hypertrophy.

Torsion is more commonly found in association with Mullerian anomalies as there are more chances of abnormal anatomic connection between ovary and other structures. The aetiology of ovarian torsion in premenarchal girls may be explained by elongated utero-ovarian ligament, congested or hypertrophied ovary or sudden rise in intra-abdominal pressure.¹¹ Laparoscopic detorsion is the treatment of choice but there is still no consensus regarding the oophoropexy.¹² Oophoropexy has been done in the present case because of recurrent torsion during the laparoscopic procedure.

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