

Ovarian Torsion in Adolescent with Chronic Immune Thrombocytopenia

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ABSTRACT

Ovarian torsions in adolescence are rarity, particularly bilateral, with mostly unknown etiology. Enlargement of the ovary contributes to torsion. Young girl presenting with abdominal pains, nausea and vomiting was for two days suspected and observed as gastroenteritis. By exclusion of gastroenteritis she was admitted for gynecological work-up. Ultrasound showed significantly enlarged right ovary, with tumor-like appearance. At the laparotomy, gynecologist found torsioned, necrotic ovary and ovariectomy was performed. Histology showed massive stromal bleeding (haemorrhage). Asymptomatic enlargement of remaining ovary occurred nine months after the ovariectomy. This enlargement was accompanied with platelets' fall and the possibility of repeated torsion impended. Thrombocytopenia was suspected from the first moment, but diagnosed after the surgery. Thrombocytopenia in adolescence requires additional attention as possible cause of intra-ovarian bleeding with consecutive enlargement and may lead to torsion. Oral contraceptives regulate dysfunctional bleeding, decrease ovarian volume and by so, may minimize risk of torsion. This strategy proved effective in the case we present.

Key words: ovarian torsion, adolescent, chronic immune thrombocytopenia

Introduction

Ovarian torsions are rare among children and adolescents¹. Usually, they are unilateral and with unknown mechanism of origin². Approximately half of them occur on the ovary with no pathological ground so ever. The other half is caused by functional cyst or tumor³.

The continuous, slow growth of both ovaries, during childhood and puberty is a normal state. Ovaries enlarge, increase in weight, and change the shape simultaneously. The mean volume increases from 0.7 cm³ at age 10 years, to 5.8 cm³ at age 17 years⁴. Conditions accompanied with enlarged ovarian size, mass or volume, increase the risk of torsion as well. As the symptoms are often unspecific, in such manner they lead to diagnostic delay and present a potential danger for the ovary.

One of the causes of ovarian torsion in adolescence is PCOS (polycystic ovary syndrome). In the study by Shah AA et al., authors analyzed ovarian torsions in premenarcheal girls. The contra lateral ovary was imaged preoperatively or inspected during the surgery and used as a

comparison for the torsioned one⁵. Almost all observed patients had both ovaries enlarged, and the peak volume of untorsioned polycystic ovary was 28.5 cm³.

Immune thrombocytopenic purpura (ITP) is a rather frequent autoimmune bleeding disorder in childhood⁶. Children account for a half of new cases per year. Often ITP is asymptomatic and can be discovered incidentally. The diagnosis is frequently made by exclusion. Visible symptoms of ITP include the spontaneous formation of bruises (purpura) and petechiae (tiny bruises), especially on the extremities, bleeding from the nostrils, bleeding at the gums, and menorrhagia, any of which may occur if the platelet count is below 20 × 10⁹/L. A very low count (<10 × 10⁹/L) may result in the spontaneous formation of hematomas in the mouth or on other mucous membranes. Bleeding time from minor lacerations or abrasions is usually prolonged. Serious and possibly fatal complications due to an extremely low count (<5 × 10⁹/L) may include subarachnoid or intracerebral hemorrhage,

lower gastrointestinal bleeding or other internal bleeding (i.e. intra-ovarian). Almost 70 percent of childhood cases end up in remission within six months, even without therapy. A third of the remaining cases usually remit during follow-up, another third develops only mild thrombocytopenia with platelets count above $50 \times 10^9/L$. Treatment is generally indicated with platelet count below $20 \times 10^9/L$. Treatment includes corticosteroids, immunoglobulin, Anti-D, immunosuppressants, thrombopoietin receptor agonists, splenectomy, eradication of *H. pylori* infection, platelet transfusion, dapsone, rituximab⁶. We strongly suspect that ITP with extremely low platelets was the reason of intra-ovarian bleeding, which caused enlargement and ended with torsion.

To our best knowledge we are presenting a first report of ovarian torsion in an adolescent patient with ITP.

Case Report

A thirteen-year-old girl was admitted to hospital with diffuse abdominal pains and abdominal wall tension, accompanied by nausea and vomiting for past two days. Few days she was observed as suspected gastroenteritis. At the admission to hospital, mother said the girl had occasionally petechiae on the skin and ecchymoses on mucous membranes and asthma at the effort. Menarche started at the age of 12 and in a past year she had regular cycles, every 28–30/5 days. Three months before torsion, menstrual bleeding patterns were heavier. Hormonal values were suggestive on PCOS: FSH 5 IU/L, LH 10 IU/L, Testosterone 3 nmol/L. At the day of admission, platelet count was slightly lower, $105 \times 10^9/L$ (normal range $158–424 \times 10^9/L$). Other blood analysis – electrolytes, liver, kidney, coagulation tests were within normal range. Ultrasound measured right ovary $8 \times 5 \times 4$ cm, with dense appearance and significantly bigger volume (86.68 cm^3) for age. Volume of the left ovary was 10 cm^3 , with numerous <9 mm follicles. Two out of three criteria according to Rotterdam consensus for PCOS, were positive⁷. But, for a PCOS to be diagnosed, recent guidelines demand 2 consecutive years with continuous symptoms, following menarche. Consequently, the PCOS was not indisputable diagnosed.

Ovarian torsion was presumed and the surgery was performed promptly. Laparotomy showed livid, three-times twisted right ovary. The tissue was necrotic, decomposed and it was bleeding during manipulation, also after the detorsion attempt. As a result, decision was to perform oophorectomy. Macroscopically, at the cross section ovary was imbued with blood. Pathohistological diagnosis was »Right ovary gangrene due to torsion with massive stromal bleeding, no visible cyst« (Figure 2).

Left ovary was found of normal size, with smooth surface and without signs of previous ovulations. On the fifth postoperative day she was discharged from hospital and suggested home care. Platelet count was within normal range $154 \times 10^9/L$.

Three months later, at the routine follow-up, platelet count dropped to $49 \times 10^9/L$, the girl had no symptoms and was without skin changes (Figure 1).

Dozen days later she presented to hematologist febrile, with platelet count $20 \times 10^9/L$. Bone marrow analysis, protein electrophoresis, serology, immunology and genetic analysis on thrombophilia were all negative.

One month later, petechiae were observed on skin and mucous membranes, platelets were $6 \times 10^9/L$. She was admitted to the pediatric department, platelet count rose spontaneously – without treatment. Diagnosis of chronic ITP was unmistakable confirmed. After that she was twice admitted to the hospital, platelet count varied from 6 to $67 \times 10^9/L$.

ITP was to be treated by immunoglobulin and methylprednisolone.

Nine months after the surgery gynecological examination with ultrasound was proposed because of mild dysfunctional bleeding. At this time platelet count was $123 \times 10^9/L$. Ultrasound revealed left ovary of homogeneous appearance, threefold enlarged, the volume was 39.20 cm^3 . No ovarian abnormality was noticed. Tumor markers (hCG, AFP, CEA, Ca 125) were all within normal range.

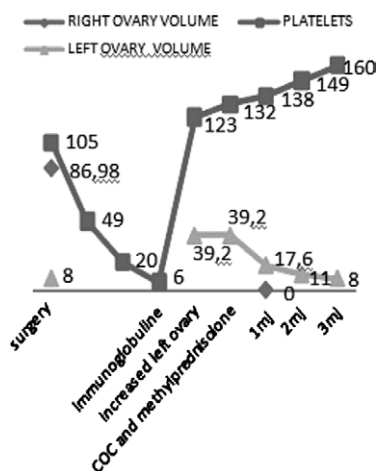


Fig. 1. Comparison of blood platelets with volume of the ovaries.



Fig. 2. Pathohistological section of right ovary.

Such significant volume enlargement correlated with another extreme fall of platelet count to $6 \times 10^9/L$.

Pediatrician and pathologist were consulted for opinion about etiology of such volume increase (4 times larger than expected for PCOS, 8 times larger than normal for the age), as well as because of possible recurrent torsion and loss of the remaining ovary. Conclusion was that earlier attack(s) of ITP, which lasted few months and preceded the torsion of right ovary, were not recognized in the right time and therefore not adequately treated.

We decided to prescribe combined oral contraception (COC) (20 μ g ethinyl-estradiol and 3 mg drospirenone) in intention to decrease ovarian volume.

As expected, 1-, 2- and 3-months later follow-up stated progressive decrease in ovarian volume from 17.6 cm³ to 11.3 cm³ and 8 cm³, respectively. As for the ITP, platelets raised to $160 \times 10^9/L$.

Discussion

Ovarian torsion with delayed surgical intervention often results with necrosis and leads to ovariectomy (or adnexectomy)¹. The rate of adnexectomy ranges from 7% to 93%⁸. Generally, the adnexectomy is performed when adnexa are strongly suspected to be necrotic or when there is no blood supply established after the detorsion^{9,10}. Necrosis is difficult to diagnose by naked eye, and there are studies which report of miss-diagnose by the surgeon^{11,12}. The incidence of pulmonary embolism due to ovarian torsion is 0.2%, and does not increase after detorsion^{13,14}. Detorsion is obligatory to be attempted (if possible laparoscopic), particular in children and younger patients¹⁵. If a truly necrotic ovary is left in abdomen, the rare complication is peritonitis and possible systemic infection¹⁶.

Puzzling, ovarian function might be preserved, even in ovaries which appear necrotic^{11,17,18}. A suitable model for surviving of the ovary is transplantation. It has been established in experimental studies that most developing follicles, and up to 50% of primordial follicles, undergo atresia during transplantation, probably due to ischemia. The differential survival of primordial follicles versus developing follicles in grafts is probably related to the lower metabolic rate of the small follicles¹⁹. As for prior, some authors claim that ovarian function has been preserved in 88–100% of the torsions. So the conservative treatment (detorsion) is strongly advised, when possible^{8,15,20}. Other potential complications of conservative procedure

are postoperative fever and finally, repeated surgery²¹. Risks and prevention of possible contralateral torsion are to be taken in consideration, as well²⁰.

Enlargement of the ovary may be due to: congenitally long supportive ligaments, mesovarium and mesosalpinx resulting in hypermobility, as well by undiagnosed PCOS. Unfortunately, in almost 50% of patients enlargement remains by unknown causes.

As for the PCOS as the possible trigger of enlargement, it cannot explain such significant increase in ovarian volume, much larger than published for PCOS⁵. According to actual guidelines, PCOS was not to be confirmed because of the requirement of two consecutive years with symptoms, for diagnose to be certain. Also to be noted is the fact that remaining ovary was not enlarged from the start, as described in the study of torsions with PCOS⁵.

In case we are presenting, bruises and petechiae occasionally appeared together with profuse menstrual periods before torsion of the right ovary. The same symptoms preceded enlargement of the left ovary, but the chronic ITP was verified. Surprisingly, nine months after the surgery, ovarian volume was 39.20 cm³, with no other sign of symptom.

Finally, as a result of sequence and development of symptoms, the changes in laboratory findings, the growth of platelets adjacent to fall of ovarian volume as a result of treatment (corticosteroids and COC), we strongly suspect ITP as the reason of ovarian enlargement.

The bleeding within ovary was most certainly caused by low count and consequently impaired platelet function, as mentioned earlier. In these patients such dangerous bleeding may be caused by corpus rubrum – luteum, as well.

Thrombocytopenia in adolescent girls requires attention as a potential cause of fatal ovarian torsion, especially in presumption of adjacent PCOS. Such girls have a predisposition for asymptomatic ovarian enlargement correlated to attack(s) of thrombocytopenia and they have high risk of possible and recurrent torsion. Therefore, we recommend regular examinations by gynecologist. With acknowledgement of ITP, we propose treatment with oral contraceptives to reduce ovarian mass and volume, as well as to regulate periods and bleeding. Decreased ovarian volume may diminish and perhaps alleviate the risk of torsion. Thus we may prevent considerable reproductive and emotional loss²².

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TORZIJA JAJNIKA U ADOLESCENTICE S KRONIČNOM IMUNOM TROMBOCITOPENIJOM

SAŽETAK

Torzije adneksa u adolescentica, a osobito bilateralne, rijedak su entitet. U skoro polovice pacijentica nepoznate su etiologije. Sva stanja koja dovode do povećanja veličine jajnika stvaraju i predispoziciju za torziju. Trinaestgodišnja djevojčica s difuznom boli u abdomenu, mučninom i povraćanjem, bila je dva dana opservirana kao gastroenteritis. Po prijemu na ginekologiju ultrazvukom je nađen značajno uvećan desni jajnik, te je na laparotomiji zbog torzije i nekroze napravljena ovariektomija. Patohistološka dijagnoza bila je masivno krvarenje u stromu. Iako su trombociti kod prijema bili ispod normale, kronična trombocitopenija (ITP) je dijagnosticirana postoperacijski. Devet mjeseci nakon ovariektomije, a nakon registriranog pada trombocita, ultrazvukom je verificirano asimptomatsko značajno povećanje volumena preostalog ovarija i time je nastala opasnost od ponovljene torzije. Trombocitopenija zahtijeva dodatnu pažnju kao mogući uzrok povećanja i posljedične torzije jajnika. Liječenje tih pacijentica oralnim kontraceptivima (uz terapiju kortikosteroidima ili/i imunoglobulinima) može regulirati menstruacijska krvarenja, smanjiti volumen ovarija i eliminirati rizik pogubne torzije.