

Ovarian cyst formation and congenital absence of the inferior vena cava: case report

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Summary

Congenital absence of the inferior vena cava (IVC) is a rare condition that presents clinically as recurrent venous thromboses and leg ulcers. We report an association with painful ovarian cysts in a 25 year old woman. The possible pathophysiology and unique management issues posed by this case are presented.

Key words: Ovarian cyst; Absent; Inferior Vena Cava.

Introduction

Anomalies of the inferior vena cava manifest as absence, agenesis, and/or interruption of a particular segment (infrahepatic, prerenal, renal, or infrarenal) [1]. Eighty-one cases of segmental absence have been reported in the English literature [1]. Of these, 89% had absence of the suprarenal segment (associated with congenital cyanotic heart disease and congenital anomalies of the upper abdominal organs), 6% had absence of the renal portion and 6% had complete absence of the inferior vena cava.

One quarter of the patients are diagnosed incidentally. Only 10% are symptomatic with recurrent venous thromboses and leg ulcerations due to the venous stasis [1]. To correct this, surgical venous grafting has been performed but is an extensive procedure with substantial risks [1].

Case Report

A 25-year-old woman was referred to our tertiary care institution for management of painful bilateral ovarian cysts.

At age 23 she experienced sudden swelling of her left leg whilst using combined oral contraceptive pills. Doppler ultrasonography confirmed deep venous thrombosis (DVT). Heparin anticoagulation was started and then switched to warfarin. The contraception was stopped. Despite anticoagulation, thrombosis recurred in the same leg.

Ultrasound and computer tomography showed only a short segment of inferior vena cava (IVC) above the common iliac veins with complete absence of the abdominal IVC. The venous return was predominantly through the vertebral veins. Above the diaphragm the IVC was normal. Serum C and S and antithrombin assays were normal. Lifelong full warfarin anticoagulation was instituted. Furosemide for pitting leg edema and a progestin - only pill for contraception were prescribed.

She was then admitted for 10 days to a community hospital with an acute exacerbation of bilateral dull pain that required

parenteral analgesia. Pelvic ultrasonography showed a simple cyst of 6.7 cm diameter on the right ovary and a simple cyst of 5.4 cm on the left ovary. The pain decreased and at an interim scan, 4 weeks afterwards, the cysts were 5.7 cm and 4.4 cm.

When referred to us 6 weeks later she had a continuous dull pelvic ache and irregular vaginal bleeding with passage of clots. Tibial pitting edema was present. Bimanual palpation disclosed a tender cystic right ovary. Vaginal probe ultrasonography identified a normal left ovary and a simple 4 cm cyst on the right ovary. It was recommended that she continue the progestin - only pill. Other management options were discussed with her in the event of recurrence of her symptoms.

She presently has mild intermenstrual bleeding and transient ovarian cyst formation.

Discussion

In this case, we hypothesize that the ovarian cysts may have formed because of sporadic ovulation (possibly due to lack of compliance in taking the progestin - only pill). The pronounced fluid transudation that occurs during the vasopermeable phase of follicular growth may have been exacerbated due to excessive vascular stasis. Venous stasis may also predispose to cyst formation in ovaries surgically translocated with a tenuous blood supply in women scheduled for pelvic radiation.

In that her cysts resolved spontaneously, continued oral progestin contraception with strict compliance has been advised. Continued suppression of ovulation is considered essential to prevent both the formation of ovarian cysts and the potential for pain, hemorrhage, rupture and torsion, and pregnancy. Pregnancy in such women is complicated by stasis-induced pain and fetal growth restriction [2].

Planning for any recurrence of ovarian cyst formation has also been instituted. Surgery should, in general, be avoided because of risk of vascular injury to extensive collateral venous channels with the superimposed anticoagulated state. Nevertheless, thermal endometrial ablation could safely remedy the uterine bleeding.

If the progestin only contraception again allows ovulation, suppression of ovulation could be provided, at least temporarily, with intranasal gonadotropin releasing hormone. Long-term use is precluded because of osteoporosis and adding back estrogen therapy is contraindicated due to risk of thrombosis. Other oral progestins such as medroxyprogesterone acetate and megestrol acetate are considered to be reasonable alternatives. Intramuscular administration of any of these therapies is considered inadvisable because of risk of hematoma. Combined oral contraception in the anticoagulated state usually carries little risk of thrombosis [3]. However, this woman has already incurred a DVT despite adequate anticoagulation, thus precluding its use.

This rare case poses a unique set of clinical management issues as well as providing some insight into ovarian cyst pathogenesis.

References

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