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CASE REPORT

Spontaneous OHSS in a Young Adolescent: A Diagnostic Dilemma

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Introduction

Ovarian hyperstimulation syndrome (OHSS) is a devastating complication seen in patients undergoing ovulation induction, usually with gonadotropins. Rarely, young patients may present with a spontaneous OHSS. This has been reported in cases of gonadotropin-secreting pituitary adenoma, primary hypothyroidism and at times even in normal pregnancy [1, 2]. Here, we report a case of a young adolescent presenting with bilateral large ovarian masses with multiple differential diagnosis but eventually detected as a case of spontaneous OHSS secondary to hypothyroidism.

Case Report

An adolescent who was 10 years and 8 months old presented to us with chief complaints of severe pain in abdomen since 1 week and amenorrhea since 6 months. There was no history of headache, vomiting, visual disturbances or seizures. She attained her menarche at the age of 10 years and had two cycles of bleeding at an interval of 15 days. On examination, she was well built with a height of 137 cm, weight of 47.2 kg, pulse rate of 96/min, BP of 100/70 mmHg, cushingoid face with thick skin and pallor was present; however, there was no edema. As per Tanner's staging, her sexual maturation score was B3 and P3 for breast and pubic hair, respectively, and sparse axillary hair were present. On per abdominal examination, abdomen appeared distended below umbilicus and a 24-week irregular mass was felt arising from the pelvis.

Investigations revealed hemoglobin of 11.5 g/dl, total leukocyte count of 9500 and ESR of 53. Hormonal investigations were performed with LH < 0.09 IU/L, FSH 9.46 mIU/L, prolactin 55.42 ng/ml, ACTH 13.2 pg/ml, DHEAS 17.1 mcg/dl and GH 0.27 ng/ml. Her thyroid function test showed FT3-135 ng/dl, FT4-8.4 ng/dl and TSH-1.82 µIU/ml. Elevated prolactin levels prompted an MRI brain which revealed a diffusely enlarged homogenously enhancing pituitary with a convexity along superior margin suggestive of a possibility of pituitary macro-adenoma or pituitary hyperplasia. As the gonadotropin values being in the normal range and lack of associated symptoms, the possibility of pituitary adenoma was ruled out.

Ultrasound revealed markedly enlarged ovaries with multiple cystic spaces with right ovary measuring 14.6 × 10.7 × 9.1 cm and left ovary 10.4 × 7.3 × 5.8 cm. However, on Doppler, fairly high-resistance flow was noted. MRI pelvis was suggestive of bilateral multiloculated cystic adnexal lesions with differential diagnosis of mucinous cystadenoma, endometriosis or hyperstimulated ovaries. Serum tumor markers done to rule out malignancy were CA-125 72.3 U/ml, alpha-fetoprotein 5.47 ng/ml and LDH 885 IU/L (Fig. 1).

Despite high LDH and CA-125 levels, the radiological findings were not coherent with malignancy. Also the characteristic soap bubble appearance noted on ultrasound was suggestive of the probability of ovarian hyperstimulation. Hence, due to high clinical suspicion, a repeat thyroid profile was done and values noted were as follows: FT3 50.08 pg/ml, FT4 1.97 ng/ml and TSH > 500 µIU/ml. TPO antibody level was 897 IU/ml, and thyroglobulin antibody 11.9 IU/ml.

A diagnosis of spontaneous OHSS secondary to hypothyroidism was made, and she was started on levothyroxine 75 µg. Subsequent ultrasonography done after 3 months revealed a significant decrease in the ovarian size and

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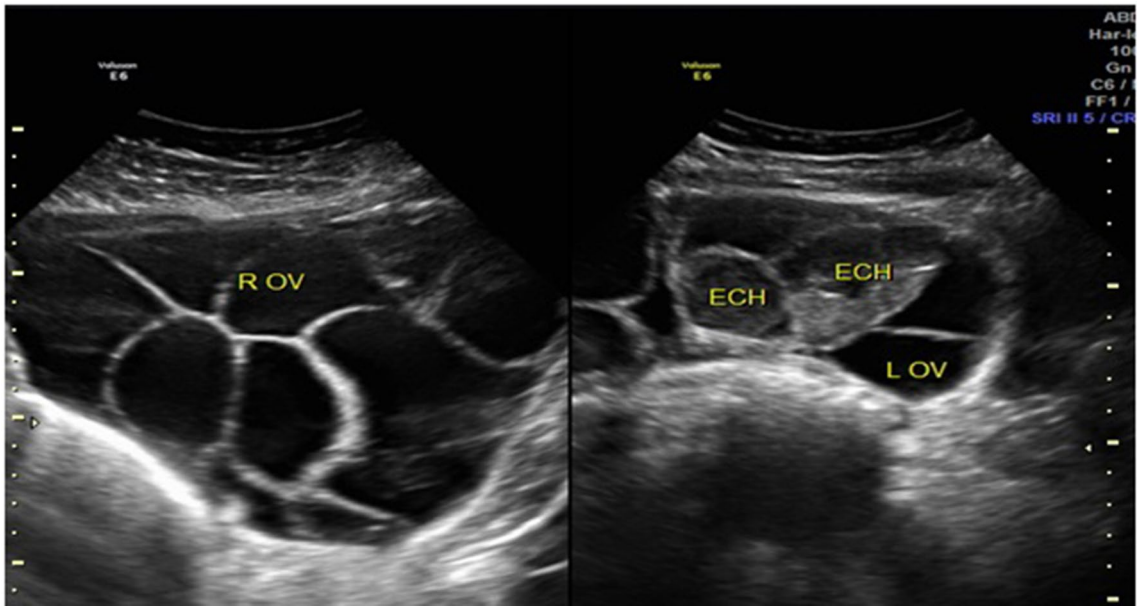

Fig. 1 Ultrasonographic image of bilateral ovaries showing classical soap bubble appearance

volume, right ovary measuring 3.6 × 3.3 × 3.1 cm (vol. 18.4 cc) and left ovary measuring 3.5 × 3.3 × 3 cm (vol. 17.3 cc). There was also a complete resolution of her symptoms. Repeat thyroid profile done after 3 months showed values in the normal range (FT3-5.97 pg/ml, FT4-1.41 ng/dl and TSH-2.20 µIU/ml) (Fig. 2).

Discussion

Spontaneous OHSS is an uncommon clinical entity occurring due to non-iatrogenic aetiologies. De Leener et al. described a classification system of spontaneous OHSS which divides it into three types:

Type I Related to mutations in the FSH receptor.

Type II Secondary to elevated levels of human chorionic gonadotropin (HCG) hormone seen in hydatidiform mole, multiple pregnancies and polycystic ovarian patients who are pregnant.

Type III Due to primary hypothyroidism.

The exact pathogenesis of this entity is complex. In type III patients, long-standing deficiency of thyroid hormone induces hyperplasia of the lactotropic as well as thyrotropic cells in the pituitary gland leading to elevated levels of prolactin and thyroid-secreting hormone (TSH). The beta subunit of TSH has identical configuration with that of FSH; hence, it stimulates the FSH receptor. In addition, it leads to pituitary hyperplasia which may mimic a pituitary adenoma. Also, there is a preferential formation of estriol which is a weak form of estrogen leading to over-production

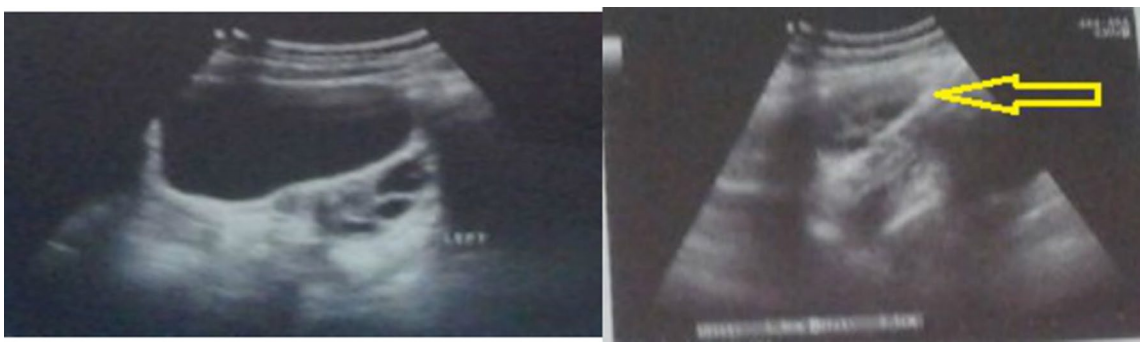

Fig. 2 Ultrasonographic image of ovaries 3 months post-thyroid supplementation, demonstrating a marked decrease in ovarian volume on either side. The yellow arrow depicts the right ovary

of gonadotropins. All these can contribute to the hyperstimulation of ovaries.

The clinical symptomatology can vary depending upon the severity of OHSS and occurs primarily due to extravascular accumulation of fluid. Hormonal evaluation in these patients reveals a markedly elevated TSH and raised anti-TPO-antibodies suggestive of autoimmune involvement. The treatment is often simple and involves a dramatic response to thyroid supplementation [3].

The suspicion of malignancy is high in a young adolescent presenting with large bilateral ovarian masses. Germ cell tumors are more common in this age group. Serum tumor markers and specific radiological findings like the presence of solid areas and ascites can aid in the diagnosis. Van Wyk-Grumbach syndrome is also an important differential diagnosis in these cases and is characterized by isosexual precocious puberty, juvenile hypothyroidism, a delayed bone age and the presence of ovarian cysts [4]. Pituitary adenoma can also present with similar complaints; however, characteristic features like visual field defects, headache, galactorrhea and unexpected bone growth help to reach the diagnosis.

A wide array of investigations need to be performed to identify the aetiology of spontaneous OHSS. Ultrasonography in these cases reveals a classical soap bubble or spokes in wheel appearance of the ovarian masses. These represent enlarged multiple follicles arranged peripherally. The final treatment in these cases depends on the underlying pathology. Knowledge of such uncommon conditions among gynecologists, radiologists and primary care physicians is essential to avoid misdiagnosis, unnecessary surgery and loss of ovarian reserve.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflicts of interest.

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