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Ovarian hyperstimulation syndrome in spontaneous pregnancy with sacrococcygeal teratoma complicated by maternal mirror syndrome: A comorbidity

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Abstract Case of ovarian hyperstimulation syndrome (OHSS) in pregnancy is rare. The complications include ascites, thromboembolism, pulmonary and hemodynamic/renal disorders, abortion, intrauterine fetal demise and preterm delivery. In this case, we found OHSS in spontaneous pregnancy with maternal mirror syndrome as complication of sacrococcygeal teratoma that increased maternal morbidity and mortality rate.

1. Introduction

Ovarian hyperstimulation syndrome (OHSS) has been extensively described after treatment with exogenous gonadotropins, clomiphene citrate, and gonadotropin releasing hormone. OHSS, not related to ovulation induction is rare. Spontaneous hyperstimulation syndrome has been reported in women with hypothyroidism, Polycystic Ovarian Syndrome (PCOS) and pregnancy, gonadotroph pituitary adenoma, and normal pregnancy. Sacrococcygeal Teratoma (SCT) is the most common newborn tumor occurring in approximately 1 in every 40,000 live births. Although postnatally diagnosed SCT carries a fairly good prognosis, fetal diagnosis of SCT is associated with extremely high prenatal and perinatal mortality. This report describes a case in which a naturally conceived pregnancy is associated with spontaneous ovarian hyperstimulation syndrome (OHSS) and mirror syndrome as complication of sacrococcygeal teratoma with previous history of polycystic ovary syndrome.

2. Case report

A 27 years old women in first pregnancy was admitted to hospital at 28–30 weeks gestation because of diarrhea and in labor and with history of Polycystic Ovarian Syndrome (PCOS). The patient complained swelling of the body followed by breathing difficulties. The current gestation started spontaneously, and no medications had been taken by the patient during the

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preceding months. On admission physical examination detected mild tenderness and distension of lower abdomen. Pelvic examination found bilaterally enlarged cystic ovaries, and a pregnancy of 28–30 weeks gestation.

Abdominal ultrasound showed a 28–30 week singleton pregnancy and on sonographic findings showed sacrococcygeal teratoma size >10 cm with polyhydramnion; there were bilateral multiloculated ovarian cysts with diameters of 20 × 16 cm (right ovary) and 16 × 10 cm (left ovary) and mild ascites (see Fig. 1).

The placental appearance was normal. Laboratory testing revealed hemoglobin 7.5 gr% and proteinuria (+3). The serum levels of beta-human chronic gonadotrophin (hCG), antithyroglobulin, and anti-peroxydase were not examined because of emergency situation. With these findings, the diagnosis of severe spontaneous ovarian hyperstimulation associated with a singleton intrauterine pregnancy and sacrococcygeal teratoma co-morbid with mirror syndrome was made. This condition leads to a decision for laparotomy and cesarean section, for both diagnosis and treatment. The patient gave birth to a 1100 g baby boy with Apgar score of 3/6 with huge sacrococcygeal teratoma (see Fig. 2).

Aspiration of a large ovarian cyst and wedge biopsy was done in laparotomy. Intraoperative histological examination of frozen section from the ovaries revealed benign cysts. Treatment in hospital given was of bed rest, close monitoring of blood pressure, pulse rate, urine output, hematocrit, electrolytes, coagulation profile, and fluid therapy (see Fig. 3).

Within 2 weeks, remarkable improvement was observed, with resolution of ascites and gradual reduction of ovarian size. The patient was discharged after 2 weeks. The patient's condition was monitored weekly for 1 month. Ten weeks after delivery, pelvic ultrasound showed normal size ovaries.

3. Discussion

Ovarian hyperstimulation syndrome in spontaneous pregnancy is an extremely rare event. Under certain circumstances such as twin pregnancies, the possibility of its existence may be higher because of higher HCG concentrations during the early pregnancy (1–3).

OHSS has life threatening complications such as venous and arterial thromboembolism. Indeed, Schenker and Ezra reported the death of patients with OHSS to be caused by these complications. Consequently, one must be aware of the rare but possible occurrence of OHSS in spontaneous pregnancy, in order to prevent its complications (4–6).

Spontaneous forms of OHSS generally develop between 8 and 14 weeks of amenorrhea, differing from iatrogenic OHSS, which usually starts between 3 and 5 weeks of amenorrhea. The recent identification of mutations in the follicle stimulating hormone (FSH) receptor gene, which display an increased sensitivity to hCG and are responsible for the development of spontaneous OHSS, helps us to understand this problem (7,8). In iatrogenic OHSS, the follicular recruitment and enlargement occur during the administration of exogenous FSH. In the spontaneous form however, the follicular recruitment and growth occur later through the promiscuous stimulation, by pregnancy-derived hCG, of a mutated FSH receptor that is abnormally sensitive to hCG or a wild type FSH receptor in the presence of abnormally high levels of hCG. Thus, the symptomatology of spontaneous cases of OHSS usually develops at 8 weeks' amenorrhea and culminates at the end of the first trimester of pregnancy (9).

In the literature, different cases were reported in which spontaneous pregnancy with OHSS and hypothyroidism was found together. It was claimed that high levels of thyroid stimulating hormone can stimulate ovaries in women with hypothyroidism and can cause ovarian hyperstimulation (10,11). In our case, hypothyroidism was not present.

Hyperstimulated ovaries release a number of vasoactive mediators under the influence of hCG. These include vascular endothelial growth factor (VEGF) and several pro-inflammatory cytokines that interact to produce the characteristic pathophysiology of OHSS. This is marked by increased capillary, permeability, leakage, of fluid from the vasculature, third space fluid accumulation and intravascular dehydration (1).

Different classification systems for OHSS have been proposed, which generally identify a mild, moderate, and severe subtype with varying internal grades of severity. In order to simplify the classification of OHSS, we used a classification system adapted from that proposed by the Royal College of Obstetricians and Gynecologists in 2006 (12,13). We identify mild OHSS, in which patients have abdominal bloating and mild abdominal pain; moderate OHSS, characterized by nausea, vomiting, moderate abdominal pain, and ultrasound evidence of ascites; severe OHSS, identifiable by clinical ascites, oliguria, hematocrit >45%, and hypoproteinemia; and critical OHSS, with tense ascites, oliguria or anuria, hematocrit >55%, and white blood count >25,000.

The management of OHSS is tailored to the degree of severity. Early recognition and prompt appropriate treatment will avoid serious sequelae. Severe OHSS requires hospital admission and prompt management to replace lost intravascular volume and prevent its potentially fatal complications namely renal failure and thromboembolic events (14). These patients should be closely monitored to ensure they do not progress into the critical category. In patients with significant ascites, paracentesis is helpful by decreasing intra-abdominal pressure and improving renal blood flow with a subsequent increased production of urine (15,16). Drainage of ascitic fluid in OHSS may be carried out abdominally or vaginally.

The vaginal route has the benefit of easier access and avoidance of ovarian trauma (17). Pleural effusions are not uncommon in OHSS. Thoracocentesis may be necessary to avoid respiratory distress, accompanied by paracentesis in order to prevent fluid from leaking back into the pleural cavity (18).

Medical treatment, undertaken in first line, may be insufficient. In these cases, invasive treatment, using surgical techniques, becomes necessary. Wedge resection of approximately one-third of the ovaries has been reported in two patients who had failed to respond to albumin infusion and paracentesis (19). In this case a recovery in the ascites, urine output and biochemical parameter was observed. However, surgical intervention should be regarded as a last resort and only considered in consultation with senior clinicians familiar with managing OHSS.

This is also an excellent example of how a fetal disorder can lead to maternal symptoms, and by treating this disorder it results in resolution of both fetal and maternal symptoms (Mirror Syndrome).

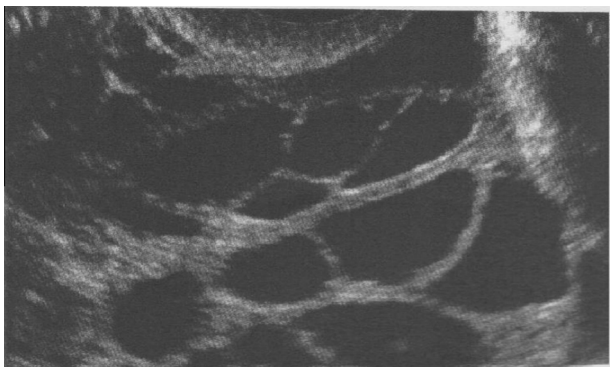

Figure 1 Ultrasound Image of multiloculated ovarian cyst.

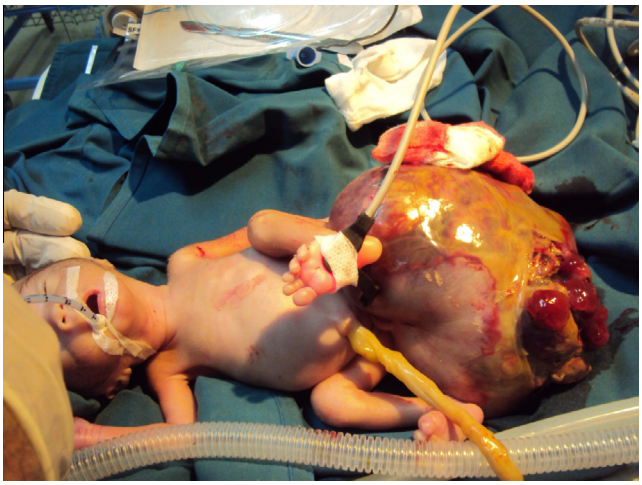

Figure 2 After cesarean section, baby with Sacrococcygeal Teratoma (SCT).

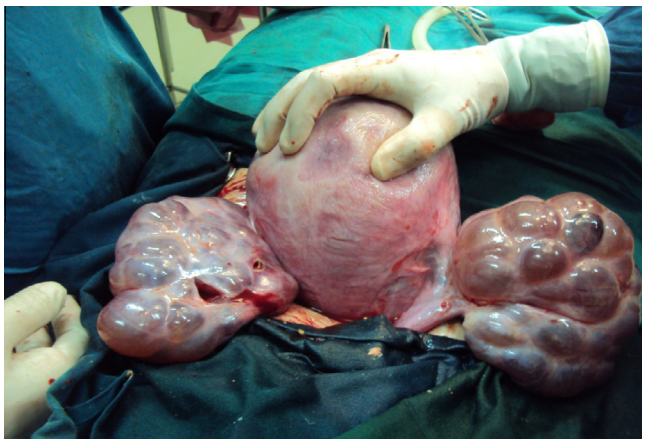

Figure 3 Durante operation, uterus with bilateral multiloculated ovary that shows sign of ovarian hyperstimulation syndrome (OHSS).

The association of SCT with incidence of spontaneous OHSS remain unknown. SCT can be either mature, immature or mixed teratoma. In immature SCT which contain type germ cell tissue that might produce hCG. This will induce OHSS event especially in woman with PCOS. As far as author's knowledge, this might be the first case reported in the literature.

4. Conclusion

The incidence of ovarian hyperstimulation syndrome in spontaneous pregnancy is rare. This case accompanied with maternal mirror syndrome as complication of sacrococcygeal teratoma increased morbidity and mortality rate. The treatment for this ovarian hyperstimulation is same with its iatrogenic case. The screening and prediction of risk factors (young women, PCOS, Multiple pregnancy, hydatidiform mole, and Hypothyroid) are the best recommended for prevention of this case. Expectant mother with history of PCOS and fetus with SCT will exposed to the risk of OHSS due to certain type of SCT which contain germ cell that might produce hCG. Cesarean section is the safest modality to terminate this pregnancy with sacrococcygeal teratoma size >10 cm to prevent further worsening progressivity.

Its comorbidity with maternal mirror syndrome caused by sacrococcygeal teratoma increased morbidity and mortality rate. There is more further investigation required to verify whether sacrococcygeal teratoma can cause ovarian hyperstimulation syndrome in spontaneous pregnancy.

Conflict of interest

There is no conflict of interest.

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