Sacrococcygeal Teratoma associated with Maternal-Mirror Syndrome

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Sacrococcygeal Teratoma (SCT) is one of the most common congenital tumors in newborns. The majority of the cases are type I and II; therefore, the diagnosis of SCT is mostly made in prenatal period especially during second-trimester ultrasound.

This is the first case reported in Bahrain of a pregnant lady in her 20th week of gestation, carrying a fetus with a rapidly growing tumor in its sacrococcygeal area. The woman developed a rare condition of severe pre-eclampsia, cardiomegaly, bilateral bronchopneumonia and pleural effusion suggestive of maternal mirror syndrome (MS).

Early prenatal diagnosis of SCT with early detection of maternal mirror syndrome is extremely challenging.


Sacrococcygeal teratoma (SCT) is considered one of the most common congenital tumors in newborns. In a study review, most of the cases showed benign nature irrespective of size, age, Altman classification or age at diagnosis; recurrence rate is 11% after surgery. SCT is originally a germ cell tumor (GCT) which commonly occurs in infants and young children. A study of SCT showed a birth prevalence of 1 in 27,000 live births. The female: male ratio is found to be 4:1. However, it can be associated with maternal mirror syndrome which is an extremely rare pathological condition; the incidence around the world is less than 100.

Prenatal diagnosis is usually performed during routine second-trimester scanning. However, some cases were reported as early as the first trimester. Most cases were Altman type I and II. If in doubt with neural tube defect, fetal MRI could provide an accurate diagnosis.

Altman Classification of SCT is as follows: Type 1: Predominantly external, minimal intrapelvic component; Type 2: Predominantly external, significant intrapelvic component; Type 3: Minor external component, predominantly internal component extending into the abdomen and Type 4: Only intrapelvic component.

The aim of this presentation is to report a rare case of Sacrococcygeal Teratoma associated with Maternal-Mirror Syndrome.

THE CASE

A thirty-one-year-old Pakistani female Gravida 5, para 1, Abortion 3 at 22 weeks of gestation was admitted to the hospital. The ultrasound had a provisional diagnosis of sacrococcygeal mass of the fetus. The possibility of termination of pregnancy was raised due to the progressive growth of the mass up to double the size of the fetus; the fundal height of 36 weeks was associated with polyhydramnios and placentomegaly. The patient reported having difficulty in passing urine for 5 days. Her blood pressure was within normal and no other warning symptoms of mirror syndrome.

She had one previous cesarean section for fetal distress 2 years prior and three previous abortions in her first trimester. All blood investigations were normal and she was Rh positive. She was given aspirin for her previous history of recurrent abortions.

US at 16 weeks of gestation revealed fetal sacrococcygeal growth. Repeated US after two weeks confirmed the diagnosis of SCT measuring 6 x 7 cm with suspected extrophy of the bladder, see figure 1.

Figure 1: Fetus Showing Sacrococcygeal Growth

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Examination at 20 weeks of gestation showed fundal height corresponding to 32 weeks. US Scan showed the SCT mass increased to 10.6 cm x 8 cm. The patient developed high blood pressure starting at 146/95 and increased to 158/99. She started to complain of severe headaches, and urine was tested positive for proteinuria.

Repeat US showed single alive fetus, hydrops, with SCT of 11 x 9 cm in size, enlarged placenta and polyhydramnios, see figure 2.

Magnesium sulfate (MgSO4) was given pre-op to stabilize her condition. Hysterotomy was performed, where a female fetus was extracted, weighed 1.330 kg. The SCT was boggy and soft but ruptured during extraction. The fetus expired immediately and showed features of hydrops with generalized edema of the face, abdomen, feet, and hands, see figures 3 and 4. Bladder extrophy was absent. The placenta weighed 2 kg, edematous and spongy, see figure 5.

Two hours post-surgery, the patient developed shortness of breath, low oxygen saturation (SPO2) approximately 83% in room air, raised pulse of 133/minute, and blood pressure 203/103 mmHg. Chest examination showed decrease air entry with bilateral crepitation and expiratory wheezes suspecting pulmonary edema. She was kept on 6 liters of oxygen, MgSO4 infusion was restarted and labetalol as per protocol. Stat dose of IV Lasix was administered. Two hours later, the patient’s condition improved; SPO2 was normal 98% on 6 liters oxygen face mask, BP decreased to 140/98, and albuminuria was negative. Echo showed normal findings with an ejection fraction of 61%.

CT angio showed no pulmonary embolism; however, there was bilateral bronchopneumonia and (mild) pleural effusion and cardiomegaly. The patient was transferred to the CCU, treated with Concord (beta blocker), Ramipril (ACE inhibitor), and Furosemide. Her blood pressure returned to normal 24 hours after surgery. Her general condition improved, and she was discharged on 5th-day post-surgery to continue with the same medications. Histopathology confirmed a benign sacrococcygeal teratoma, see figure 6.
DISCUSSION

Mirror syndrome, originally called Ballantyne syndrome, was first reported in 1892 showing maternal edema in a pregnancy complicated with fetal and placental hydrops due to rhesus isoimmunization. It is called early-onset mirror syndrome pre-eclampsia when the fetus has severe hydrops with placentomegaly of immune and non-immune causes. It can lead to fetal mortality in 56% of cases; while it can cause maternal morbidity in 21.4% such as pulmonary edema and oliguria.

The exact pathophysiology is still unknown. However, one of the theories was that the placentomegaly was the key cause for the development of pre-eclampsia. This large placenta could cause more debris than usual in the maternal circulation, followed by an inflammatory response due to the debris and oxidative stress would lead to endothelial dysfunction. Clinically, it presents with hypertension and its consequences. Unlike pre-eclampsia, mirror syndrome has higher fetal mortality rate.

Mirror syndrome can be associated with rhesus isoimmunization (29%), twin-twin transfusion syndrome (18%), and viral infection (16%). However, fetal placental tumors, such as sacrococcygeal teratoma and placental chorioangioma are more common, approximately 37.5%. Other non-structural includes fetal arrhythmias. In a systematic review of 56 reported cases of maternal mirror syndrome, the earliest gestational age of diagnosis was made around 16 weeks in a case of twin pregnancy.

The average gestational age at the time of diagnosis of SCT is between 22.5 to 27 weeks of gestation by ultrasound and can be confirmed with MRI. Besides the morbidity of the mother of mirror syndrome, diagnosing SCT during the antenatal period is important to save the fetus by early intervention. These tumors are highly vascularized and can transform into malignancy if not excised in early life.

Cesarean section or hysterotomy is the recommended safe delivery, especially if the mass is more than 5 cm.

CONCLUSION

The decision for termination of a pregnancy with non-immune hydrops fetalis due to SCT complicated with early-onset of mirror syndrome was made for maternal sake regardless of the fetal outcome, which carried poor prognosis due to severe hydrops fetalis and the rapid growth of the tumor associated with placentomegaly.

REFERENCES