Maternal Mirror Syndrome as a Consequence of Fetal Sacrococcygeal Teratoma: A Case Report and Literature Review

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Background

Sacrococcygeal teratomas (SCTs) are the most frequently recognized fetal neoplasm, with an estimated incidence of approximately 1 in 30,000 live births (1). Prenatal diagnosis typically occurs during the second trimester via routine ultrasound, which reveals a mass near the lower spine. Perinatal complications may include preterm labor, spontaneous tumor hemorrhage or rupture, hydrops fetalis, IUFD, or maternal mirror syndrome. Mirror syndrome (Ballantyne syndrome) is an exceedingly rare and life-threatening condition, with few cases diagnosed and documented in the academic literature (2). It is characterized by generalized maternal edema, often with pulmonary involvement, that "mirrors" the edema of the hydropic fetus and placenta. Its clinical course and presentation may be similar to severe preeclampsia. Delivery is usually required to induce remission of maternal symptoms.

Objective

- Present a case of this rare phenomenon and discuss the presentation of maternal Mirror Syndrome
- Stress the importance of close antepartum surveillance for these patients

Case Presentation

- A 24 year old female, gravida 4 para 2012 presented to the High Risk OB office at 20 weeks gestation for evaluation of a fetal spinal mass noted during an outside ultrasound.
- A SCT was diagnosed, measuring 6.7 x 4.7 x 7.8 cm. Bilateral pyelectasis and abdominal ascites were also noted (Figure 1).

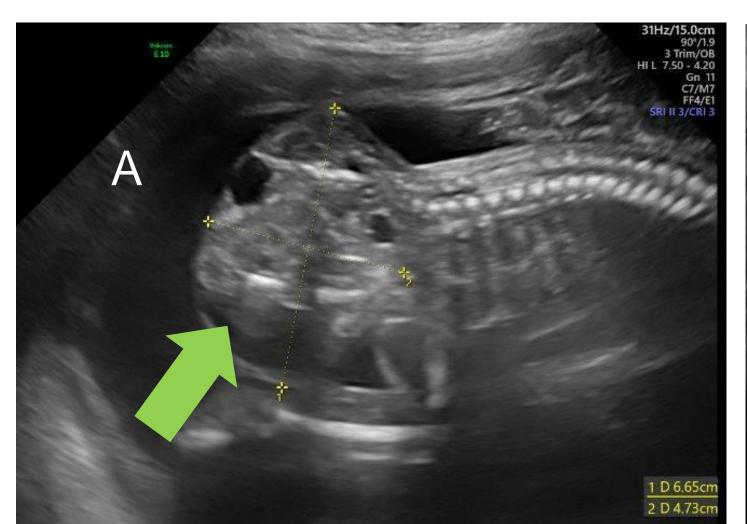




Figure 1. Ultrasound images: SCT (arrow) with measurements (A) and abdominal ascites (arrow) (B)

Case Presentation (continued)

- By 24 weeks EGA, the SCT measured 10.2 x 7.6 x 9.8 cm (Figure 2)
- There was evidence of worsening ascites, a pericardial effusion, and ventricular wall hypertrophy. Lower extremity equinovarus was also noted, consistent with compression or infiltration of nerve roots.

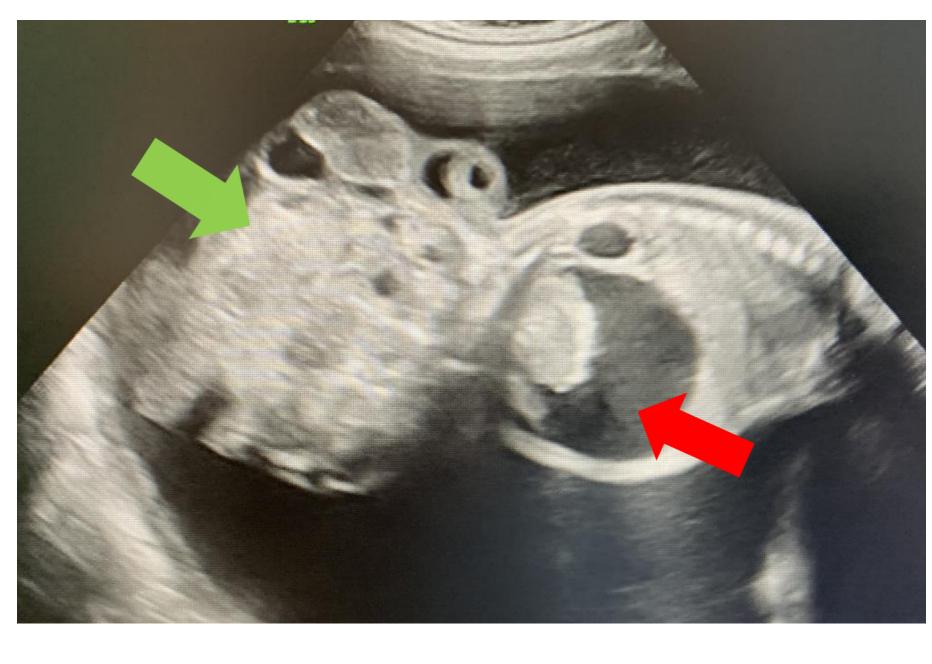


Figure 2. Ultrasound image: enlarging SCT (arrow), worsening abdominal ascites (arrow) and polyhydramnios

- A second opinion was obtained within the same week. Cardiomegaly and left and right ventricular dysfunction were noted in addition to a further enlarging SCT. Polyhydramnios and placentomegaly were also appreciated. The prognosis was deemed "lethal" with no in utero or postnatal surgery recommended.
- The patient shortly thereafter developed transaminitis and abdominal ascites consistent with maternal Mirror Syndrome.
- Delivery was recommended and occurred at 25 weeks gestational age by repeat low transverse cesarean section.
- The neonate received palliative care before expiring shortly after delivery (Figure 3).
- The mother recovered uneventfully with eventual resolution of her transaminitis.

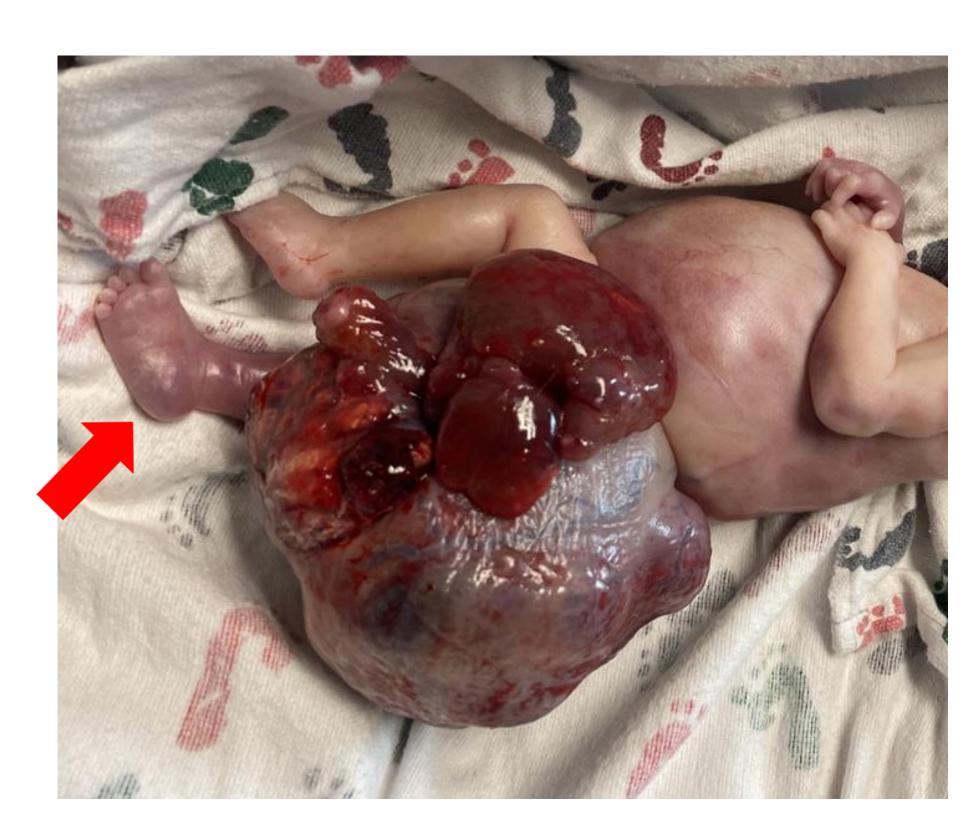


Figure 3. Neonatal image of SCT and right equinovarus (arrow)



Discussion

- Sacrococcygeal teratomas (SCT) are one of the most common neoplasms diagnosed prenatally
- They are germ cell tumors and are thought to arise from the totipotent cells along Hensen node
- There are 4 types, classified by external vs internal components
- The tumor histological type may be mature, immature, or malignant
- Sonographically, SCTs appear as a solid and/or cystic mass that arises from the anterior sacrum and usually extends inferiorly and externally as it grows
- They may enlarge rapidly with advancing gestation
- Fetuses may develop hydrops from high-output cardiac failure
- Mirror Syndrome, also known as Ballantyne Syndrome, is a rare complication of pregnancy and is associated with fetal hydrops
- Mirror Syndrome has many characteristics similar to preeclampsia including edema, hypertension, proteinuria, and elevated liver enzymes. Some even consider Mirror Syndrome to be a form of severe preeclampsia
- In most cases with Mirror Syndrome, prompt delivery is indicated which is typically followed by resolution of maternal symptoms

Conclusion

- SCTs are relatively common tumors found sonographically in a fetus that may enlarge rapidly and cause high-output cardiac failure leading to hydrops fetalis
- Close antepartum surveillance is required given the high morbidity for the mother and fetus and the potential for rapid progression of worsening disease

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