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Fetal Sacrococcygeal Teratoma: A Case Report of a Giant Tumor with an Excellent Outcome

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Abstract. Sacrococcygeal teratoma (SCT) occurs in approximately 1 per 20,000–40,000 births and is the most frequently encountered fetal teratoma, with 75% of cases observed in female fetuses. SCT can be detected on ultrasound as early as the first trimester, presenting as a large mass originating from the sacrococcygeal area, with or without an intrapelvic component. The prenatal course for most fetuses with SCT is generally uneventful, with only a few cases experiencing obstetric and fetal complications. We present the case of a 19-year-old woman who was in good health and had no relevant family or medical history. She was gravida 2 and para 1. During the first trimester scan, an examination revealed a heterogeneous mass in the presacral area with a predominantly multicystic appearance, measuring 12 mm in diameter. At 21+6 weeks of gestation, the Type 2 fetal SCT showed an increase in volume with the size of 49×37×36 mm and contiune to increase in size. The male fetus was delivered by elective Cesarean section at 38 weeks of gestation. The resection of the tumor and coccyx was performed when the newborn was 7 days old. The tumor measured 190×160×100 mm and weighed 1100 g. Pathological examination confirmed the diagnosis of a mature teratoma (Grade 0), and the resection margins were negative. Our case report highlights a fetus with a large and rapidly growing SCT, yet the outcome was excellent.

Keywords: sacrococcygeal teratomas, prenatal diagnostic, vascular index, alpha-fetoprotein.

Fetalinė sakrokokcigealinė teratoma. Atvejo pristatymas: didžiulis auglys, puiki baigtis

Santrauka. Sakrokokcigealinė teratoma (SCT) būna apytiksliai vienu iš 20 000–40 000 gimimų ir yra dažniausia fetalinė teratoma – 75 proc. atvejų būna moteriškos lyties embrionuose. SCT gali būti nustatyta ultragarsu jau net pirmąjį nėštumo trimestrą. SCT – tai didelis auglys, kurio kilmės vieta yra sakrokokcigealinis plotas, jame gali būti intrapelvinis komponentas ar jo nebūti. Daugelio gemalų su SCT raida iki gimimo įprastai yra neprobleminė ir tik nedaugeliu atvejų pasitaiko akušerinių ar vaisiaus komplikacijų.

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Pateikiame devyniolikametės moters atvejį. Jos sveikata buvo gera ir nebuvo jokių šeimos istorijos ar asmeninių medicininių sąsajų. Tai buvo moters antrasis nėštumas ir pirmasis gimdymas. Pirmojo semestro ultragarsinio tyrimo metu presakralinėje srityje buvo aptikta nevienalytė masė, aiškios multicistinės išvaizdos. Masės skersmuo buvo 12 mm. Po 21+6 savaičių nėštumo buvo nustatyta antrojo tipo gemalo 49×37×36 mm dydžio SCT, kuri ir toliau didėjo. Buvo pasirinkta daryti cezario pjūvį. Po 38 savaičių nėštumo gimė vyriškosios lyties kūdikis. Auglys ir uodegikaulis naujagimiui išpjauti 7 dienų amžiaus. Auglio dydis buvo 190×160×100 mm, jo svoris 1100 g. Patologiniu tyrimu nustatyta subrendusi teratoma (lygis 0). Rezekcijos vietoje patologijos nebuvo. Šiuo atveju pristatytos situacijos, kai vaisius buvo su didele ir sparčiai augančia SCT, baigtis itin sėkminga.

Raktažodžiai: sakrokokcigealinė teratoma, prenatalinė diagnostika, kraujagyslių indeksas, alfafetoproteinas

Introduction

Incidence and Etiology

Teratomas are the most common congenital tumors and often involve various locations such as the coccyx and sacrum (referred to as sacrococcygeal teratomas), as well as the neck (cervical teratomas), brain, mediastinum, heart, and abdomen along the midline [1]. Sacrococcygeal teratoma (SCT) occurs in approximately 1 per 20,000–40,000 births and is the most frequently encountered fetal teratoma, with 75% of cases observed in female fetuses [2], [3].

Histology and Types of Sacrococcygeal Teratoma

Histologically, SCT can be classified into three groups: mature (or benign), immature, or malignant [4]. Overall, about 90% of SCT cases are benign.

There are four types of SCT that can be distinguished. According to the Surgical Section of the American Academy of Pediatrics, the following four types of SCT are recognized (Figure 1):

- Type 1: Predominantly external, with a minimal presacral component (45.8%).
- Type 2: Predominantly external, with a significant intrapelvic component (34%).
- Type 3: Predominantly internal, with abdominal extension (8.6%).
- Type 4: Entirely internal, with no external component (9.6%) (5).

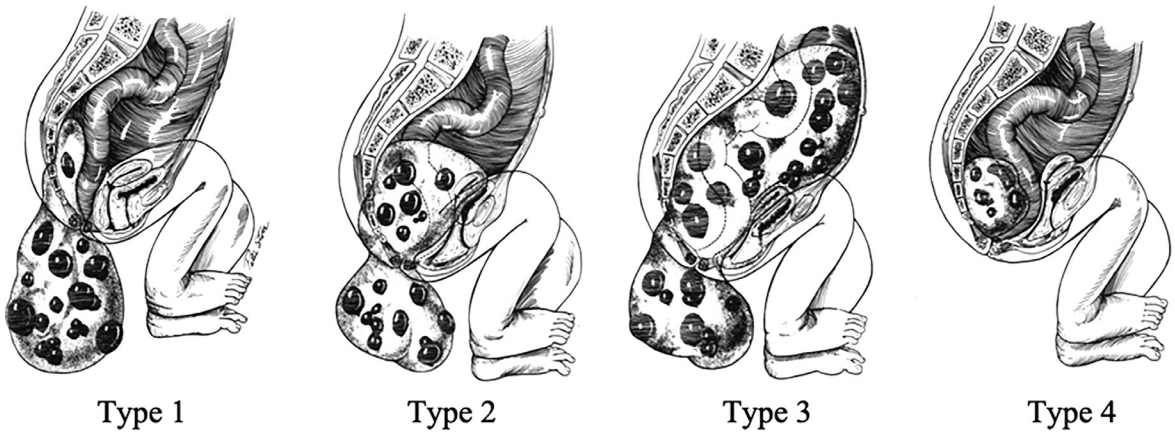

Figure 1. Types of sacrococcygeal teratomas.

Diagnostics

SCT can be detected on ultrasound as early as the first trimester, presenting as a large mass originating from the sacrococcygeal area, with or without an intrapelvic component [6], [7]. It is recommended to perform serial ultrasonography, including Doppler ultrasonography, to monitor the tumor's growth and vascularization over time. MRI (magnetic resonance imaging) provides a comprehensive assessment of SCT due to its characteristic appearance [8].

The ultrasound machine's virtual organ computer-aided analysis (VOCAL) software can utilize the vascular index (VI) to assess SCT. It calculates the VI by comparing the number of colored voxels representing blood vessels with the total voxel count of the SCT, expressed as a percentage (VI%) [9]. A VI% above 8% indicates an adverse outcome.

Prognosis

Approximately 50% of prenatally diagnosed SCT patients survive. Fetal SCT diagnosed in utero carry a high risk of preterm delivery (50%), a mortality rate of 15–35%, and a morbidity rate of 12–68%.

Vaginal delivery alone may result in traumatic hemorrhage, which can be life-threatening if not anticipated and promptly managed. Therefore, the mode of delivery for infants with SCT should be carefully considered [9].

In survivors, there is a risk of long-term complications, such as urinary and fecal incontinence, which can occur due to nerve injury during surgery. Additionally, there is a possibility of malignant transformation of the tumor if surgery is delayed or if incomplete excision occurs.

Recurrence has been reported even after resection of mature SCT, with recurrence rates ranging from 0% to 26%, averaging around 10% [1].

Management strategy

Various interventions have been described, including coiling, embolization, sclerotherapy, monopolar cautery, laser ablation, and radiofrequency ablation [10]. The overall survival rate for cases undergoing minimally invasive in-utero procedures is 44%, while for open fetal surgery, it is 50% [6], [10].

There is a greater consensus regarding the risk of malignancy after birth. Timely resection of the SCT is crucial, as the risk of malignancy increases with delayed surgery. The postnatal surgical approach involves complete removal of the tumor, including the coccyx, to minimize the chances of recurrence.

The aim of our case report is to present fetus with a large SCT where the diagosis was made during early pregancy and the outcome was excellent.

Case Report

We present the case of a 19-year-old woman who was in good health and had no relevant family or medical history. She was gravida 2 and para 1 and was referred to Riga Maternity Hospital for further evaluation. During the first trimester scan, an examination revealed a heterogeneous mass in the presacral area with a predominantly multicystic appearance, measuring 12 mm in diameter (Figure 2). The first trimester combined screening indicated a low risk for trisomies 21, 13, and 18.

During the evaluation, all cranial sonographic markers for open spina bifida were found to be negative. The ratio between brain stem thickness and its distance from the occipital bone (referred to as BS-to-occipital bone distance or BSOB) was within normal limits. The BS/BSOB ratio was less than 1, further supporting the diagnosis of SCT and ruling out open spina bifida in the current fetus.

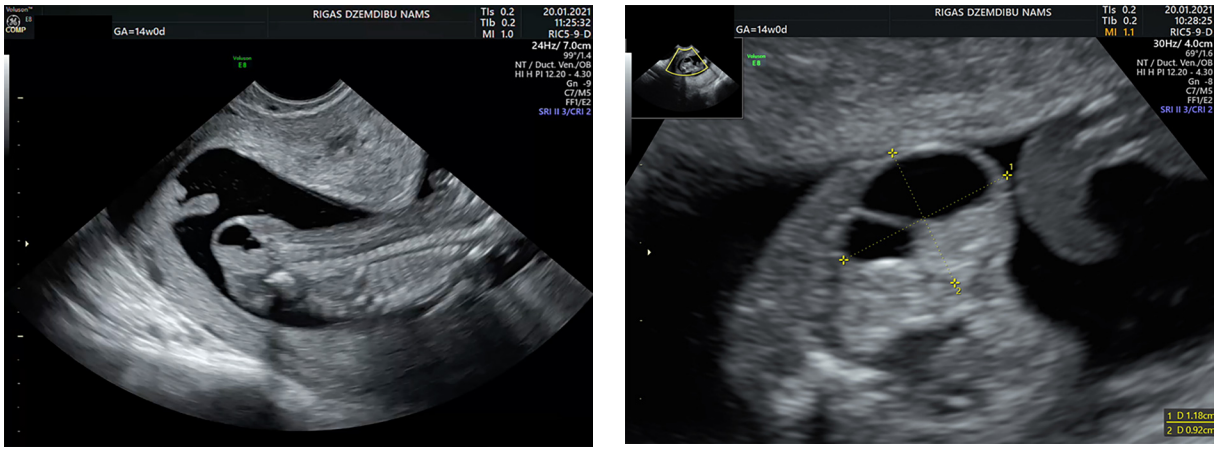

Figure 2. A heterogeneous mass with a predominantly multicystic appearance, measuring 12 mm in size, was detected in the presacral area.

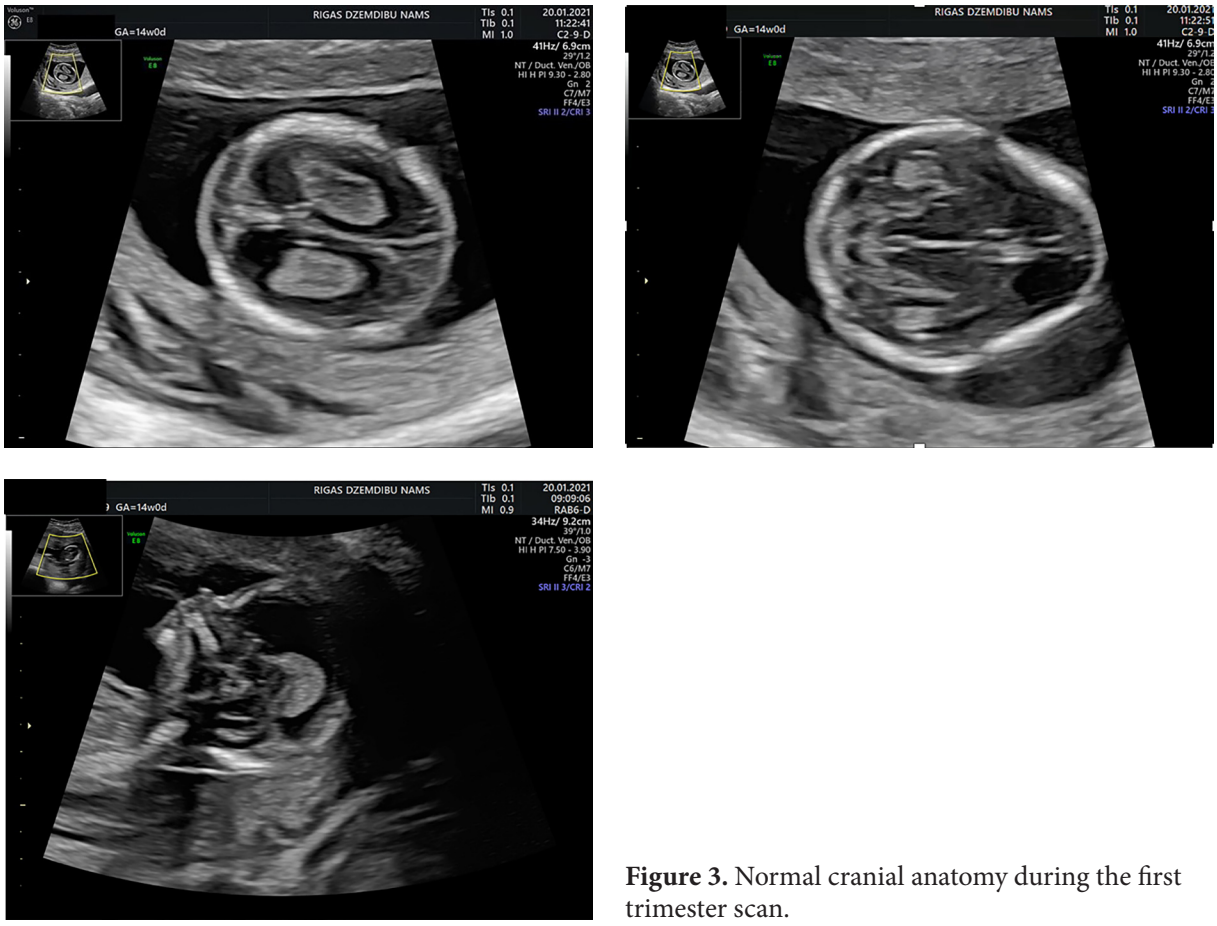

Figure 3. Normal cranial anatomy during the first trimester scan.

During the follow-up ultrasound examination at 21+6 weeks of gestation, the Type 2 fetal SCT showed an increase in volume. The size of the SCT was now measured at 49×37×36 mm, with a calculated volume of 65.2 cm^3. The fetal weight was 450 g, resulting in a tumor volume-to-fetal weight ratio of 0.60. The SCT displayed a complex appearance with both solid and cystic components. Minimal vascularization within the tumor was observed on Doppler ultrasound, with a VI of 15%. There were no indications of any additional fetal anomalies.

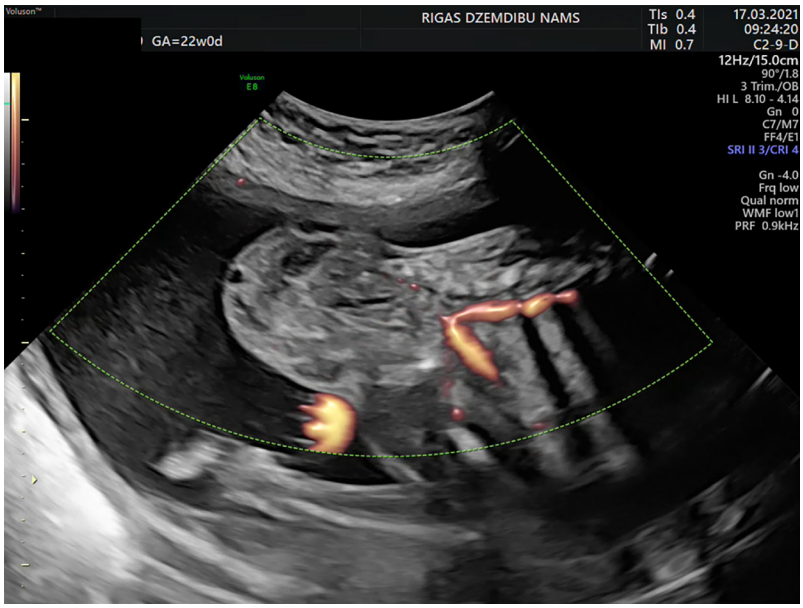

Figure 4. Sacrococcygeal teratoma with the size of 49×37×36 mm and volume 65.2 cm^3, minimal vascularization in the second trimester scan.

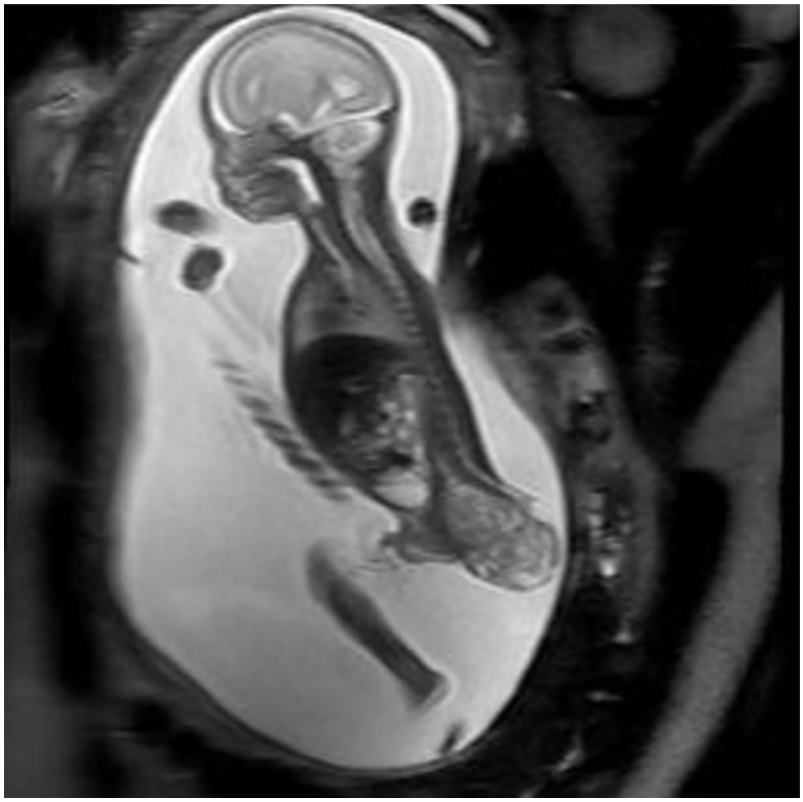

Figure 5. Magnetic resonance imaging revealed a heterogeneous cystic sacrococcygeal teratoma measuring 47×46×30 mm, located below the bladder.

At 22 weeks of gestation, an MRI scan was conducted, showing a heterogeneous cystic tumor with dimensions of 47×46×30 mm. The tumor was located below the bladder, with its smallest part near the coccyx and rectum. The majority of the tumor extended outside the body into the gluteal area. No further genetic testing was performed, and following thorough counseling, the family made the decision to proceed with the pregnancy.

Throughout the follow-up period, both the external and internal components of the SCT gradually increased in size. By 32 weeks of gestation, the cystic-solid mass had grown to 677 cm^3. The

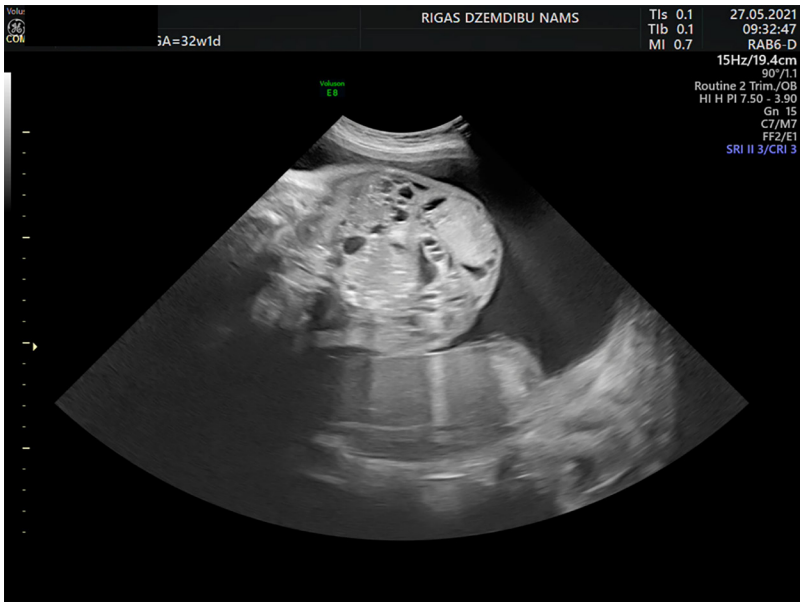

Figure 6. At 32 weeks of gestation, there was observed enlargement of both the external and internal components of the cystic-solid tumor.

Figure 7. At 32 weeks of gestation, the vascular index measurement of the sacrococcygeal teratoma was found to be 15%, indicating a moderate level of vascularity within the tumor.

measurement of the tumor at this stage was 15%, indicating a moderate level of vascularity within the SCT. The perianal complex was clearly visible during ultrasound examination. Mild polyhydramnios was noted, with an amniotic fluid index of 28 cm, but there were no signs of fetal anemia.

The male fetus was delivered by elective Cesarean section at 38 weeks of gestation.

The resection of the tumor and coccyx was performed when the newborn was 7 days old. The tumor measured 190x160x100 mm and weighed 1100 g. The surgical procedure successfully removed the tumor along with the coccyx. Pathological examination confirmed the diagnosis of a mature teratoma (Grade 0), and the resection margins were negative, indicating complete removal of the tumor without any residual disease.

On the 8th day of age, the alpha-fetoprotein (AFP) level was measured and found to be 38450 IU/mL.

However, it can be inferred that the surgical procedure was successful in removing the sacrococcygeal teratoma based on the subsequent developments.

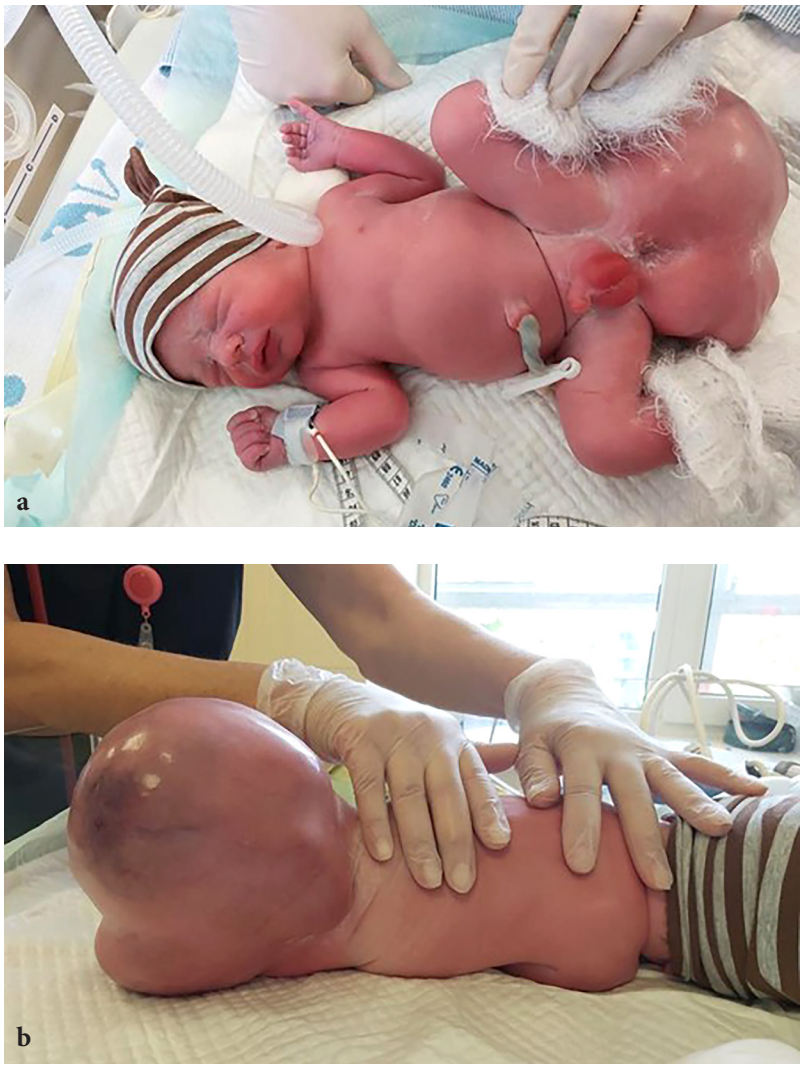

Figure 8a, 8b. (a) At the time of delivery, the newborn had a large tumor in the sacral area with an intact anus. (b) The presence of hemorrhage within the tumor was evident.

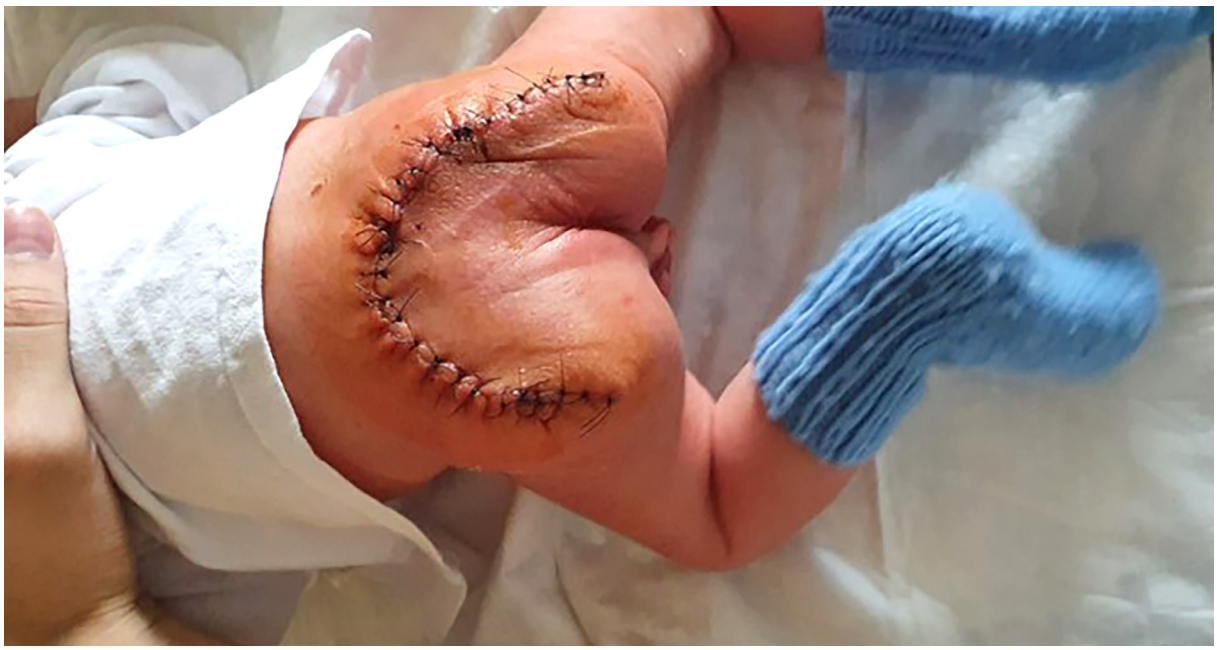

Figure 9. The gluteal area shortly after the surgery is not described in the provided information.

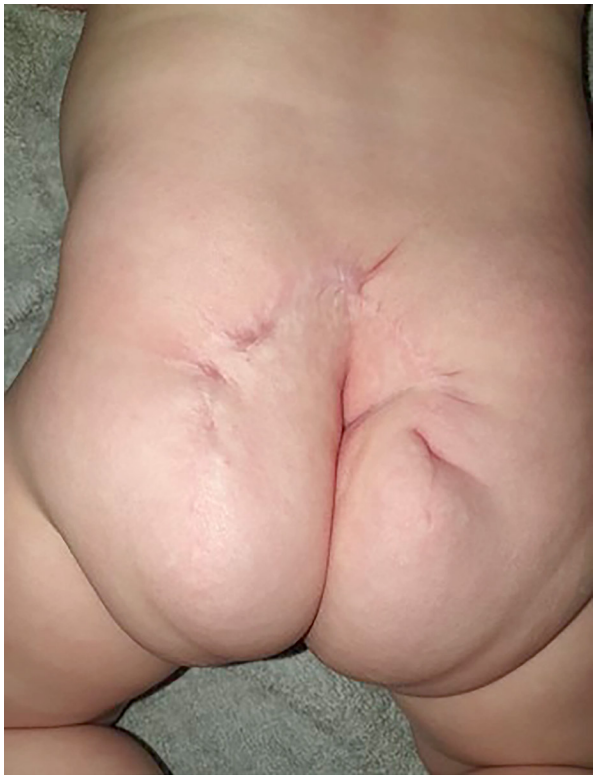

Figure 10. Gluteal area one year after the surgery.

At the age of 1 year, the AFP level was measured at 12.1 IU/mL, which is within the normal range. This is a positive predictor, indicating that there are no signs of tumor recurrence.

At the time of writing, the 2-year-old infant shows normal bladder and bowel functions, suggesting that there are no long-term complications related to the resection of the SCT. The scars resulting from the surgical procedure have a good cosmetic appearance, indicating satisfactory wound healing.

The absence of tumor recurrence, normal bladder and bowel functions, and good wound healing suggest a successful removal of the teratoma without any collateral damage to surrounding tissues and organs. Overall, the toddler's condition appears to be favorable with a positive outcome after the treatment of the SCT.

Discussion

Our case report highlights a fetus with a large and rapidly growing SCT, yet the diagosis was made during first trimester scan and outcome was excellent.

Detecting suspicions of fetal structural abnormalities in early pregnancy provides several benefits, including facilitating diagnostics, allowing for more time for parental counseling and decision-making, and establishing a follow-up and treatment plan.

The latest guidelines from the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG) and the World Association of Perinatal Medicine (WAPM) recommend an approach that focuses on comprehensive and systematic ultrasound examination of fetal anatomy during the first trimester [11], [12]. This approach aims to detect a significant proportion of structural anomalies, including central nervous system and neural tube abnormalities.

Meningomyelocele is the most common differential diagnosis for SCT. Differentiating between SCT and open spina bifida (OSB) is crucial for appropriate management and counseling. The latest guidelines from the ISUOG and the WAPM emphasize the early diagnosis of OSB, ideally in the first trimester [13], [14], [15], [16].

Following targeted fetal neurosonography with high-resolution transvaginal assessment, the diagnosis of OSB was excluded. Fetal MRI is a valuable imaging modality that provides detailed information about the content and extent of the tumor, aiding in counseling parents and planning postnatal treatment. The visualization of an intact perianal complex during mid-pregnancy ultrasound is an important prognostic indicator for both the medical team and the family. It suggests that the tumor is not affecting the surrounding structures and that there is a good likelihood of normal bladder and bowel functions after surgical removal.

Although the VI was 15% and the tumor showed rapid growth from 12 mm in diameter at week 14 of gestation to 190 mm after birth, it was encouraging that there were no fetal or maternal complications, allowing the pregnancy to reach full term.

Type 2 SCT is generally considered a benign tumor, but it is important to note the small probability (6%) of metastasis. In this case, the histological examination after surgery confirmed a mature teratoma (Grade 0), further supporting the benign nature of the tumor.

The successful surgical resection of the tumor along with the coccyx, negative resection margins, and absence of tumor recurrence are positive indicators of a favorable outcome. The absence of collateral damage to surrounding tissues and organs during the surgery suggests that long-term complications related to the resection of the SCT are unlikely.

Serum AFP level is a useful diagnostic tool in evaluating germ cell tumors, including SCT, before surgery [1], [3]. However, it is important to exercise caution when interpreting AFP levels, especially in neonates and infants, as their levels are naturally elevated due to hepatic production. Therefore, AFP levels alone cannot definitively confirm or exclude the presence of SCT. Following surgical resection of SCT, monitoring serum AFP levels can be helpful in detecting potential recurrences. It's important to note that the interpretation of AFP levels should be done in conjunction with other clinical findings and imaging studies to make an accurate assessment.

Overall, it appears that this case had a positive outcome with successful diagnosis, appropriate management, and satisfactory results after treatment of the SCT. The management of pregnancies with fetuses diagnosed with SCT requires a multidisciplinary approach and individualized care. As there are currently no standardized clinical guidelines specifically tailored for SCT management, each case must be carefully evaluated, taking into account various factors such as gestational age, coexisting pathologies, maternal condition, and potential complications.

The involvement of an experienced multidisciplinary team is crucial in providing comprehensive care to these patients. This team may include fetal medicine experts, geneticists, pediatric surgeons, neonatologists, and other specialists as needed. Their collective expertise allows for a evaluation of the risks and benefits of different management options, as well as the development of a tailored treatment plan.

Furthermore, the sharing of experiences and knowledge among medical professionals is vital for advancing the understanding and management of SCT. Collaboration and communication between experts in the field can contribute to the development of best practices, standardized protocols, and improved outcomes for patients and their families.

Given the complexity and variability of SCT cases, ongoing research and collaboration are necessary to further enhance our understanding of this condition and refine management strategies. Future studies and advancements in medical technology may provide additional insights and potential guidelines for the management of pregnancies with SCT.

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