

## Giant sacrococcygeal teratoma in a neonate: illustrative case

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**BACKGROUND** Sacrococcygeal teratomas (SCTs) are tumors that emerge in the sacrococcygeal area and contain tissue from all three germ layers. SCT affects about 1 in every 35,000–40,000 live births, with malignant transformation becoming more common as the patient gets older. Ultrasound helps in prenatal diagnosis. Surgical resection is the mainstay of treatment.

**OBSERVATIONS** A couple gave birth to a neonate with a small mass over his sacral region that progressively increased in size. Diagnostic magnetic resonance imaging was performed, and a diagnosis of giant SCT was established. Complete resection with flap reconstruction was performed. In regular follow-up, he is in a good state of health.

**LESSONS** One of the most common tumors in infancy, SCT should be carefully diagnosed. SCT is often confused with neural tube defects such as myelocystocele or myelomeningocele. Complete resection with appropriate reconstruction can ensure better treatment, and close follow-up until adulthood is recommended to keep a close view on its possible recurrence and to improve prognosis. Postoperative complications such as infection, bleeding, and urethral complications should be carefully watched.

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**KEYWORDS** sacrococcygeal teratoma; tumor; resection; prognosis

Sacrococcygeal teratomas (SCTs) are tumors that emerge in the sacrococcygeal area and contain tissue from all three germ layers. SCT affects about 1 in every 35,000–40,000 live births, with malignant transformation becoming more common as the patient's age increases.<sup>1,2</sup> Females are more prone to develop the tumor, with a male-to-female ratio of approximately 1:3–4.<sup>3,4</sup> SCT is assumed to have developed from "totipotent somatic cells that derived from the primitive knot (Hensen's node),"<sup>5</sup> and they are commonly attached to the coccyx.<sup>5,6</sup>

Obstetric ultrasound allows the clinician to make an antenatal diagnosis, typically in the second trimester via routine sonography. It is also an essential tool in the tumor assessment and surveillance during pregnancy in order to detect fetuses at elevated risk of problems and, when necessary, arrange a multidisciplinary therapy or intervention.<sup>7</sup>

In contrast to the neonate with SCT, the fetus with SCT is at risk of perinatal complications and mortality.<sup>8</sup> Compared with neonates diagnosed postnatally, fetuses with SCT detected prenatally have a threefold mortality rate.<sup>9</sup> Neonatal deaths can occur as a result of

obstetric complications such as tumor rupture, preterm labor, or dystocia in the mother.<sup>10</sup>

The fetus is in danger of developing high-output cardiac failure, placentomegaly, and hydrops, all of which can lead to fetal death due to the metabolic demands and vascular steal of a rapidly growing solid tumor. The mainstay of treatment is surgical resection, and recurrence is infrequent after complete excision.<sup>5</sup>

We report a case of giant SCT in a neonate. He was treated with complete resection and flap reconstruction and discharged after the seventh postoperative day. In regular follow-up, he is in a sound state of health.

### Illustrative Case

A 27-year-old woman gave birth to a male baby via spontaneous vaginal delivery. At the time of delivery, the weight of the baby was 3.5 kg, and he cried immediately after birth with Apgar scores of 9/10 at 1 minute and 10/10 at 5 minutes. The couple was not consanguineous

**ABBREVIATIONS** AFP = alpha-fetoprotein; beta-HCG = beta-human chorionic gonadotropin; SCT = sacrococcygeal teratoma.

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and had no reported history of medication, hereditary disease, substance abuse, or a family history of congenital anomalies and teratoma. The patient's serology was negative for human immunodeficiency virus, Venereal Disease Research Laboratory, and hepatitis B surface antigen, and she had no diabetes mellitus. An antenatal ultrasonic scan of the fetus was otherwise normal.

The baby had a swelling over his sacrococcygeal area that progressively increased in size and, on presentation, measured 14 × 3 cm as shown in Fig. 1. The swelling was hard in consistency, globular in shape with a smooth surface, and had a negative result of a transillumination test. The physical examination of the baby revealed a normal head. The cardiovascular system was not involved. On abdominal examination, the baby had a soft, depressible abdomen without any organomegaly. A diagnosis of SCT was established with the help of magnetic resonance imaging. When the baby was approximately 2 weeks old, teratoma resection, coccyx resection, and skin flap plasty were performed. Figure 2 shows the intraoperative image, and Fig. 3 shows the gross section of the resected teratoma. The excision of the coccyx and mass of 5 kg was complete. Histopathology showed a mature SCT with negative margins. The baby was discharged 7 days after surgery. In regular follow-up, he and his mother are in a good state of health.

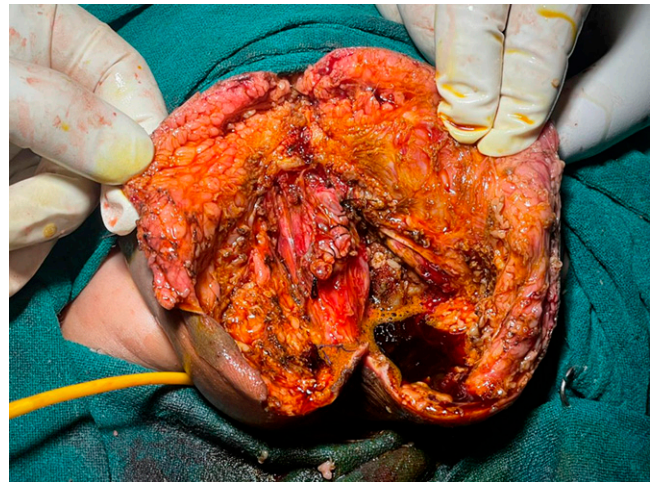
## Discussion

### Observations

A newborn with SCT has a favorable prognosis, depending on the timing of diagnosis, the tumor's malignant potential, and the simplicity of surgical excision.<sup>3</sup> The prenatal diagnosis was not feasible in our patient's case, because the tumor was small and not detectable by antenatal ultrasound.

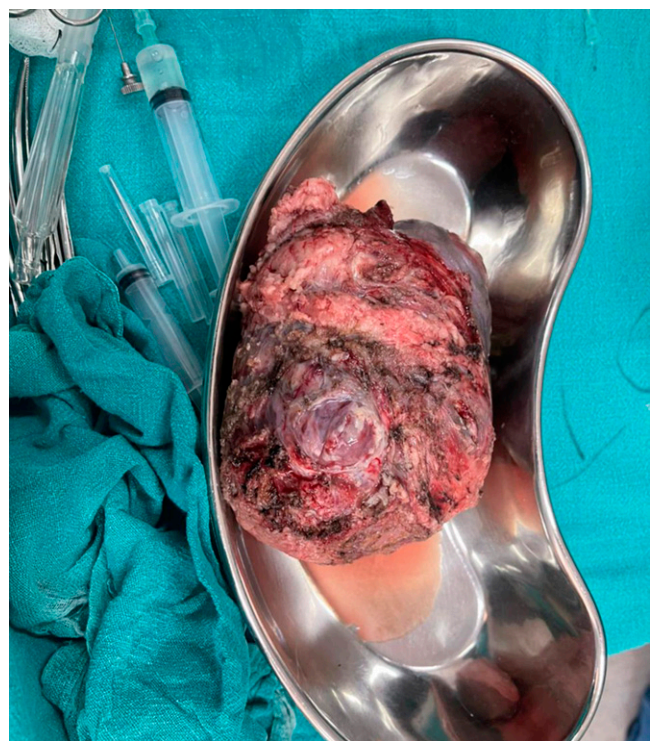


**FIG. 1.** A neonate with giant SCT.



**FIG. 2.** Intraoperative image.

SCTs are extragonadal neoplasms that present as “a mass in the midline caudal end of the fetus”<sup>9</sup> in the presacral area. They can be solid, cystic, or mixed in nature. In the majority of instances, they are asymptomatic or cause rectal or bladder blockage because of tumor development. Male infants are more commonly affected by malignant degeneration than female infants, despite female infants being more frequently reported.<sup>9,11</sup> In line with the aforementioned discussion, our case was a male neonate with a mass in the midline caudal end with its solid consistency.



**FIG. 3.** Gross section of SCT.

SCTs are divided into four types based on the external or internal components of the tumor according to the American Academy of Pediatrics Section on Surgery. Type I is primarily external, type II is external and has a significant intrapelvic component, type III is external and has a pelvic mass, and type IV is completely internal. Types I and II tumors are detected easily prenatally, whereas type IV tumors are more likely to be malignant.<sup>3,12</sup> Our case was completely external and thus met the type 1 classification.

Mature teratomas (fully differentiated tissues such as bone, teeth, and hair), immature teratomas (embryonal components or incompletely differentiated structures that confer a high risk of malignancy on the tumor), and malignant teratomas, which contain one or more of the malignant germ cell tumors (yolk sac tumor, choriocarcinoma, embryonal carcinoma, and others), are the three types of SCTs classified histologically.<sup>13–15</sup>

Antenatal diagnosis is often made by routine sonography in the second trimester, though it can also be detected in the first trimester. A growth in the vicinity of the distal spine is the most common finding, but there could also be an erosion in the spine or a collection of calcifications. As a result of the existence of the tumor, other structural abnormalities, such as hydronephrosis, rectal stenosis, or cardiomegaly, may be discovered.<sup>16</sup>

Magnetic resonance imaging can determine the tumor's extent and the pressure on adjacent organs, and it can distinguish this disease from the most prevalent differential diagnosis, a distal neural tube abnormality (myelocystocele or myelomeningocele). The major distinction is the site of the mass effect, which in SCTs is presacral and in neural tube defects is posterior.<sup>17</sup> Perinatal morbidity and death in SCT-affected fetuses remain high because of the frequent involvement of prematurity. Premature labor, malignant infiltration, tumor hemorrhage or rupture, amniotic fluid blockage, and heart failure are the most serious perinatal complications.<sup>18</sup>

SCTs are generally treated with the tumor and coccyx operative resection to prevent a recurrence. In most cases, this surgery is done after the baby is born, and it is only done in utero as a last resort in specialized facilities and in a small number of cases with a tumor that is at high predisposition of developing hydrops as well as gestational age less than 32 weeks.<sup>19</sup> For SCTs with no malignant components, total surgical resection is usually adequate, followed by 3 years of monitoring alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (beta-HCG) levels.<sup>20</sup> Adjuvant chemotherapy involving a platinum-based regimen is recommended in malignant cases. The recurrence rate of SCTs is approximately 4%.<sup>21</sup> Complete resection with flap reconstruction was the mode of treatment adopted in our case.

The incidence of malignant transformation within 2 months after birth is 20%, and it is 40% within 4 months. As a result, SCT should be removed entirely as soon as possible after delivery to prevent malignant change. The most common risk factors for recurrence include residual coccyx and tumor rupture during surgery.<sup>22</sup> As a result, during the surgery, the tumor and coccyx should be removed as thoroughly as feasible. Most doctors now believe that eliminating the coccyx and minimizing residual damage to the tumor cyst wall during surgery are critical to preventing a recurrence of benign teratoma. Even after full excision of an SCT, it might recur several years later. Patients should now be continuously monitored until they reach maturity.<sup>22</sup> According to other researchers, tumor size, percentage of solid components, rate of growth, vascular density, grade of cardiac function impairment, fetal edema, polyhydramnios, and maternal comorbidities are all associated with a poor fetal outcome.<sup>23,24</sup> In

addition to the aforesaid factors, the period of gestation at the time of delivery is also an independent prognostic variable.<sup>25</sup> Complete resection and regular follow-up are ensured in our case.

The prognosis of a fetus or newborn diagnosed with an SCT is based on three factors: the simplicity with which the teratoma may be surgically resected, the time of the diagnosis, and the kinds of tissue contained in the tumor.<sup>8</sup> Once the fetal lung matures, the route of delivery is determined by the size and vascularity of the tumor. To reduce the risk of hemorrhage, infants with massive or extremely vascular tumors should be delivered through cesarean section.<sup>26,27</sup> Because of the small size, spontaneous vaginal delivery was preferred in our case. Plain radiographs should be taken before surgery to determine the tumor volume and extent and to identify any dissemination. Computed tomography or magnetic resonance imaging may be used to detect pelvic involvement. Angiography can also be used to map the tumor's blood supply.<sup>21</sup>

While the patient waits for surgery, care must be taken to preserve the tumor from trauma or abrasion, which might result in substantial hemorrhage and the emergence of disseminated intravascular coagulation.<sup>28</sup> Wound infection is one of the most frequent postoperative complications.<sup>4</sup> Additional focus should be placed on severe urethral complications involved with teratoma excision.

## Lessons

SCT, one of the most frequent tumors in childhood, should be diagnosed carefully. Complete resection with proper reconstruction can provide better therapy, and regular follow-up until maturity is indicated to keep a close eye on its likely recurrence and to improve prognosis. The removal of an SCT requires careful consideration of potential scarring and lingering effects on nerve and muscle function. The manner in which such a fetus is delivered should be considered carefully. If the teratoma is large enough, a cesarean delivery can reduce the chance of hemorrhage. A careful watch is advised to avoid postoperative problems such as infection, hemorrhage, and serious urethral complications. Reviewing the literature and investigating this isolated case that arrives at the second level of care is most likely the most effective tool for keeping health professionals up to date and assisting them in providing better treatment to their patients. However, limitations such as the inability to suspect and diagnose it during pregnancy are unavoidable. Similarly, AFP and beta-HCG are not routinely tested due to the patient's financial constraints.

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#### Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

#### Author Contributions

Conception and design: all authors. Acquisition of data: Neupane, Lageju, Jaiswal, Bista, Sapkota. Analysis and interpretation of data: Neupane, Lageju, Jaiswal, Sapkota. Drafting the article: Neupane, Lageju, Jaiswal, Bista, Sapkota. Critically revising the article: Neupane, Lageju, Jaiswal, Bista, Sapkota. Reviewed submitted version of manuscript: Neupane, Lageju, Jaiswal, Sapkota. Approved the final version of the manuscript on behalf of all authors: Neupane. Administrative/technical/material support: Dahal. Study supervision: Dahal.

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