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Prenatal Diagnosis and Management of Sacrococcygeal Teratomas

Anca Budusan, Horatiu Gocan and Roxana Popa-Stanila

Abstract

Sacrococcygeal teratomas (SCT) represents a group tumors deriving from the primordial germ cells. It is the most common tumor affecting neonates, with a female to male ratio of almost 4:1.78. SCT are either benign (mature) or malignant (immature) with different outcome. With advancements in ultrasonography, more SCT are diagnosed prenatally. magnetic resonance imaging (MRI) is more accurate in describing the intrapelvic and abdominal extent of the tumor. Most fetal teratomas could be managed by planned delivery and postnatal surgery. The earlier the diagnosis and surgical intervention, the better the prognosis. A complete surgical excision of the tumor is necessary, including coccygectomy, to prevent recurrence. At the time of birth, most lesions are benign and surgical resection can be accomplished with relatively low morbidity and mortality. Recurrence is reported as 2–35% in patients with immature teratomas, tumor spillage, incomplete resection or failure to remove the coccyx. A long-term follow-up is required for any urinary or bowel dysfunction.

Keywords: sacrococcygeal, teratomas, surgery, children, tumor

1. Introduction

Sacrococcygeal teratoma is the most common congenital neoplasm. The word "teratoma" is derived from the Greek word "teratos" meaning monster. The first reported case was described on a cuneiform tablet dated approximately 2000 BC. Advances in antenatal imaging have let to prenatal detection of most SCTs and may avoid early mortality. Delayed presentation and presence of malignant elements continue to be poor prognostic factors, while surgical goal remains complete resection.

2. Background

Although rare, SCT is the most common tumor of the fetus and the newborn, with a reported prevalence of 0.25–0.28:10,000 live births. With more advancements in US (ultrasonography), more SCT are diagnosed prenatally.

2.1 Etiology

SCT arises from pluripotent cells of Hense's node that is present/located anterior to coccyx, a remnant of the primitive streak in the coccygeal region. The primitive

streak is a longitudinal ridge of ectodermal cells at the caudal end of the bilaminar embryonic disc. It consists of totipotent cells, which are able to transform into any type of cells. This structure determines the future craniocaudally axis of the embryo and demarcates the embryo into left and right halves. If totipotent cells of the primitive streak remain after the fourth week, these cells give rise to a SCT [1].

SCT is a relatively uncommon tumor affecting neonates, infants and children.

SCT represents a group of benign and malignant tumors deriving from the primordial germ cells. Pediatric germ cell tumors (GCTs) are neoplasms derived from primordial germ cells and may occur both inside the gonads and extragonadal organs. The five main histologic categories of GCTs are: dysgerminomas (in the ovary), seminomas (in the testes), teratomas, choriocarcinomas and endodermal sinus tumor (ESTS) or Yolk sac tumor. The most common site of extragonadal GCTs in the pediatric population is the sacrococcygeal region and the most common type are teratomas.

The sacrococcygeal region is the most frequent location for teratomas, but teratomas may occur in almost any organ, tending to develop more commonly in midline or paraxial location and can be observed from the brain (cephalad) to the coccyx (caudal). Less common sites are the mediastinum, testes, ovary, retroperitoneum, head [2, 3].

Females are affected more frequently with a female to male ratio of almost 4:1.78. SCT are either mature, immature or malignant, composed of embryonic elements. A mature SCT is a benign tumor containing only mature components, while immature SCT contains immature tissues. SCT that contains malignant elements are considered to be malignant tumors. Mature and immature teratomas are considered as benign tumors and may undergo malignant transformation. At birth, the great majority of SCTs are benign. They can manifest malignant transformation with advanced age.

They appear as cystic tumors or solid. The cystic may be filled with serous fluid, mucoid or sebaceous material, or even cerebrospinal fluid. Virtually any tissue can be present in a SCT. Neuroglial tissue, skin, respiratory and enteric epithelium cartilage, smooth muscle and striated muscle fibers are the most common elements found. Also bone, pancreatic tissue, choroid plexus and adrenal tissues are less commonly identified.

2.2 Classification

Size of a SCT (average 8 cm diameter, range 1–30 cm) does not predict its biological behavior. Altman et al. defined the size of SCT as follows:

Small: as 2–5 cm diameter

Moderate: 5–10 cm

Large: >10 cm.

Antenatal diagnosis is important to avoid complications during delivery. Fetal US and MRI are the mainstay of antenatal diagnosis of SCT. MRI is more accurate in describing the intrapelvic and abdominal extent of the tumor and provides more information on compression of adjacent organs. Prenatal assessment of the fetus is critical for counseling the parents and planning surgical options. Because of acoustic shadowing by the fetal pelvic bones, US cannot always define the extent of SCT. Fetal MRI has been successfully performed to evaluate anatomy, content and extent of the tumor, but just a few small cases series have been published yet.

SCT arise from the base of the coccyx and may continuously grow in the posterior direction forming an external protrusion, or in the anterior direction, dissecting and distorting surrounding structures such as the rectum, vagina and bladder, but without invading them. Based on this morphologic characteristic, Altman et al.

have been defined The American Academy of Pediatrics Surgical Section (APPSS) classification [3–5]:

Type I: predominantly external with minimal presacral component—45.8%

Type II: present externally, but with significant intrapelvic extension—34%

Type III: apparent externally but predominantly a pelvic mass extending into the abdomen—8.6%

Type IV: presacral mass with no external presentation—9.6%.

2.3 Histology

SCT are graded histologically as follows:

Grade 0—tumor contains only mature tissue

Grade 1—tumor contains rare foci of immature tissues

Grade 2—tumor contains moderate quantities of immature tissues

Grade 3—tumor contains large quantities of immature tissue with or without malignant yolk sac elements.

3. Diagnosis

3.1 Intrauterine diagnosis

The majority of SCT present between the 22nd and the 34th week of gestation. The diagnosis of SCT on routine US is associated with a greater than expected incidence of prenatal and perinatal complications. Close antenatal follow up is needed to optimize patient counselling and treatment in the presence of a completely solid tumor and the onset of polyhydramnios. A poor outcome is usually correlated with placentomegaly, cardiomegaly or non-immune hydrops fetalis.

3.2 Associated anomalies

Associated congenital malformations are observed in 12–15% of cases and occur more frequently with presacral tumors. The incidence of various congenital malformations associated with SCT ranges from 5 to 26%. Of these, anorectal and genital malformations are most commonly. A growing SCT during the first weeks of embryonic life will encroach between the layers of the cloacal membrane and prevent descent and fusion of the urorectal septum to the cloacal membrane, resulting in a high anorectal malformation with a rectourethral or rectovestibular fistula. The presence of SCT in the same period of time (47th weeks), when cloaca is subdivided by the urorectal septum to form the anorectal canal and the primitive urogenital sinus, could prevent fusion of the genital folds, resulting in a bifid scrotum or hypospadias. The most commonly observed anorectal defects are: imperforate anus, anorectal stenosis and common vertebral anomalies are: sacral hemivertebrae, absence of the sacrum and coccyx [6].

Other associated anomalies include spinal dysraphism, sacral agenesis, dislocation of the hips and meningocele. Rarely, gastrointestinal or cardiac defects are associated with SCT.

Currarino triad represents association of anorectal malformation, sacral dysplasia and presacral mass. Delay in diagnosis of the presacral lesion is common because a rectal examination may not be possible in many cases with anorectal stenosis. Presenting symptoms in some of these unusual cases include perirectal abscess or fistula in ano (**Figure 1**).
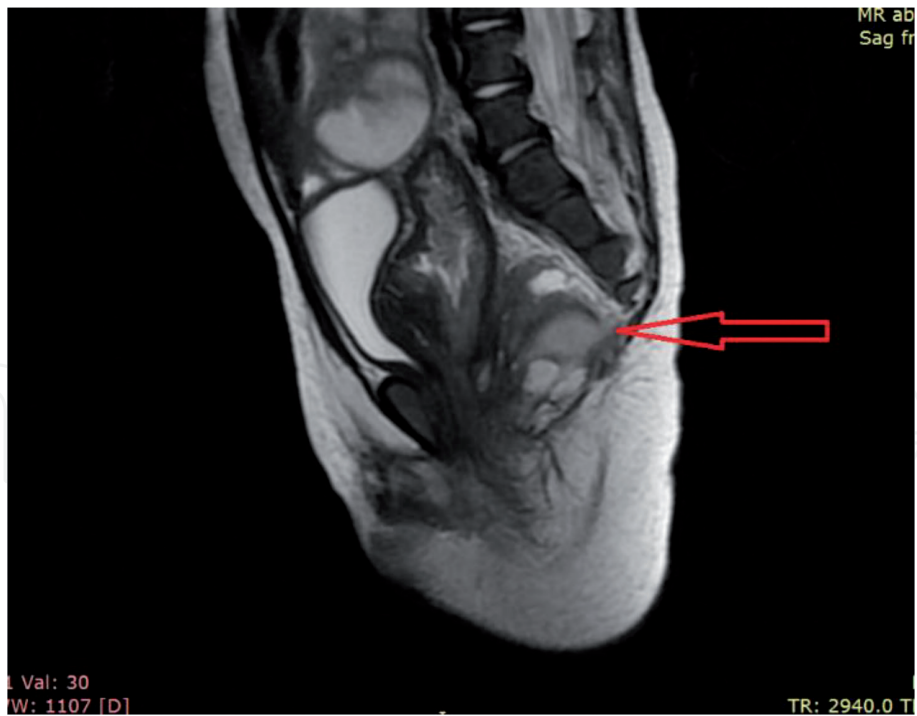
Figure 1.
Currarino triad in a female patient with SCT—MRI. Red arrow points the SCT. The girl presented with imperforate anus, recto-vestibular fistula and SCT was misdiagnosed until MRI.

3.3 Clinical presentation

Most external tumors are asymptomatic, with the exception of the presence of a visible exophytic large mass at the sacral region with occasional surface ulceration and hemorrhage and with anus displaced anteriorly (**Figure 2**). Sometimes, rupture of the tumor may occur as a result of a difficult delivery. Pelvic tumors or tumors that extend into the abdominal cavity may present with compression of the rectum or recto-sigmoid and urinary tract obstruction (constipation, frequent stools, obstruction of the bladder neck).

3.4 Diagnostic tools

Except for clinical examination, there are a variety of radiographic studies that can help. Plain X-Ray may show the presence of calcification within the tumor and anterior displacement of the rectum by the tumor. The sacrum may appear abnormal (such as hemivertebrae, agenesis).

Computer tomography (CT) or MRI of the pelvis with intravenous contrast material may reveal urinary tract displacement or obstruction and outlines the extent of the tumor more accurately. MRI is also a useful diagnosis of spinal cord extension of tumor (**Figure 3**).

A chest X-Ray or CT thoracic scan is obtaining to rule out the presence of pulmonary metastases.

Malignant SCT may have elevated tumor markers. The most commonly produced tumor marker is AFP (alpha-fetoprotein) because yolk sac components are the most common malignant elements. Other malignant elements may produce beta HCG (human chorionic gonadotropin). Serum AFP and beta HCG should be evaluated at the initial diagnostic work-up and assessed to monitor tumor relapse during the follow up period. The use of AFP as a tumor marker is well established and persistent, elevated level may indicate a residual tumor, recurrence or malignant degeneration. Because AFP is produced by fetal liver and fetal gastrointestinal tract,

Figure 2.
SCT type II in a newborn girl. Large mass visible in sacral region, with displacement of anal orifice.
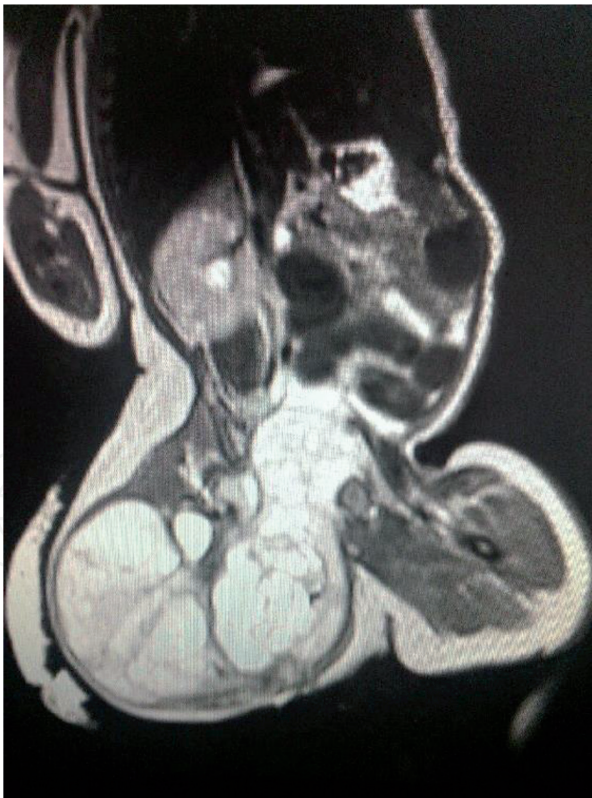

Figure 3.
Type II SCT with intrapelvic component—MRI.

its level is normally elevated in the first 8 months of life and after that age it rapidly falls to normal adult level (10 ng/ml). The mean time required for AFP to normalize after SCT resection is about 9 months [7, 8].

The differential diagnosis of SCT include rectal duplication, meningocele, lipoma, chordoma, epidermoid cyst, neuroblastoma.

4. Management

4.1 Prenatal management

Fetal MRI is a powerful addition to the prenatal evaluation of fetuses with SCT. Due to the fact that, in most cases, neonatal surgery is required soon after cesarean section, the anatomic details of tumor extent and involvement of adjacent structures may affect the surgical approach. Patients with significant intrapelvic extension of the tumor may need a combined abdominoperineal approach to control the blood supply and achieve complete resection. All these may contribute to avoid resection-related complications during surgery [9] (**Figure 4**).

Monitoring for fetal distress during pregnancy is very important. Some large tumors have a very high blood flow that causes a shift in blood flow away, producing fetal hydrops. Other possible complications are bleeding inside the tumor, polyhydramnios and preterm labor. A rare condition is called "mirror syndrome" where the mother mirrors the baby's sickness, leading to fluid retention, preeclampsia, high blood pressure, heart failure [10, 11].

4.2 Surgical management

Most fetal teratomas could be managed by planned delivery and postnatal surgery. The earlier the diagnosis and surgical intervention, the better the prognosis. A complete surgical excision of the tumor including coccygectomy is necessary, in order to avoid recurrence. A long term observation and follow-up is required for any urinary or bowel dysfunction.

Mature teratomas should not recur, if complete surgical excision and coccygectomy were achieved properly. Recurrence is reported in literature, as 2–35%, in patients with immature teratomas operated after the age of 5 months and is related to tumor spillage or incomplete excision.

At the time of birth, most lesions are benign and surgical resection can be accomplished with relatively low morbidity and mortality. The incidence of
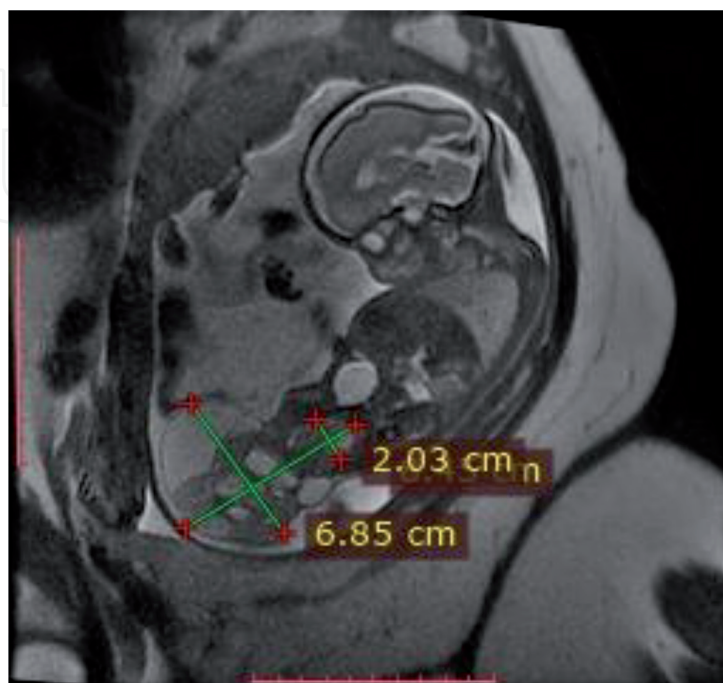

Figure 4.
Antenatal diagnosis of SCT using MRI and measurements.

malignancy in SCT is increasing with age. Failure to remove the coccyx results in 30–40% recurrence rate, with a higher probability of malignancy. Alpha fetoprotein may be used to detect early occurrence of malignancy.

Management of SCT depends on fetal lung maturation and presence of placental enlargement and/or fetal hydrops. When maturity of fetal lung without placental enlargement and/or hydrops fetalis, planned cesarean section is indicated. Some authors recommend preventive early delivery by cesarean section when the tumor exceeds the diameter of 5 cm, to avoid complications such as rupture and hemorrhage. The primary treatment of SCT is early surgical resection with complete excision of the coccyx. Early surgical intervention is associated with better prognosis. The surgical approach depends on the degree of pelvic extension. Posterior sacral approach is recommended in type I and II, and combined abdominal and posterior sacral approaches in type III and IV. The technique of wide resection of benign lesions with coccygectomy is helpful in preventing recurrence and has changed little over the last four decades.

The goals of surgical resection of an SCT are:

- Complete resection of the tumor

- Removal of the coccyx

- Reconstruction of the pelvic floor and ano-rectal sphincter

- Acceptable cosmetic appearance

After inserting a urinary catheter, the patient is placed in the prone jack-knife position. A V-shaped incision is made at the superior margin of the tumor. It is important to identify the course of the anus by placing a tube in the anal canal. After raising skin flaps, the muscles are dissected from the tumor which must be resected with the coccyx, after ligation of its main blood supply, which are middle sacral artery or branches from the hypogastric arteries [4, 7, 12].

Figure 5.
Large SCT in a newborn—preoperative. SCT represents more than 50% of birthweight, with visible ulceration of the skin.

Figure 6.
Same newborn—postoperative view, after removal of SCT.

The multiorgan involvement makes the anesthetic management challenging. Prematurity and hypothermia are risk factors for coagulopathy and can lead to fatal consequences. Management of intraoperative bleeding and early extubation are good outcome predictors [5, 13, 14] (**Figure 5**).

Patients with malignant SCT are managed after surgery with irradiation if residual disease is present, and chemotherapy. Most tumors have a plane of dissection and can be removed easily. It is safer and recommended to catheterize the bladder to keep it away from the tumor and place a large rubber catheter in rectum for identification. Levator ani muscles are often stretched over tumor and should be reconstructed after tumor is excised. Drainage is necessary as there is a large raw area and collections should be avoided (**Figure 6**).

Preservation of the autonomous nerve supply to the bladder and rectum may be difficult. Therefore, postoperative complications (31%) that may be expected are bladder dysfunction, incontinence for feces and dysesthesia. The main postoperative early complication is wound infection because of the proximity to the anus and the skin flaps that may be needed.

5. Conclusions

SCT is a common neonatal neoplasm. Antenatal diagnosis is essential, for avoiding complications and high morbidity, mortality. Delivery in a tertiary center by cesarean section, when needed, should be emphasized. Early diagnosis and complete resection of the tumor with removal of the coccyx is associated with good prognosis.

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