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## RESEARCH ARTICLE

### SACRO-COCCYGEAL TERATOMA: WHAT DO WE EXPECT FROM IMAGING?

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#### Abstract

Sacro-coccygeal teratomas (SCT) are rare congenital tumors, although they are the most common in the neonatal period. The revealing clinical situation is variable. It may be a hydramnios before birth, a sacral or gluteal mass at birth, or signs of urinary or digestive compression later. Ante-natal diagnosis of SCT requires regular ultrasound monitoring to assess the evolution of the tumor and its possible impact on the fetus, and to decide the modality and timing of delivery. Before a sacro-coccygeal mass, CT and MRI can evoke the diagnosis, by visualizing a very heterogeneous tumor. They make it possible, among other things, to appreciate its extension and its reports, and to establish a classification. Tumor excision is the only effective treatment for TSC. It is indicated as soon as the diagnosis is made, even in the first days of life. The purpose of this article is to illustrate the importance of imaging techniques in the diagnosis, characterization, evaluation of lesion extension, management orientation and postoperative monitoring.

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#### Introduction: -

Sacro-coccygeal teratoma is a germinal tumor containing tissues from two or three embryonic layers (ectoderm, mesoderm and endoderm). Although rare, it remains the most common tumor of the caudal region of the child (7). The etiology of teratomas is not yet clear, but may result from the migration of totipotent cells near the Hensen node in the embryo.

The sacrococcygeal teratoma is considered a surgical emergency because of its potential for malignant transformation from the 4th month of life [4], the prognosis is generally excellent in the neonatal period but gradually becomes darker with advanced age of the child.

In addition to the histological type of the tumor, which is benign in 90% of cases, the other prognostic factors of SCT are: its size as well as its extent, the degree of prematurity and finally the complete resection of the tumor [5,6]. Until recent years, the discovery of SCT was fortuitous at the time of delivery, and they could ask sometimes, depending on their volume, serious obstetric problems.

The antenatal risk of sacro-coccygeal teratoma is that of haemodynamic decompensation that can lead to fetal death in utero or neonatal death (9; 10).

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Altman's classification distinguishes four types according to the location and the pelvic extension of the tumor (1,8). Nevertheless, the advent of ultrasound has allowed the antenatal diagnosis of SCT [7], and thus a better obstetric and neonatal management of patients.

For example, ultrasonography and fetal MRI allow prenatal discovery, whereas postnatal imaging determines the importance of an endopelvic component, its impact on neighboring organs and looks for signs of degeneration (1, 4, 6, 7).

### **Epidemiology**

Despite the rarity of SCT, it ranks first among teratomas of the newborn [44]; the 2nd rank of neonatal tumors after lymphangiomas and the 3rd rank of retroperitoneal tumors in children, after neuroblastoma and nephroblastoma [30], with an incidence of about 1: 40,000 and 1: 35,000 live births [2] ]. There is a clear female predominance with a sex ratio of 1/3 to 4 (1,3,7).

### **Pathology**

The sacro-coccygeal teratoma develops from elements of the caudal cell mass that have a potential for multiple differentiation which explains the heterogeneity of the tissues contained in the mass (bone, hair, teeth ...). It is about a tumor reaching preferentially the fetuses of female sex. Malignant degeneration is observed during childhood, so it is essential to surgically remove the tumor before the age of 6 months and completely.

Anatomical classification of Altman

In 1974, ALTMAN et al [11]. Proposed a topographic classification of sacrococcygeal teratomas in their report to the American Academy of Pediatric Surgery:

1. Type I: Type I teratomas are almost exclusively external with a minimal pelvic component.
2. Type II: Type II teratomas have a significant pelvic component.
3. Type III: the intra-abdominal and intra-pelvic component is much larger than the external component.
4. Type IV: They are exclusively pre-sacral without external component; this is the most difficult form to diagnose clinically as well as ante- or post-natal ultrasound.

### **What are the support of imaging techniques?**

#### **Antenatal diagnosis**

Formerly the discovery of SCT was done during delivery, which usually poses serious obstetric problems requiring surgical management which explains the dark perinatal prognosis of these tumors. Advances in ultrasound and fetal medicine currently make it possible to recognize this pathology during pregnancy, but also to specify certain prognostic factors in order to give better information to the couple. The prenatal knowledge of these SCTs also makes it possible to ensure better obstetrical follow-up and, finally, to predict the delivery in a specialized structure where the newborn will be entrusted to the pediatric and surgical team informed, in optimal conditions. Genetic counseling remains useful since family cases have been reported.

#### **Fetal ultrasound (13). It allows:**

1. To appreciate the size of the tumor and its dynamic growth.
2. To specify its cystic, solid or mixed nature.
3. Look for calcification, intra-tumoral hemorrhage or necrosis.
4. To appreciate the intra-pelvic extension.
5. To make the differential diagnosis.
6. Look for malformations and associated complications.
7. To evaluate the prognosis and to orient towards the mode and the moment of delivery.
8. Doppler examination makes it possible to appreciate the importance of tumor vascularization

#### **Positive ultrasound diagnosis:**

A tumor syndrome with a mass at the distal end of the sacrum, more or less heterogeneous, with mixed contents, with hypo- and hyperechoic zones, irregular borders with cystic components and sometimes calcifications. Sometimes it is a well-limited tumor with hypoechoic content, sometimes purely cystic in appearance, sometimes heterogeneous mixed component.

#### **Ultrasound classification**

According to the classification proposed by the American Academy of Pediatric Surgical Section Survey (AAPSS).

1. type I: exopelvic tumor;
2. type II: intrapelvic tumor with exopelvic extension;
3. type III: predominantly intrapelvic tumor but visible in exopelvic;
4. type IV: exclusively endopelvic tumor.

#### **Ultrasound differential diagnosis**

The differential diagnosis of sacro-coccygeal teratomas with exclusive endo-pelvic extension will mainly occur in cystic forms, with other intra-abdominal cystic masses:

1. Obstructive uropathies
2. Obstructions or digestive duplications
3. Cystic lymphangioma
4. Ovarian cyst in female fetuses

#### **Fetal MRI (14; 15):**

MRI can provide additional information; it allows to:

1. Better evaluate the size of the tumor.
2. Specify the nature of the tumor when the ultrasound can not conclude.
3. Look for intra-tumoral hemorrhage.
4. Differential diagnosis especially with anterior meningocele.
5. Enjoy intra-pelvic or intra-spinal extension.
6. Study relationships with neighboring structures.
7. Properly classify the tumor according to the Altman classification.

The MRI aspect of the SCT is characteristic, usually a large mass containing well-defined rounded areas of variable intensity signal representing the cystic, solid or calcified components.

#### **Postnatal diagnosis**

The clinical presentation of SCT is variable, ranging from a simple induration or wheelbase or asymmetry of the size of the buttock, to a bulky mass that can reach the size of the fetus. Other signs are related to the compression of neighboring organs and degeneration with secondary locations.

#### **Standard radiography:**

On the pelvis images, the tumor is in the form of a heterogeneous opacity, multi-lobed, with fuzzy limits. Within this opacity, calcifications or even bone fragments (teeth) may exist.

#### **Ultrasound (13; 16):**

Ultrasound: The polymorphism of ultrasound images is related to the nature of the elements that make up the teratoma.

#### **Ultrasound can:**

To specify the echo-structure of the mass, it can be liquid, solid or mixed:

1. Fluid echo-structure: the ultrasound shows a rounded mass, of liquid tonality, often multilocular.
2. Solid echo-structure: this is the rarest form. The solid components have an echogenic echo structure, most often inhomogeneous, they may contain calcifications or bone fragments of variable size and shape.
3. Mixed echo-structure: this is the most common form; within an echogenic mass, we find cystic images, well limited, of variable size. Indicate the existence of calcification.
4. Show the presence of hemorrhage, single or multilocular cysts, fatty areas
5. Objectiver intra pelvic extension.
6. Identify the relationship with the bladder, the rectum
7. Explore the urinary shaft for possible bladder dilatation or ureterohydronephrosis.
8. Follow and detect any recurrence

#### **Computed tomography (17):**

It determines the tumor composition: cyst, fat, and calcifications (Figure 1).

It also allows:

1. To locate the teratoma in relation to the pelvic organs, especially the rectum and the bladder.

2. To delimit the pelvic bone structures thus to show the coccygeal attachment.
3. It shows small endo-pelvic extensions that may have eluded ultrasound (Figure 2)

**MRI: (2; 17)**

MRI allows to characterize the nature of the mass, to specify the links with the muscular and bony structures of the pelvis (the upper strait) and to reveal an intra pelvic extension although the relations with the rectum are less visible than with the scanner.

It is particularly effective for the study of the spine therefore to visualize an intra spinal extension and / or to remove the doubt on a meningocele.

**Differential diagnosis:**

The differential diagnosis of SCT is made with other sacro-coccygeal expansive processes such as: antero-cellular meningocele, cystic formations (dermoid, epidermoid, lymphangioma), lipomas, pelvic neuroblastoma, lymphoma, rhabdomyosarcoma (1 4).

**Surgical management:**

A SCT, whether diagnosed prenatally or after birth, should benefit from a surgical resection carrying the coccyx in the first days of life, to avoid the risk of malignant transformation that increases with time (2; 5). For degenerate forms or in case of incomplete excision, chemotherapy is coupled with surgery. Technological advances indicate an excision of the in utero mass or an embolization of the tumor vessels by radiofrequency (11; 12). Post-treatment follow-up is clinical and paraclinical with imaging and determination of alpha feto-protein. Monthly monitoring is recommended in the first year, every three months for at least three years, and every year thereafter.

**Conclusion: -**

Sacrococcygeal teratoma is a rare congenital embryonic tumor with a high potential for malignant transformation and therefore considered a neonatal surgical emergency. Hence the need for antenatal diagnosis and early management. Imaging remains essential for the diagnosis, the evaluation of tumor extension and the search for elements of malignancy. Early surgical treatment prevents progression to malignant transformation.

**Conflicts of interest.**

The authors do not declare any conflict of interest.

**Contributions from The Authors**

All authors contributed to this article. All authors have read and approved the final version of the manuscript.

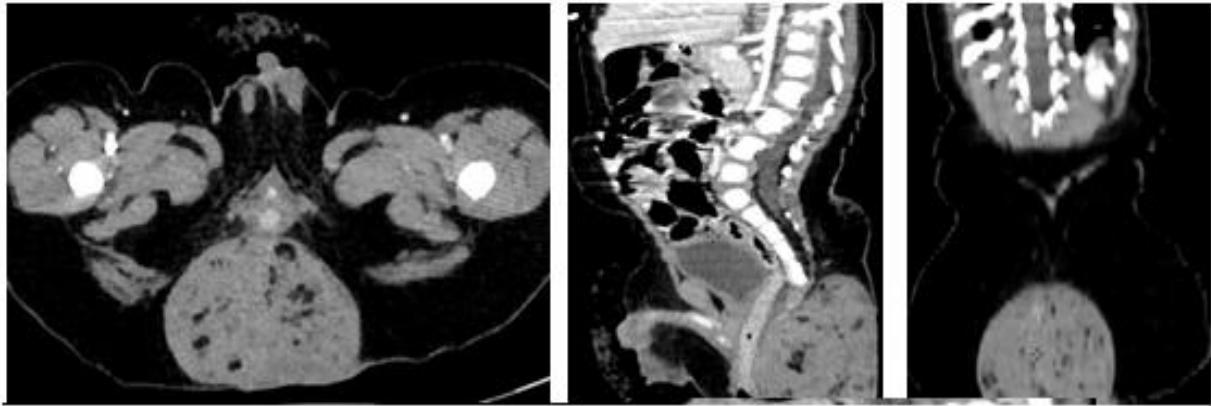


Figure 1: C + abdominal-pelvic CT scan in axial (A), sagittal (B) and coronal (C) sections showing a malignant sacrococcygeal teratoma containing an externally developing type I fatty substance of Altmann infiltrating the striated sphincter, the perianal region and retrorectal in a 9-month-old infant.

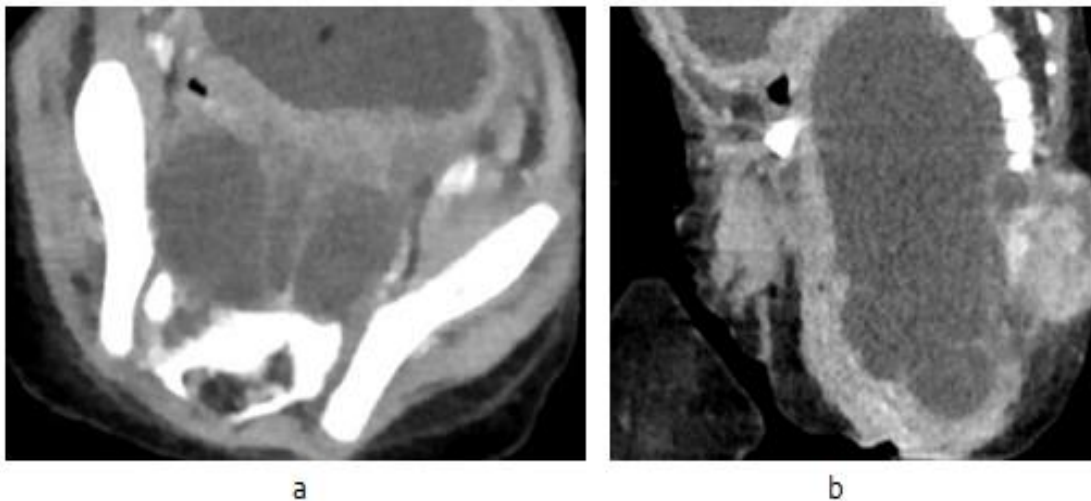


Figure 2: Abdominal C + CT (axial slices (a), coronal section (b)), an 11-month-old infant admitted for abdominal mass management with rectal and bladder compression, and CT abdominal reveals a voluminous process corneal sacrococcygeal process retro rectal heterogeneous two-component fleshy and cystic including the perineum and presents an endo-pelvic development, retro-uretero-vesical responsible for ureteral compression.

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