PRENATAL DIAGNOSIS AND MANAGEMENT OF SACROCCYGEAL TERATOMA

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Abstract

Sacrococcygeal teratoma (SCT) is one of the most common fetal tumors, with a birth prevalence varying from 1 in 22000 to 1 in 40000 live births. Recently, the perinatal outcome has improved significantly with the introduction of routine fetal anomaly scan. We present the prenatal diagnosis and management in a case of SCT that was referred to our centre at 24 weeks’ gestation. Serial fetal ultrasound evaluations were performed to assess fetal wellbeing, tumor growth and vascularization, amniotic fluid volume and to diagnose specific complications. In addition, MRI exam was performed at 32 weeks to evaluate de intrapelvic extension of the tumor. The tumor size markedly increased throughout pregnancy (from 49/30/32 mm at 24 weeks to 130/80/90 mm at 37 weeks), however the tumor vascularization was reduced and no complications were detected, except for moderate polyhydramnios in the late third trimester. A male newborn was delivered by emergency Caesarean section at 37+1 weeks’ gestation, with good Apgar scores. The tumor was resected in the early neonatal period with uneventful postoperative course and the child was doing well at the time of writing.

Rezumat: Diagnosticul şi conduita prenatală în teratomul sacrococcigian

Teratomul sacrococcigian este una dintre cele mai frecvente tumori fetale, cu o prevalenţă variind de la 1: 22000 la 1:40000 născuţi vii. Prognosticul perinatal s-a îmbunătăţit semnificativ în ultima perioadă, odată cu introducerea screeningului de rutină pentru anomalii fetale. Prezentăm diagnosticul şi conduita în cazul unei sarcini cu teratom sacrocociggian fetal, ce s-a prezentat în clinica noastră la 24 săptămâni de gestaţie. Examinări ecografice fetale seriate au fost efectuate pentru evaluarea creşterii fetale, a dimensiunilor şi vascularizaţiei tumorale şi pentru diagnosticarea complicaţiilor specifice. La 32 de săptămâni de sarcină s-a practicat examinarea fetăla prin rezonanţă magnetică pentru determinarea extensiei întrapelvine a tumorii. Deşi dimensiunile tumorale au crescut considerabil pe parcursul sarcinii (de la 49/30/32 mm la 24 săptămâni la 130/80/90 mm la 37 săptămâni), vascularizaţia tumorii a fost redusă, fără apariţia complicaţiilor specifice, cu excepţia unui polihidramnios moderat la sfârşitul trimestrului al treilea. Un nou-născut de sex masculin a fost extras prin operaţie cezariană de urgenţă la 37săptămâni + 1 zi de gestaţie cu scor Apgar bun.

Tumora a fost rezecată în totalitate după naştere cu evoluţie favorabilă postoperatorie, iar în prezent, la vârsta de 2 ani, copilul se prezintă fără semne de recidivă.

Cuvinte cheie: teratom sacrococigial, ecografie fetălu, tumori fetale, teratom.

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KEY WORDS: sacrococcygeal teratoma, fetal ultrasound, fetal tumors, teratoma
Introduction

Sacrococcygeal teratoma contains elements derived from all three germ cell layers (ectoderm, mesoderm and endoderm) and it affects approximately 1 in 27000 pregnancies, being four to nine times more common in females than in males (1-3). Although the aetiology is not completely known, sacrococcygeal tumours are believed to originate from either Hensen’s node or ectopic primordial germ cells (1-6).

Histologically, SCT’s are classified as mature (include well-differentiated mature structures such as skin, nails, hair, bone teeth etc.) and immature (undifferentiated cells, with nuclear atypia and increased mitotic activity) (1-3). The majority of SCT’s diagnosed perinatally are benign, malignancy being uncommon and frequently associated with delayed and/or incomplete postnatal resection (2,6). American Academy of Paediatrics Surgery Section (AAPSS) classified the sacrococcygeal teratomas according to development (Table 1). The classification describes the surgical anatomy and it does not carry any relevance regarding the likely prenatal course, nor the ultimate prognosis (2,3,7).

Prenatal diagnosis is typically made during routine anomaly scan, SCT’s presenting as well circumscribed complex masses, localized in close proximity to the coccyx that could demonstrate mixed echogenic structure with cystic, solid, calcified and vascular regions (3,8). The incidence of associated anomalies varies from 0% to 35%, involving nervous, cardiac, gastrointestinal, genitourinary and musculoskeletal systems. In isolated cases of SCT, aneuploidy has not been reported. (1).

Whereas some SCT’s do not cause prenatal complications, rapidly growing tumours, with rich vascularization can cause high-output cardiac failure, fetal anaemia and hydrops (4,9). The perinatal outcome has improved with advances in fetal ultrasonography that have enabled early diagnosis of SCT and associated complications.

Following the diagnosis of a SCT the patients need to be referred to a tertiary care centre specialized in multidisciplinary perinatal management of this condition (maternal–fetal medicine specialist, obstetrician, radiologist, neonatologist, paediatric surgeon).

We present the prenatal diagnosis and management and a short summary of postnatal follow-up in a case of SCT referred to our centre in the second trimester of pregnancy.

CASE REPORT

A 22-year-old woman, gravida 1 para 0 without medical or surgical history, was referred for detailed fetal ultrasound at 24 weeks’ gestation for a fetal sacrococcygeal tumour. A complex mass, with cystic and solid components, measuring 49.2/29.8/40 mm in sacrococcygeal area was identified. Doppler interrogation revealed poor vascularization, with a single feeding vessel originating probably from the middle sacral artery. No additional structural defects were seen, the fetal growth, amniotic fluid volume and Doppler studies were within normal ranges. The

<table>
<thead>
<tr>
<th>Class</th>
<th>Definition</th>
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<tr>
<td>Type I</td>
<td>Predominantly external with minimal intrapelvic extension</td>
</tr>
<tr>
<td>Type II</td>
<td>Mainly external, with significant intrapelvic extension</td>
</tr>
<tr>
<td>Type III</td>
<td>Visible external, but predominantly internal</td>
</tr>
<tr>
<td>Type IV</td>
<td>Internal, although external parts may be seen</td>
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</table>

Table 1. AAPSS Staging classification of sacrococcygeal teratomas
Echocardiography revealed normal structural and functional heart. The normal intracranial anatomy excluded an open neural tube defect. The findings were consistent with sacrococcygeal teratoma type I (Fig. 1). The patient was counselled in regard to prenatal and postnatal management, prognosis, risk of malignancy and recurrence by a multidisciplinary team including obstetrician, fetal medicine specialist, neonatologist and paediatric surgeon. We decided to perform follow-up scans every 3 to 4 weeks to evaluate the fetal wellbeing, tumour growth rate and vascularization. We also assessed the hemodynamic impact of the tumour by evaluating the cardiac function and by measuring middle cerebral artery-peak systolic velocity (MCA-PSV), in order to diagnose hyperdynamic circulation.

At 32 weeks’ gestation, fetal magnetic resonance imaging (MRI) was performed to establish the intrapelvic/intraabdominal extension of the tumour. The MRI examination revealed a complex mass measuring 110/70/77mm with cystic and solid content, well circumscribed, localized in the sacrococcygeal area, with external development and minimal intrapelvic extension up to the level of S3-S4 with no signs of local compression, confirming the diagnosis of sacrococcygeal teratoma type I (Fig. 2).

The prenatal course was uneventful, although the tumour size increased significantly throughout pregnancy. A moderate polyhydramnios was
diagnosed at 36 weeks with no other signs of specific complications (Table 2).

At 37+1 weeks’ gestation, the patient presented with regular uterine contractions, in labour. The foetus was in vertex presentation, with normal growth, normal Doppler studies and moderate polyhydramnios (deepest vertical pocket of 10 cm). The teratoma measured 130/80/90 mm with no signs of hemodynamic decompensation. An emergency caesarean section was performed due to the large tumor and the risks of rupture and haemorrhage. A male new-born of 3190 g and 49 cm was delivered with Apgar scores of 8, 9, 9 at 1, 5 and 10 minutes respectively (Fig. 3). After the neonatal first care, the new-born was transferred to the paediatric surgical center for further management. Postnatal computed tomography (CT) scan showed a perineal voluminous mass of 123/75/100 mm with cephalad extent up to vertebrae S5, enveloping the coccyx that was consistent with the diagnosis of sacrococcygeal teratoma with extra-abdominal development.

After the new-born stabilization, the tumor was resected in the third day of life with uneventful postoperative course (Fig. 4, 5). On pathology, this was an immature teratoma and the postsurgical follow-up was performed by serial measurements of tumor markers levels – serum alpha-fetoprotein (AFP) and â-human chorionic gonadotropin (â-hCG). No signs of recurrence, malignancy or functional impairment were detected till the time of writing.

**Discussion**

Advances of ultrasound techniques have made possible the accurate prenatal diagnosis of SCT, detection rates reported by large studies varying around 88 to 90 % in early second trimester (4, 8).

![Fig. 3 Postnatal aspect of sacrococcygeal teratoma type I- large exophytic external component](image)

### Table 2  Prenatal fetal ultrasound details

<table>
<thead>
<tr>
<th>Gestational age (week)</th>
<th>Tumour size(mm)</th>
<th>MCA-PSV (cm/s)</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>24+5</td>
<td>49.2/29.8/30</td>
<td>33.45</td>
<td>No</td>
</tr>
<tr>
<td>26+1</td>
<td>67.2/41.2/40</td>
<td>39.24</td>
<td>No</td>
</tr>
<tr>
<td>29+1</td>
<td>80/60/59</td>
<td>49.30 (+1 SD)</td>
<td>No</td>
</tr>
<tr>
<td>32+2</td>
<td>110/70/73</td>
<td>57.89 (+1 SD)</td>
<td>No</td>
</tr>
<tr>
<td>36+1</td>
<td>121/65/70</td>
<td>70.61 (+1 SD)</td>
<td>Polyhydramnios</td>
</tr>
</tbody>
</table>

SD- Standard deviation
Ultrasound examination with Doppler studies has an important role in assessing the location, content and possible hemodynamic repercussions of the tumor (2). However, the acoustic shadowing of the bone structures could impede the detection of intrapelvic/intraabdominal extent of the tumor. MRI exam has a superior resolution in such cases, providing detailed information about the intrapelvic extension and relationship to adjacent structures (2,3).

Prenatal diagnosis of SCT should prompt referral to a multidisciplinary team experienced in perinatal management of this condition. Parents could be reassured that in the majority of the cases, infants with SCT have excellent prognosis, only few of them developing specific prenatal or postsurgical complications. Genetic testing is not mandatory in isolated SCT, as the incidence of abnormal karyotype is extremely low (9).

Serial prenatal ultrasound imaging is important in identifying cases at high risk for developing fetal and obstetrical complications. The most important prognostic factors are tumor size, tumor growth rate and vascularization. Small and poorly vascularized tumours will not have a significant impact on the fetus, whereas highly vascularized and fast growing tumours are associated with adverse outcomes including high output cardiac failure, prematurity and fetal death. The latter type of tumours increases the metabolic demands and the circulating volume, causing fetal anaemia, high output cardiac failure, polyhydramnios and fetal hydrops, increasing the risk for maternal “mirror” syndrome. The pathophysiologic mechanism is referred to as “vascular steal”, similar to that seen in the arteriovenous malformations such as chorioangioma or Galen vein aneurysm (2,3,10). Less common causes of fetal anaemia could be intratumoral or intraamniotic haemorrhage (2).

In SCT’s associated with fetal complications, few symptomatic prenatal interventions have been described. Amniodrainage in sever polyhydramnios can prevent premature labour and reduce maternal discomfort, while fetal anaemia can be diagnosed measuring MCA-PSV and easily corrected by intrauterine fetal transfusion in order to improve fetal outcome. Only limited experience is available concerning intrauterine surgical procedures such as intratumoral cyst aspiration, sclerosis or ablation of the feeding vessels, tumor embolization, or shunting.
in cases of secondary urinary tract obstructions, these
being performed in selected cases in specialized
centres (1-3).

The time and mode of delivery depends on
tumor size and/or configuration and associated
maternal and fetal complications. In absence of
obstetrical indication, vaginal delivery is an option for
small tumours. Elective Cesarean delivery at term is
recommended for SCT’s measuring more than 5 cm
in diameter to reduce the risk of dystocia, tumor
rupture, haemorrhage and fetal death (3). After
neonatal stabilization and postnatal diagnosis, tumor
excision is usually performed during the first days of
life. Immediate resection in early neonatal period is
critical, as delayed and incomplete resection is
associated with high risk of malignancy and
recurrence (2-4). Postoperative follow-up is important
not only for detection of recurrence or malignancy,
but also for early diagnosis and treatment of pelvic
floor dysfunction. Functional urinary or rectal
problems may be more frequent when a significant
portion of the tumour is located in the pelvis, due to
sacral nerves and pelvic plexus stretching. In addition,
secondary compression lesions of the pelvic could
develop in utero or following surgical excision (2).

References

1. Mancuso MS, Biggio J. Fetal Tumors. In: James D, Steer
High risk pregnancy management options. Elsevier

2. Gucciardo L, Uyttebroek A, De Wever I, Renard M, Calus
F, Devlieger R, Lewi L, De Catte L, Feprest J. Prenatal
assessment and management of sacrococcygeal teratoma.

Management of fetal teratomas. Pediatr Surg Int 2006; 32:
635-647.

4. Lee MY, Won HS, Hyun MK, Lee HY, Shim JY, Lee PR,
Kim A. Perinatal outcome of sacrococcygeal teratoma.
Prenat Diagn 2011; 31: 1217-1221.

JC, Baud D, O’Brien K, Beecroft R, Chaturvedi R, Jaeggi E,
Fish J, Ryan G. Minimally invasive therapy for fetal
sacrococcygeal teratoma: case series and systematic
review of the literature. Ultrasound Obstet Gynecol 2014;
43: 611-619.

over two decades: Birth prevalence, prenatal diagnosis

7. Deprest J, Flake AW, Gratacos E, Ville Y, Hecher K,
Nicolaides K, Johnson MP, Luks FI, Adzick NS, Harrison
MR. The making of fetal surgery. Prenat Diagn 2010; 30:
653-667.

8. Makin EC, Hyett J, Ade-Ajayi. Outcome of antenatally
diagnosed sacrococcygeal teratomas: single centre

N. Sacrococcygeal teratoma in a fetus with prenatally
diagnosed partial trisomy 10q (10q24.3->qter) and partial

10. Benachi A, Durin L, Maurer SV. Prenatally diagnosed
sacrococcygeal teratoma: a prognostic classification. J
Pediatr Surg 2006; 41: 1517-1521.