

Prenatal diagnosis of sacrococcygeal teratoma: A review of cases between 1993 and 2000

R. Axt-Fliedner¹, H. J. Hendrik¹, H. Reinhard², A. K. Ertan¹, M. Friedrich¹,
K. Remberger³, W. Schmidt¹

¹Division of Prenatal Diagnosis and Ultrasound, Department of Obstetrics and Gynecology;

²Division of Pediatric Oncology and Hematology, Department of Pediatrics;

³Department of General Pathology, Medical School, University of the Saarland, Homburg/Saar (Germany)

Summary

Sacrococcygeal teratoma is the most common fetal neoplasm with a reported incidence of 1 in 30,000 to 40,000 births. Affected fetuses carry a high perinatal mortality and morbidity. The aim of this retrospective study was to assess prenatal sonographic aspects and pathological details of our cases with sacrococcygeal teratoma. Over the last seven years we identified six cases by retrospective chart review in our institution. Four fetuses were electively aborted, in two of these four fetuses the diagnosis was made before the 16th week of gestation. Two caesarean sections were performed at 35+5 and 37+0 weeks of gestation, respectively. In those two cases the diagnosis was only made in the late second and third trimester. No case of neonatal mortality occurred. In one of the two resected sacrococcygeal teratomas potential malignancy was diagnosed. A multidisciplinary approach seems advisable for optimal perinatal management.

Key words: Fetal tumour; Sacrococcygeal teratoma; Prenatal ultrasound.

Introduction

Sacrococcygeal teratomas consist of tissue from all three germinal cell layers, ectoderm, endoderm and mesoderm, and are known to be the most common congenital tumour occurring in one of 30,000 to 40,000 births [1-7]. Sacrococcygeal teratomas appear three to four times more frequently in females than in males. Histological classification of these tumours includes different forms of mature and immature types with later risk of malignancy and primarily malignant types. In 1974 Altman *et al.* presented a classification of sacrococcygeal teratomas with regard to their location [2]. Type I tumours are mostly external with only a minimal pre-sacral component and represent the commonest type. Type II tumours are partly external and partly intrapelvic masses. Type III tumours are visible externally but the predominant mass is located within the pelvis extending into the abdomen. Type IV tumours are completely intrapelvic without any external component. Associated anomalies and chromosomal aberrations are rare [7-9]. Perinatal mortality and morbidity are mostly related to high-output cardiac failure because of arteriovenous shunting within the tumour, subsequent fetal hydrops, polyhydramnios and preterm delivery [4-7]. Other risk factors include cervical dystocia during vaginal delivery, tumor rupture, fetal hemorrhage or hemolysis caused by the tumor and placentomegaly. The prognosis of fetal sacrococcygeal teratoma detected antenatally has been reported as poor [8-10]. Nevertheless, improved fetal ultrasonographic scanning might help to optimize paren-

tal counselling as well as pre-, peri-, and postnatal management by identifying sonographic factors associated with adverse or favourable outcome of the fetus. We report our experience of six cases with sacrococcygeal teratoma managed at our institution over the last seven years.

Materials and Methods

A retrospective analysis of prenatally diagnosed fetal sacrococcygeal teratomas between January 1993 and September 2000 was performed. Six cases were included. Maternal and neonatal records as well as ultrasound scans were reviewed. Information from these data included: gestational age at diagnosis, gestational age at delivery, initial tumour size, maximal tumour size, associated anomalies, antenatal complications, development of fetal hydrops, presence of polyhydramnios calculated from the amniotic fluid index, appearance of the tumour with regard to solid and cystic components, result of amniocentesis and AFP- and ACHE-value, mode of delivery, Apgar scores, arterial pH, histologic result of the surgically excised tumours and results of the post-mortem examination of the aborted fetuses. The Division of Prenatal Diagnosis and Ultrasound is a regional referral institution. All suspected fetal anomalies have a targeted ultrasound scan. Follow-up ultrasound scans are scheduled, if necessary, in short intervals and appointments for the parents together with the neonatologist and pediatric surgeon are offered. A multidisciplinary team including an obstetrician, a neonatologist, a pediatric surgeon and an experienced anesthetist is present in the delivery room. Ultrasound scans are performed with Siemens-Sonoline-Elegra (Siemens, Erlange, Germany) or Acuson-XP-128 (Acuson, Mountain View, CA, USA) equipment.

Revised manuscript accepted for publication August 1, 2001

Table 1. — Antenatal parameters of six fetuses with sacrococcygeal teratoma.

Case	GA at diagnosis (weeks)	GA at delivery (weeks)	Tumour size at diagnosis (cm)	Tumour size at delivery (cm)	Type	Hydrops	Polyhydramnios	Associated US anomalies	Amniocentesis
1	26.1	35.5	6.0x4.0x6.0	13.0x10.0x11.0	II, cystic>solid	no	yes	bilateral hydronephrosis	46, XY; AFP: 0.87 MOM; ACHE: negative
2	31.1	37.0	9.2x10.7	13.0x12.1x10.5	II, cystic	no	no	—	—
3	15.0	15.5	2.6x1.9x1.8	3.2x2.2	I, cystic>solid	no	no	pes equinovarus	—
4	22.4	22.5	5.5x6.3x7.1	5.4x6.5x7.5	I, cystic-solid	no	no	—	—
5	21.0	22.2	5.5x7.2	7.0x7.2	I, cystic-solid	no	no	—	46, XY; AFP: 1.56 MOM; ACHE: negative
6	14.2	14.6	3.5x4.8x5.8	3.6x5.5x6.2	I, solid	no	no	thoraco-gastroschisis	—

GA: gestational age; US: ultrasound.

Results

In six cases the diagnosis of sacrococcygeal teratoma was made during the study period after referral for detailed ultrasound. One case with suspected sacrococcygeal teratoma by the referring gynecologist had to be excluded because targeted ultrasound at our institution did not confirm the suspected diagnosis. The male to female ratio was 1:2. The median maternal age at diagnosis was 31 years (range 22 to 38). The median gestational age at diagnosis was 21.6 weeks (range 14.2 to 31.1 weeks of gestation). Antenatal characteristics are summarized in Table 1. There were two live births and four terminations of pregnancy. The diagnosis in the two surviving cases was only made after 26 weeks of pregnancy, whereas the latest diagnosis of sacrococcygeal teratoma in the four terminated pregnancies was made at 22.4 weeks of gestation. There was neither a stillborn baby nor a neonatal death (Table 1).

In subject 1, the tumour initially measured 6.0 cm x 4.0 cm x 6.0 cm at 26.1 weeks of gestation and increased rapidly to 13.0 cm x 10.0 cm x 11.0 cm at 35.5 weeks of gestation. Additionally, bilateral hydronephrosis was diagnosed. At 26.4 weeks of gestation a genetic amniocentesis was performed with the fetal karyotype being normal, alpha-feto-protein (AFP) being at 0.87 MOM and acetyl-cholin-esterase (ACHE) being negative. The type II tumour appeared to be cystic with few solid components and showed intensive vascularization on Doppler flow study. The rapidly growing teratoma was associated with the development of polyhydramnios with subsequent preterm labour. Neither cardiomegaly nor hydrops fetalis occurred. Amniocentesis for volume reduction was performed twice at 29.6 and 33.5 weeks of gestation. A fetomaternal Doppler flow examination at 31.5 and 35.1 weeks of gestation was normal. After repeated induction of fetal lung maturity elective caesarean section was performed at 35.5 weeks of gestation. Vaginal delivery was considered inappropriate because of the tumour size and the risk of fetal trauma or fetal hemorrhage. A male newborn, arterial pH 7.29, Apgar score 8, 10, 10 at 1 min, 5 min and 10 min, respectively, weighing 3,060 g (with

tumour) was delivered. After stabilization of the neonate the tumour was resected completely by the pediatric surgeon on the second day of life. Histology revealed a potentially malignant cystic sacrococcygeal teratoma weighing 1,170 g with 60% mature and 40% immature components.

In subject 2, the tumour measured 9.2 cm x 10.7 cm at initial diagnosis at 31.1 weeks of gestation increasing to 13.0 cm x 12.1 cm x 10.5 cm at 37.0 weeks of gestation and consisted predominantly of cystic components. This was a type II sacrococcygeal teratoma. There were no additional anomalies nor signs of fetal hydrops or cardiomegaly. Fetomaternal Doppler flow examination was unremarkable at 32.6 and 35.6 weeks of gestation. After repeated induction of fetal lung maturity elective caesarean section was performed with 37.0 weeks of gestation because the size of the tumour precluded a vaginal delivery. A female newborn, arterial pH 7.40, Apgar score 7, 9, 9 at 1 min, 5 min and 10 min, respectively, weighing 3,990 g (with tumour) was delivered. Complete resection of the tumour was performed on the third day of life after stabilization of the newborn. Histologic examination

Table 2. — Histology, post-mortem findings and outcome features.

Case	Histology findings	Post-mortem	Outcome
1	1,170 g, 60% mature, 40% immature components, potentially malignant	—	elective caesarean section, live birth, normal follow-up at 6 years
2	930 g, mature benign teratoma, 14x12x12 cm, hydrops placentae	—	elective caesarean section, live birth, normal follow-up at two months
3	immature benign teratoma	pes equinovarus	TOP
4	immature benign teratoma	—	TOP
5	78 g, immature benign teratoma	fetus of 462 g, no further malformations	TOP
6	immature benign teratoma	complete eventration of stomach, liver, heart	TOP

TOP: termination of pregnancy.

revealed a mature benign teratoma weighing 930 g and hydrops of the placenta. This newborn is now eight weeks old and has recovered without problems from the operation.

In one of the terminated pregnancies severe additional malformations were found (thoraco-gastroschisis, ectopia cordis). In three of the four terminated cases intense arterial and venous perfusion of the tumour was seen by Doppler flow. In all terminated pregnancies parents were counselled extensively together with the neonatologist and pediatric surgeon.

Discussion

Sacrococcygeal teratomas are tumours deriving from multipotent cell lines containing embryonic ectodermal, endodermal and mesodermal components [4]. Perinatal mortality is high when sacrococcygeal teratomas are diagnosed prenatally mainly due to high-cardiac output failure associated with arterio-venous shunting and fetal hydrops [8-10]. Although rare, malignant fetal sacrococcygeal teratoma in utero has been described [11]. Sacrococcygeal teratomas diagnosed accidentally at birth have a favourable outcome. According to Altman *et al.*, over 90% of the latter group are full-term births with an overall mortality of 7% being primarily related to tumour haemorrhage or malignant invasion of the tumour [2].

About 40% of germ cell tumors in childhood present as sacrococcygeal teratomas. Of these tumors, 70% are in females and just over half occur in neonates. Histological examination reveals potential malignancy in 30% of the cases and the tendency of malignancy is greater in males [2].

Although rare, malignant fetal sacrococcygeal teratoma in utero has been described [11]. Perinatal mortality varies between 12% and 68% according to different authors.

The prenatal sonographic detection of sacrococcygeal teratoma is based on the identification of a tumour on the sacral pole of the fetus or an intraabdominal caudal mass and might be of cystic, solid or mixed appearance [4, 11, 12]. Associated sonographic findings may be polyhydramnios, placentomegaly, hydrops fetalis, hydronephrosis or bladder anomalies. The differential diagnosis includes urogenital anomalies and caudal myelomeningocele [4-7].

With the more frequent application of second trimester anomaly screening the diagnosis is being made earlier in pregnancy [4, 13, 14]. In our series, as in others, there was a wide range of gestational age at which the diagnosis was made. Sonographic detection of fetal sacrococcygeal teratoma during the first trimester has been reported to be difficult because of small tumour size and often diagnosis is made only later in gestation when the tumour increases remarkably in size [4, 13, 14]. In our small series the earliest diagnosis of a fetal sacrococcygeal teratoma was made at 14.2 weeks of gestation at an early second trimester anomaly screening by vaginal ultrasound. Kirkinen *et al.* reported that prenatal MRI was useful in the antenatal topographic evaluation of the

intrapelvic part of fetal sacrococcygeal teratoma but reliable differentiation of mature and immature sacrococcygeal teratoma was not possible [14].

The size of fetal sacrococcygeal teratoma is variable. In the two surviving fetuses the tumour consisted predominantly of cystic components and increased remarkably in size during gestation. In one of the two surviving fetuses bilateral hydronephrosis was observed eventually due to a local compression effect of the tumour but we did not see any cardiovascular sequelae in the fetuses. This is in accordance to the known association of large vascular tumours with the development of cardiac hypertrophy, high-output cardiac failure, pleural and pericardial effusions, placentomegaly, hydrops fetalis and fetal anasarca [4-6]. These complications have been regarded as preterminal with an associated mortality of up to 100%. Therefore only a few authors have reported on intrauterine intervention as a therapeutic alternative in cases with early hydrops fetalis or rapidly growing sacrococcygeal teratoma, with different outcomes [15-17]. Langer *et al.* reported on successful intrauterine resection of fetal sacrococcygeal teratoma by hysterotomy in a case with a large tumour and fetal hydrops early in gestation (21st week of gestation), but preterm labour led to the delivery of a preivable infant [15]. In another case report Garcia *et al.* described the in utero decompression of a cystic grade IV sacrococcygeal teratoma at 22 weeks of gestation ending in preterm delivery at 28.5 weeks of gestation [16]. Hecher and Hackeloer repeatedly performed intrauterine laser surgery to interrupt the blood supply of a sacrococcygeal teratoma at 20 weeks and 26 weeks of gestation resulting in a live birth at 37 weeks of gestation [17]. To our knowledge there are no data dealing with intrauterine therapy of sacrococcygeal teratomas that turned out malignant. We feel that this point should be addressed clearly in case of intrauterine surgery for sacrococcygeal teratoma.

Despite the texture of fetal sacrococcygeal teratoma (cystic and poorly vascularized or solid and highly vascularized with possible subsequent hydrops fetalis), other factors having an impact on perinatal survival in cases of fetal sacrococcygeal teratoma are described in the literature [1, 5, 6, 10, 11]. Brace *et al.* recently reported that in the absence of polyhydramnios or hydrops fetalis 70% of affected fetuses survived and conversely only 7% and 25% survived, respectively, in the presence of hydrops and preterm delivery < 34 weeks of gestation. Late prenatal diagnosis (> 30 weeks of gestation) and a tumour size of less than 10 cm at delivery were associated with a survival rate greater than 80% [4]. The diagnosis of sacrococcygeal teratoma in the two surviving fetuses of our report was also only made in the third trimester. In our series one of six patients underwent repeated amniocentesis for decompression and caesarean section was performed at 35.5 weeks of gestation.

Recommendations of the optimal mode of delivery have been made with regard to the size of tumour. Gross *et al.* suggest caesarean section in cases with sacrococcygeal teratomas > 5 cm, Brace *et al.* recommend

performing a caesarean section in cases with a tumour > 10 cm except cystic tumours where aspiration under ultrasound control might allow vaginal delivery [1, 4]. The aim should be in any case an atraumatic delivery of the fetus and to minimize the risk of tumour rupture and fetal haemorrhage.

Children with malignant sacrococcygeal teratoma need chemotherapy in case of primary inoperability. Prognosis of sacrococcygeal teratoma is pretty good if the correct diagnosis is established within two months and complete resection is possible [2]. Long-term deficits concerning urinary and anorectal functions have been described [18, 19].

Modern perinatal management of fetal sacrococcygeal teratoma requires a multidisciplinary approach including a specialist in prenatal diagnosis, an obstetrician, a neonatologist, a pediatric oncologist as well as the pediatric surgeon. Serial ultrasound examinations are necessary to assess tumour growth and to detect fetal compromise. In addition, Doppler flow is helpful in monitoring blood flow through the tumour and to diagnose fetal cardiac decompensation.

In conclusion, fetal sacrococcygeal teratoma remains a congenital anomaly with high perinatal mortality and morbidity. First trimester anomaly screening might become a valuable tool in detecting fetal sacrococcygeal teratoma at early stages of gestation. This could have an important impact on parental counselling and appropriate management of the affected pregnancies.

If possible, delivery should be delayed to allow for fetal lung maturity. In case of fetal hydrops or rapid increase of tumour size elective delivery after administration of corticosteroids seems advisable. Termination of pregnancy should be discussed if fetal hydrops or a large tumour occurs early in gestation.

References

- [1] Gross S. J., Benzie R. J., Sermer M., Skidmore M. B., Wilson S. R.: "Sacrococcygeal teratoma: Prenatal diagnosis and management". *Am. J. Obstet. Gynecol.*, 1987, 156, 393.
- [2] Altman R. P., Randolph J. G., Lilly J. R.: "Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey-1973". *J. Pediatr. Surg.*, 1974, 9, 389.
- [3] Sepulveda W. H.: "Prenatal sonographic diagnosis of congenital sacrococcygeal teratoma and management". *J. Perinat. Med.*, 1989, 17, 93.
- [4] Brace V., Grant S. R., Brackley K. J., Kilby M. D., Whittle M. J.: "Prenatal diagnosis and outcome in sacrococcygeal teratomas: a review of cases between 1992-1998". *Prenat. Diagn.*, 2000, 20, 51.
- [5] Holterman A. X., Filiatrault D., Lallier M., Youssef S.: "The natural history of sacrococcygeal teratomas diagnosed through routine obstetrics sonogram: a single institution experience". *J. Pediatr. Surg.*, 1998, 33, 899.
- [6] Westerburg B., Feldstein V. A., Sandberg P. L., Lopoo J. B., Harrison M. R., Albanese C. T.: "Sonographic prognostic factors in fetuses with sacrococcygeal teratoma". *J. Pediatr. Surg.*, 2000, 35, 322.
- [7] Chisholm C. A., Heider A. L., Kuller J. A., von Allmen D., McMahon M. J., Chescheir N. C.: "Prenatal diagnosis and perinatal management of fetal sacrococcygeal teratoma". *Am. J. Perinatol.*, 1998, 15, 503.
- [8] Flake A. W., Harrison M. R., Adzick N. S., Laberge J. M., Warsaw S. L.: "Fetal sacrococcygeal teratoma". *J. Pediatr. Surg.*, 1986, 21, 563.
- [9] Flake A. W.: "Fetal sacrococcygeal teratoma". *Semin. Pediatr. Surg.*, 1993, 2, 113.
- [10] Bond S. J., Harrison M. R., Schmidt K. G., Silverman N. H., Flake A. W., Slotnick N. *et al.*: "Death due to high-output cardiac failure in fetal sacrococcygeal teratoma". *J. Pediatr. Surg.*, 1990, 25, 1287.
- [11] Sheth S., Nussbaum A. R., Sanders R. C., Hamper U. M., Davidson A. J.: "Prenatal diagnosis of sacrococcygeal teratoma: sonographic-pathologic correlation". *Radiology*, 1988, 169, 131.
- [12] Winderl L. M., Silverman R. K.: "Prenatal identification of a completely cystic internal sacrococcygeal teratoma (type IV)". *Ultrasound Obstet. Gynecol.*, 1997, 9, 425.
- [13] Tongsong T., Wanapirak C., Piyamongkol W., Sudasana J.: "Prenatal sonographic features of sacrococcygeal teratoma". *Int. J. Gynaecol. Obstet.*, 1999, 67, 95.
- [14] Kirkinen P., Partanen K., Merikanto J., Rynänen M., Haring P., Heinonen K.: "Ultrasonic and magnetic resonance imaging of fetal sacrococcygeal teratoma". *Acta Obstet. Gynecol. Scand.*, 1997, 76, 917.
- [15] Langer J. C., Harrison M. R., Schmidt K. G., Silverman N. H., Anderson R. L., Goldberg J. D. *et al.*: "Fetal hydrops and death from sacrococcygeal teratoma: rationale for fetal surgery". *Am. J. Obstet. Gynecol.*, 1989, 160, 1145.
- [16] Garcia A. M., Morgan W. M., Bruner J. P.: "In utero decompression of a cystic grade IV sacrococcygeal teratoma". *Fetal. Diagn. Ther.*, 1998, 13, 305.
- [17] Hecher K., Hackeloer B. J.: "Intrauterine endoscopic laser surgery for fetal sacrococcygeal teratoma". *Lancet*, 1996, 347, 470.
- [18] Rintala R., Lahdenne P., Lindahl H., Siimes M., Heikinheimo M.: "Anorectal function in adults operated for a benign sacrococcygeal teratoma". *J. Pediatr. Surg.*, 1993, 28, 1165.
- [19] Havranek P., Rubenson A., Güth D., Frenckner B., Olsen L., Kornfält S. A., Hansson G.: "Sacrococcygeal teratoma in Sweden. A 10-year national retrospective study". *J. Pediatr. Surg.*, 1992, 27, 1447.

Address reprint requests to:
 R. AXT-FLIEDNER, M.D.
 Division of Prenatal Diagnosis and Ultrasound
 Department of Obstetrics and Gynecology
 University of the Saarland
 66424 Homburg/Saar (Germany)