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Outcome of antenatally diagnosed sacrococcygeal teratomas: single-center experience (1993-2004)

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Abstract

Aims: Sacrococcygeal teratomas (SCTs) are the commonest neonatal tumors with an incidence of approximately 1:30,000. There are few large single-center series and even fewer describing both their antenatal and postnatal course. We report the outcome of all fetuses investigated at a tertiary fetal medicine center with this diagnosis.

Method: Demographic details were obtained from a prospectively maintained database. Patient records were examined for additional data including antenatal and postnatal interventions. Data were described as median (range).

Results: Forty-one SCTs were diagnosed antenatally during the period 1993 to 2004. Twelve were excluded from subsequent analysis (single antenatal visit or attending for second opinion [n = 6] and termination of pregnancy [n = 6]). Twelve underwent fetal intervention (laser vessel ablation [n = 4], alcohol sclerosis [n = 3], cyst drainage [n = 2], amniodrainage [n = 2], vesicoamniotic shunt [n = 1]) for fetal hydrops and polyhydramnios to aid in delivery and to prevent obstructive uropathy developing in the fetus. Of these, 3 died in utero and 9 survived to be born (median gestational age, 33 weeks [27-37 weeks]). A further 3 died in the neonatal period. There are 6 long-term survivors (50%) from this group. Seventeen infants, without intervention, were born at median gestational age 38 weeks (26-40 weeks). One infant with severe cardiac anomalies died on the day of birth. All surviving infants had definitive excisional surgery at a median of 2 days (1-16 days). Current median follow-up of survivors is 39 months (8-86 months). There have been no recurrences. One child has mild constipation, and 3 are awaiting cosmetic revision of their scars.

Conclusions: The overall survival of antenatally diagnosed SCT is approximately 77%, with the development of hydrops and others requiring in utero intervention carrying a poor prognosis. Otherwise, the outcome after surgical excision is excellent.

A sacrococcygeal teratoma (SCT) is the commonest solid neonatal tumor with an incidence of approximately 1:30,000 live births [1]. Their embryological origin is from the pluripotent cells in Hensen's node of the primitive streak and contain components arising from the 3 layers: endo-

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Table 1 Outcome of pregnancies with SCT that had some antenatal intervention

Case	Indication for intervention	Intervention	GA delivery (wk)	Outcome
1	Polyhydramnios	Amniodrainage	31	Alive
2	Fetal bladder outflow obstruction	Vesicoamniotic shunt inserted	33	Alive
3	To facilitate delivery	Cyst drainage	37	Alive
4	Polyhydramnios (also cardiomegaly)	Amniodrainage	34	Alive
5	To facilitate delivery	Cyst drainage	36	Alive
6	Hydrops	Laser ablation	32	Alive
7	Hydrops	Laser ablation	24	IUFD
8	Hydrops	Laser ablation	32	IUFD
9	Hydrops	Laser ablation	28	NND
10	Hydrops	Alcohol sclerosis	27	IUFD
11	Hydrops	Alcohol sclerosis	32	NND
12	Hydrops	Alcohol sclerosis	27	NND

IUFD indicates intrauterine fetal death; NND, neonatal death.

derm, mesoderm, and ectoderm. Their midline distribution can be explained by the arrest or aberrant migration of the primordial germ cells [2]. Such pluripotent cells differentiate into embryonic (mature and immature teratomas) and extraembryonic (choriocarcinoma or yolk sac teratoma), with the latter associated with the increased risk for malignant transformation [3]. Metastatic spread is rare and is found in the regional and abdominal lymph nodes, liver, lungs, and very infrequently the vertebrae and bone marrow [3]. The commonest site for extragonadal teratomas is the sacral region, followed by the anterior mediastinum, pineal, retroperitoneum, neck, stomach, and vagina [4].

Sacrococcygeal teratomas diagnosed postnatally have historically been associated with an excellent prognosis after surgical excision [5,6]. However, the diagnosis is now more commonly made within the late second and third trimesters with the advent of improved antenatal imaging techniques and screening. More recent reviews in this population recognize the increased incidence of complications of pregnancy and a high rate of fetal mortality [7-10]. Fetal demise is associated with the onset of high-output cardiac failure secondary to the metabolic demands and vascular steal of the growing tumor. This leads to the development of polyhydramnios, cardiomegaly, and fetal hydrops, resulting in either intrauterine death or preterm labor of an infant in extremely poor condition. If these complications arise before 37 weeks' gestation then mortality approaches 100% [9-13].

The aim of this article was to review the current outcome of antenatally diagnosed SCT from a single-center perspective, particularly their overall survival and specifically in those fetuses that required antenatal intervention. An assessment of functional outcome was also made.

1. Materials and methods

A retrospective analysis of antenatally diagnosed SCTs between 1993 and 2004 at Kings College Hospital, London, was performed. Demographic details were collected from a prospectively maintained fetal medicine database. Hospital and general practitioner records were assessed to collate information regarding antenatal and postnatal interventions, outcome, and long-term follow-up.

After local diagnosis by routine ultrasound in the mid second trimester, women were referred to the Harris Birthright Research Centre for Fetal Medicine to confirm the presence of an SCT and plan the need for antenatal intervention if required. Ultrasound assessment included measurement of the size of external and internal parts and its composition (solid, cystic, or mixed), and were subsequently assigned a type corresponding to the Altman criteria. Vascularity of the tumor was also assessed by Doppler scanning. Serial measurements were used to assess the rate of growth and change in vascularity of the tumor and to identify the early signs of developing hydrops or complications caused by the compression of the tumor. Intrauterine intervention was offered if serial antenatal surveillance identified significant fetal compromise. All intrauterine interventions involved percutaneous ultrasound–guided or fetoscopic techniques.

After delivery, surgical resection was performed via a perineal approach in all cases. A Mercedes/inverted "Y" or chevron incision was made, and skin flaps were mobilized. In each case, there was preservation of the anorectal muscle sling. The wound reconstruction resulted in a predominantly sagittal scar with buttock reconfiguration. Excision of the coccyx was performed in all cases [14].

All infants then entered a follow-up program based on serial α-fetoprotein estimations and at least 1 magnetic resonance imaging scan of the pelvis during the first year of life. Yearly review thereafter consisted of clinical examination supplemented by urologic, neurologic, or orthopedic review where necessary.

Data are expressed throughout as median (range).

2. Results

There were 41 antenatally diagnosed SCTs in this series. Twelve of these were excluded from subsequent analysis; 6 because the parents elected to terminate the pregnancy on confirmation of the diagnosis, and 6 that were seen antenatally on a single occasion in our unit to give a second opinion about further management before being delivered elsewhere. The median maternal age for the remaining 29 cases was 29 years (25-46 years), and median gestational