Objective: Sacrococcygeal teratoma (SCT) is the most common congenital tumor of the newborn. About 20% of SCTs are malignant. We report the case of a dizygotic twin with a large immature SCT who was successfully delivered by cesarean section at 34 weeks’ gestation and who underwent surgical removal of the tumor.

Case Report: A 21-year-old, gravida 2, para 0, abortus 1, woman was referred to Chung Shan Medical University (CSMU) Hospital at 33 weeks’ gestation because of the diagnosis of a twin pregnancy with SCT in one twin. There were no remarkable findings by ultrasound examination during the second trimester. At 30 weeks of gestation, a sacral mass measuring 8.4 cm in diameter and with multi-cystic and solid components attached to the coccyx was identified sonographically. The patient was referred to CSMU Hospital and underwent planned cesarean section at 34 weeks of gestation. The sick twin was delivered smoothly and underwent surgical resection of the tumor at 1 week of age after magnetic resonance imaging studies. Immature SCT with free surgical margins was confirmed by pathologic examination. The infant did well postoperatively.

Conclusion: SCT diagnosed before birth can be managed by planned abdominal delivery and postnatal surgery. Patients with immature teratomas should be followed-up long-term for recurrence and distant metastases.

Key Words: immature teratoma, postnatal surgery, sacrococcygeal teratoma, twin pregnancy

Introduction

Sacrococcygeal teratoma (SCT) is a rare subset of germ cell neoplasms, but is the most common congenital tumor of the newborn, with an incidence of one in 35,000–40,000 live births. SCT is more common in female than male children, and may occur in twin pregnancies [1,2]. Antenatal diagnosis of SCT can be made by ultrasound during the second trimester or even earlier in the first trimester [3,4]. SCT parasitizes blood supply from the internal and external iliac systems and results in vascular shunting to the rapidly growing tumor. Large tumors during early gestation cause significant prenatal mortality due to placentomegaly, fetal hydrops or high-output cardiac failure [2,3]. Most SCTs are benign, but about 20% are malignant, though this is rare in the neonatal period (i.e. <1 month old) [2]. Complete surgical excision of the tumor is the appropriate primary therapy in patients with SCT and should avoid damage to pelvic-associated structures [2]. We describe a dizygotic twin who was diagnosed with fetal SCT by prenatal ultrasound examination, and who underwent postnatal surgical removal of the tumor after planned cesarean section.

Case Report

A 21-year-old, gravida 2, para 0, abortus 1, woman was diagnosed with a fetal SCT in a dizygotic twin pregnancy at 30 weeks of gestation by ultrasound examination.
There were no remarkable findings during regular prenatal care. At 16 weeks of gestation, maternal α-fetoprotein (AFP) was 226.83 ng/mL and human chorionic gonadotropin (hCG) was 106,482.77 mIU/mL (Down syndrome risk, 1/1,739; neural tube defect risk, 1/305). At 30 weeks of gestation, a sacral cystic mass measuring 8.4 cm in diameter and with solid components was identified incidentally by ultrasound in the local obstetric clinic.

The patient was referred to the obstetrics and gynecology clinic of Chung Shan Medical University Hospital for further evaluation at 33+2 weeks’ gestation due to rapid growth of the tumor. A large SCT (11.9 × 9.5 cm) in one twin fetus was diagnosed by abdominal ultrasound at the time of the clinic visit (Figure 1A). After detailed ultrasound examination, vascularization in the central solid component was identified, but there was no appreciable vascular shunting within the tumor (resistance index, 1.47; pulsatility index, 2.65) (Figure 1B). Neither cardiomegaly nor genitourologic anomalies were found under ultrasound examination.

At 34+5 weeks of gestation, the estimated body weight of each twin was greater than 2,000 g, and planned cesarean section was performed. Twin A (female; birth weight, 2,500 g; Apgar scores, 7 and 9 at 1 and 5 minutes) and twin B with a large sacral mass (female; birth weight, 2,694 g; Apgar scores, 7 and 9 at 1 and 5 minutes) were delivered smoothly (Figure 2A). Twin A was normal in appearance without congenital anomalies. Twin B presented with an external sacral mass 15 cm in diameter, and the anus was displaced anteriorly (Figure 2B); otherwise, no additional congenital anomalies were identified.

Tumor markers of the delivered twins demonstrated markedly high serum AFP (twin A, 50,028.1 ng/mL; twin B with sacral tumor, 41,054.5 ng/mL). Magnetic resonance imaging (MRI) revealed a large external mass with multi-cystic and solid components, and no pelvic or intra-abdominal mass in twin B (Figure 1C). The neonatal SCT was categorized as Altman’s type I according to the location of the tumor [2].

After consultation with the pediatric surgeon, twin B underwent complete surgical removal of the tumor at 1 week of age. Immature solid teratoma composed of immature embryonal-type tissue mixed with mature tissues was confirmed by pathologic examination. The

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**Figure 1.** (A) Prenatal ultrasonography at 33+2 weeks’ gestation revealed a cystic complex with solid components and calcification, measuring 11.9 × 9.5 cm. (B) Detailed ultrasound examination of the vascularization of the solid component showed minimal vascular shunting within the tumor (resistance index, 1.47; pulsatility index, 2.65). (C) Postnatal magnetic resonance imaging revealed an external sacral complex with multi-cystic and solid components, with no pelvic or intra-abdominal lesions (c = cyst; s = solid component; sc = sacrococcygeum).

**Figure 2.** Dizygotic twins were delivered by cesarean section at 34 weeks of gestation: (A) twin A was normal in appearance without congenital anomalies, and twin B presented with a large sacrococcygeal mass without additional congenital anomalies; (B) the tumor measured 15 × 10 × 9.5 cm and the anus was displaced anteriorly (arrow).
surgical margin was free of tumor and biopsy of coccygeal bone was negative for malignancy. Surgical removal of the tumor was complete and twin B did well postoperatively. Both twins were closely followed-up.

Discussion

The exact etiology of SCT is unknown [2]. Ronald et al first described SCT diagnosed by prenatal ultrasonography in a male twin in 1985 [5]. The lesion was not of sufficient size for detection during the second trimester, but was clearly visualized at 30 weeks of gestation. The sick twin underwent complete resection of the tumor and pathologic examination revealed three germ cell layers with abundant immature neuroectodermal tissue. Postoperatively, the male infant progressed well [5]. The female twin B presented in this study is similar to that reported by Ronald et al [5].

Malignancy of SCT is very rare in the neonate (< 1 month of age). Surgery and chemotherapy are the suggested treatments [2,6–8]. Altman et al found that when the tumor is discovered before the age of 1 month, the risk of malignancy is only about 5% [2]. Between 1 and 12 months of age, the malignancy rate is about 60%. In most patients, surgery is the principal therapy for SCT and should be performed in toto within the first week after delivery. The long-term survival rate of neonates after surgery for benign tumors is 92–95%, and that of malignant tumors is about 20% [6,9]. About 23% of immature teratomas recur after surgical resection (as determined by the detection of an elevated serum AFP). Multi-agent chemotherapy may be beneficial for treatment of tumor recurrence [6,7]. Hedrick et al described a patient with mixed mature/immature SCT and subsequent sacral recurrence who had an excellent response after treatment with four cycles of etoposide, carboplatin and bleomycin [8]. Serum AFP levels, computed tomography (CT) or MRI may be helpful in detecting occult recurrences or metastases of malignant tumors [7,8].

Altman et al defined four types of SCT according to the location of the tumor [2]. Our patient had an Altman type I tumor, i.e. predominantly external on the buttocks with a minimal presacral component. Altman type I tumors account for most SCTs (47%). In large clinical series, more than 90% of type I SCTs are successfully removed at the initial operation. All SCTs should be regarded as having malignant potential. About 5% of patients with SCT have metastatic disease at the time of diagnosis [2,6].

Large SCTs cause prenatal death due to high-output cardiac failure resulting from vascular stealing; they need immediate intervention before childbirth. Prenatal intervention in selected fetuses with SCT includes open fetal surgery or radiofrequency ablation in utero to prevent fetal death secondary to vascular steal in a rapidly growing solid tumor [8,10]. Chen and Shih reported that three-dimensional color power angiography allows the visualization of blood flow between the SCT and fetal circulation, and facilitates the targeted ablation of the feeding tumor vessels to interrupt blood flow [11]. Hedrick et al reported that surgical debulking of the tumor in utero has proven successful in carefully selected patients based on ultrasonographic and echocardiographic evidence of impending heart failure and favorable anatomy [8]. Large SCT did not cause a problem for this twin B fetus. This may have been because the tumor was mainly composed of cystic tissue and had no profuse vascularity within it. Vascular steal was not a cause of significant effects in high cardiac output.

AFP levels in our delivered twins were markedly elevated and demanded close follow-up as AFP is an indicator for malignancy. In newborns, the use of AFP in this manner is somewhat complicated because AFP is normally high in neonates [12]. Wu et al reported a mean AFP level in healthy newborns of 48,000 ng/mL, and that AFP in neonates 8 months of age had returned to the normal value of 8.5 ng/mL [12].

In this twin B with SCT, adequate follow-up of serial AFP levels will be performed at the age of 8 months after delivery. The normal twin will also be closely followed-up until AFP returns to normal. Elevated AFP levels indicate recurrence or metastases, so twin B with SCT will receive complete investigative oncologic studies.

References