症例報告

SUCCESSFULLY COMPLETE EXCISION OF A HUGE SACROCOCCYGEAL TERATOMA: A CASE REPORT

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Abstract : A female infant was born with a huge sacrococcygeal teratoma (SCT) at 36 weeks of gestation with a weight of 4546g (combined weight of the infant and tumor). The patient's mother developed polyhydramnios and cesarean section was performed at 36 weeks of gestation. MRI showed Altman classification type II SCT. We performed abdominoperineal resection at 3 days after birth. The mass was completely excised, and the size and the weight of the mass were 18×12×15cm and 1100g (Weight of SCT against body weight; 393 g/kg), respectively. The pathological findings revealed a grade II immature teratoma with no malignant elements such as yolk sac tumor. The patient was discharged at 45 days after birth with fecal continence, but neurogenic bladder dysfunction. At 9 months after surgery, the patient was doing well with no evidence of tumor recurrence, and AFP levels had returned to normal values.

Key words : sacrococcygeal teratoma, polyhydramnios, alpha-fetoprotein

In the management of sacrococcygeal teratomas diagnosed in utero, it is difficult to decide the timing of operation in association with fetal hydrops or malignant potential of the tumor. Several reports describe that polyhydramnios or fetal cardiomegaly may be an early sign of fetal distress, and hydrops often predisposes preterm labor, and solid and primary mass indicate a poor prognostic factor [1-3].

CASE REPORT

In August 2005, a 24-year- old woman was referred to our hospital for evaluation of a fetal tumor. At 27 weeks of gestation, fetal ultrasonography showed a tumor in the hip. Afterwards the woman was followed in an outpatient clinic until 31 weeks of gestation, when magnetic resonance imaging (MRI) revealed an enlarged sacrococcygeal tumor (Fig. 1a). After the patient's mother was admitted, polyhydramnios was noticed and cesarean section was performed at 36 weeks of gestation. A female infant was born with a huge sacrococcygeal teratoma (SCT) at 36 weeks of gestation with a weight of 4546g (combined weight of the infant and tumor) (Figs. 1b, 2a). MRI showed Altman classification type II SCT presenting with a mixed pattern, of which the majority was a cystic mass (Figs. 1c, 1d). We performed abdominoperineal resection with the patient in the supine position after

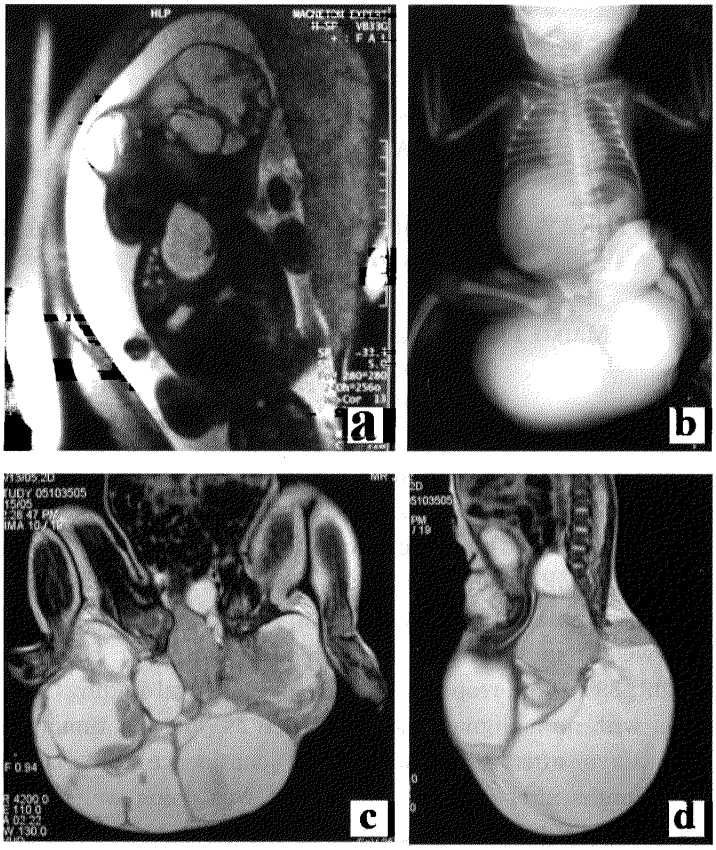

Fig. 1. Prenatal and postnatal radiography of an infant.
a:, Magnetic resonance imaging (MRI) at 31 weeks of gestation revealed an large sacrococcygeal tumor.
b: X-ray film at 36 weeks of gestation after cesarean section showed a huge sacrococcygeal tumor.
c, d: MRI showed Altman classification type II SCT presenting with a mixed pattern, of which the majority was a cystic mass (c: coronary section, d: sagital section).

devascularization with laparotomy at 3 days after birth. A transverse incision above the umbilicus was done and the peritoneal reflection was opened to the right of the sigmoid colon. A large cystic mass was confirmed in the presacral space (Fig. 3a). When the mass was compressed to the visceral side, the median sacral artery was noticed and then ligated (Fig. 3a). After devascularization, a partial resection of the tumor was done in a prone position with 4-cm distance from the anus to the excised skin margin (Figs. 2a, 3b, 3c). Afterwards the patient was placed in a supine position, via a sacral chevron incision, underwent a complete resection of the mass with completion coccygectomy (Figs. 3d, 3e). The middle sacral artery was also a feeding artery of SCT and ligated. The size and the weight of the mass were 18×12×15cm and 1100g (Weight of SCT against body weight; 393 g/kg), respectively (Fig. 3f). Estimated blood loss was neglible. The operating time was 10

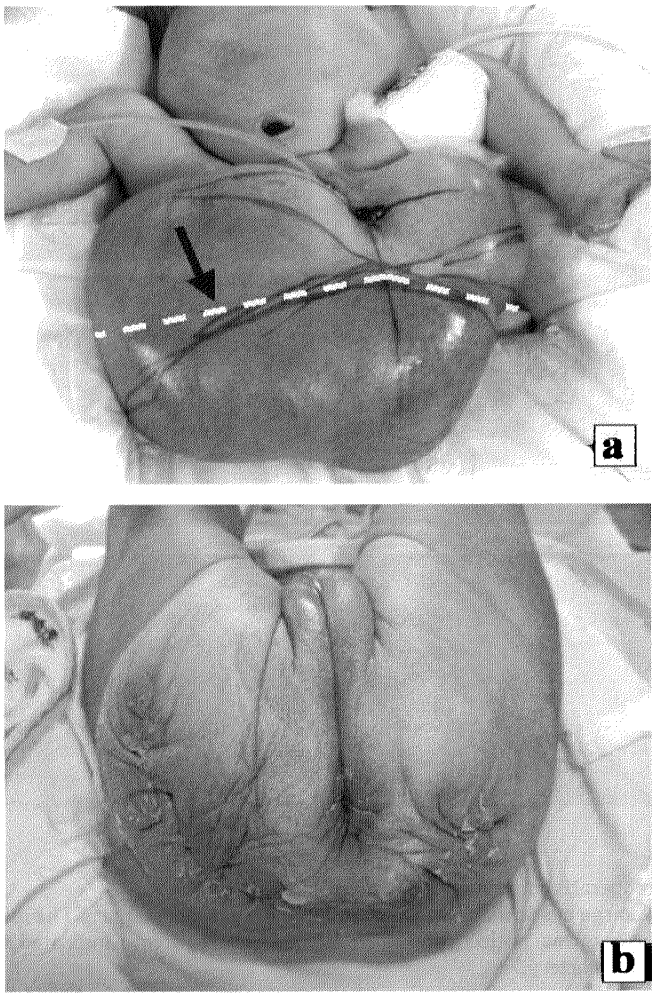

Fig. 2. Pre- and post-operative findings of an infant.
a: . The weight of an infant was 4546g with combined weight of the infant and tumor. A partial resection of the tumor in a prone position was done at 4-cm distance from the anus to the excised skin margin (black arrow).
b: In a supine position, via a sacral chevron incision, a complete resection of the mass was done. The reconstruction of the hip skin was cosmetically pleasing without any excess skin.

hours. The reconstruction of the hip skin was cosmetically pleasing without any excess skin (Fig. 2b). In the pathological findings, the solid parts of the tumor were found to be composed mainly of immature glial elements (Fig. 4a), associated with foci of neuroblastoma-like cells (Fig. 4b), immature mesenchymal cells and glandular epithelial cells (Fig. 4c). In the dilated cystic parts of the tumor, relatively mature skin tissues (Fig. 4d) and adipose tissues including lipoblast-like cells (Fig. 4e) were present. These pathological findings revealed a grade II immature teratoma with no malignant elements such as yolk sac

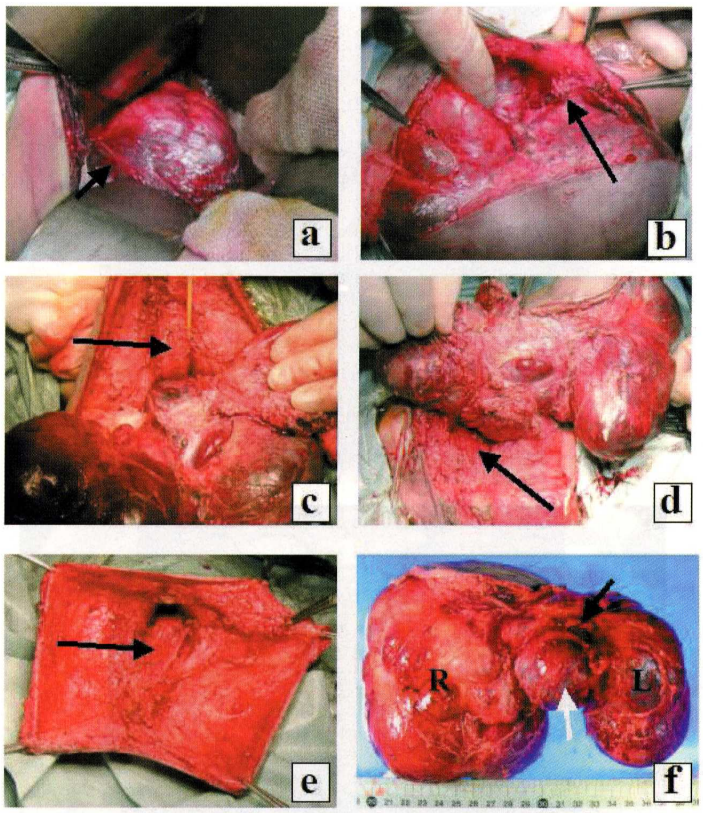

Fig. 3. Intraoperative findings and resected specimen.
a: A large cystic mass was confirmed in the presacral space. The median sacral artery was noticed and then ligated (Black arrow indicated the median sacral artery.)
b, c: A partial resection of the tumor was done in a prone position. To prevent an injury of the rectal muscle layer and levater muscles, a large nelaton tube was inserted into the rectum (Black arrow indicated rectal muscle layer).
d,e: Afterwards the patient was placed in a supine position, via a sacral chevron incision, underwent a complete resection of the mass including resection of the coccyx and ligation of the middle sacral artery by transfixing suture. (Black arrow indicated rectal muscle layer).
f: The size and the weight of the mass were 18×12×15cm and 1100g, respectively (R: right, L: left. White arrow indicated a large cystic mass in the presacral space. Black arrow indicated a resected coccyx.)

tumor. The patient was discharged at 45 days after birth with fecal continence, but neurogenic bladder dysfunction. The postoperative alpha-fetoprotein (AFP) levels decreased slowly (127347.0 ng/ml at birth to 73.6 ng/ml at 75 days after birth). At the last follow-up, 9 months after surgery, the patient was doing well with no evidence of tumor recurrence, and AFP levels had returned to normal values (9.1 ng/ml).

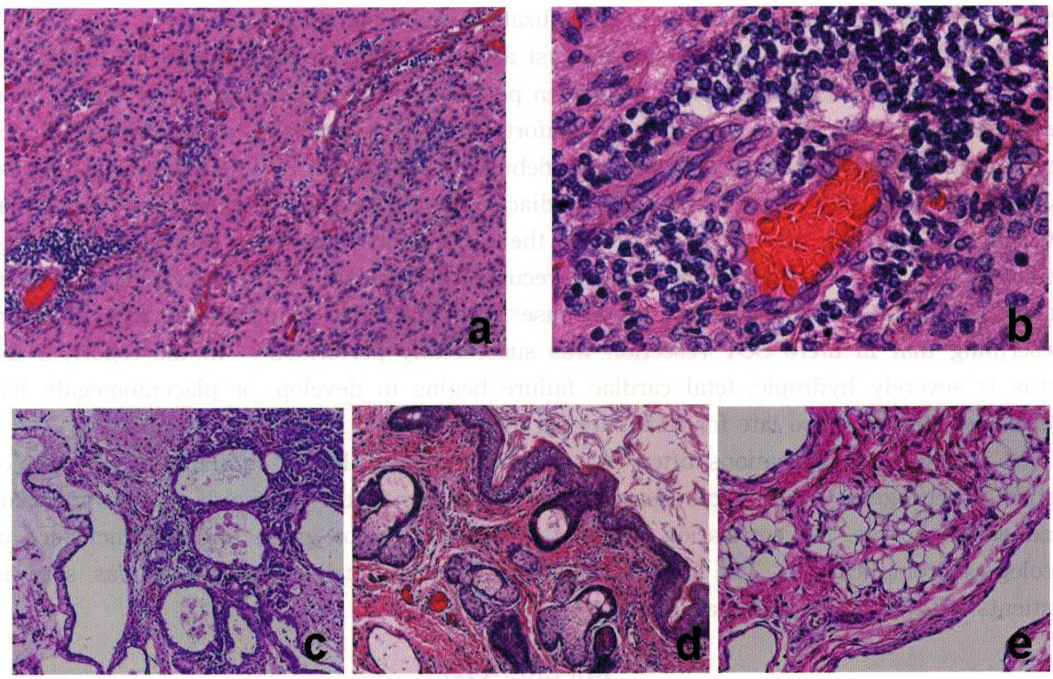

Fig. 4. Pathological findings (Hematoxylin-eosin staining)
a; The solid parts of the tumor were found to be composed mainly of immature glial element (×100).
b; Foci of neuroblastoma-like cells were present (×400).
c; Immature mesenchymal cells and glandular epithelial cells were also found (×200).
d, e; In the dilated cystic parts of the tumor, relatively mature skin tissues (d) and adipose tissues including lipoblast-like cells (e) were present (×200).
These pathological findings revealed a grade II immature teratoma with no malignant elements such as yolk sac tumor.

DISCUSSION

On prenatal follow-up of sacrococcygeal teratoma (SCT), the most valid predictors of poor outcome in fetuses with SCT are delivery prior to 34 weeks gestation, and the development of fetal hydrops with survival rates of 7 and 25%, respectively. On the other hand, the absence of hydrops and polyhydramnion, prenatal diagnosis after 30 week's gestation, tumor size <10cm at delivery, and delivery after 34 week's gestation are all associated with a survival rate of >80%[3]. The size of any SCT may increase considerably with gestational age. A mainly solid and highly vascularized tumor may lead to the development of fetal hydrops. In neonates and young infants, the presence of malignant elements and the size of the tumor are important prognostic factors. More commonly, microscopic malignant foci lead to recurrence of a tumor initially thought to be benign. Therefore, infants with SCT should be followed closely for at least 3 years based on serum alpha-fetoprotein and β-hCG levels[3]. In level normalization of AFP, it was described that

the mean time required for AFP level normalization was 9 months[4].

For SCT, fetal interventions, including cyst aspiration, amnioreduction and amnioinfusion except open fetal surgical resection have been performed. The indications for cyst aspiration and amnioreduction were maternal discomfort, preterm labor, and prevention of tumor rupture at delivery[2]. On open fetal surgery (debulking of external SCT; Altman classification type Ⅰ and Ⅱ) because of an increased cardiac output, placental thickness or pericardial effusion, no attempt was made to dissect the intrapelvic component of the tumor or to remove the coccyx. Postnatally, metastases, recurrence or failure to thrive occurred, but most cases are alive without evidence of disease[2]. On the other hand, there was a report describing that in utero SCT resection was successfully performed[5]. In the report, if the fetus is severely hydropic, fetal cardiac failure begins to develop, or placentomegaly has developed, then it is too late for fetal surgical intervention.

In urological complications after resection, neurogenic bladder is common. The major cause of neurogenic dysfunction seems to be related to surgical trauma of the pelvic plexus and sacral nerves. No correlation was noted between tumor grade and the incidence of urological complications. The highest incidence of urological complications was seen in patients with type IV (presacral) disease[6,7].

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