A case of a giant sacrococcygeal teratoma

H. Gozar¹, M. Ardelean², V. Giliga³, M. Turcu³, S. Bancu³

¹County Emergency Clinical Hospital Tg-Mureș
²University of Medicine Paracelsus Salzburg
³University of Medicine Tg-Mureș

Introduction

Sacrococcygeal teratomas (SCTs) are the most frequent solid congenital tumors in the fetus and newborn (1). They originate during embryonic development from totipotent cells (2) and are composed from one or more of the three germinal layers. SCTs originate from the primitive knot and are attached to the coccyx (3). These tumors contain multiple tissues foreign to the sites in which they arise. The tumors appear generally as disorganized islands of cells with cystic spaces. The tumor may contain skin, hair, teeth, cartilage, bones, fat, intestinal or neural tissue. The newborn’s teratoma are benign in up to 70% of cases, composed from mature cells (4). However some SCTs contain immature cells, but if they are completely excised they do not recur.

The incidence reported is of approximately 35,000-40,000 live births, with a male: female ratio of about 1:3.
As associated anomalies with sacrococcygeal teratomas there are described anorectal malformations, Hirschprung’s disease, congenital dislocation of the hip, nervous system lesions, urogenital anomalies (hypospadias, vesicouretral reflux, vaginal or uterine duplications) (4).

SCTs are commonly diagnosed in the prenatal period, during a routine ultrasound (5,6). Complications may occur in utero: polyhydramnios and tumor hemorrhage, which can lead to anemia and nonimmune hydrops fetalis. Postpartum morbidity associated with SCT is attributable to associated congenital anomalies, mass effect of the tumor, recurrence, intraoperative and postoperative complications.

Fetus with sacrococcygeal teratomas detected antenatally have three times mortality rate compared with postnatally diagnosed neonates (7,8). The causes of death are tumor rupture, cardiovascular failure, metabolic problems (9).

The treatment of teratomas is surgical. Complete excision of the tumor and the coccyx is very important. Type III also has external and internal components, but the internal portion extends into the abdominal area. These types, II and III are also evident at birth, but the resection may be more difficult requiring access both from the back of the baby and from the front. Type III will spread in about 20% of cases.

Case report

A neonate girl, born in another hospital, was presented at 16 days with a giant type III SCT (Fig. 1). Mother was 22 years old and it was her first pregnancy. The pregnancy was normal, without any events, the mother had five examinations to an obstetrician and he did four ultrasounds. The tumor was diagnosed at 36 weeks of gestation and the cesarian section was performed at 40 weeks. The baby proved to be in a good condition, with a mass on the sacrococcygeal region. She had 4040 g in weight and 50 cm length. The cranial perimeter was 35 cm. The tumor had 15 cm in diameter and it prolonged more to the left hip. The baby had also an arterio-venous malformation on the left forearm. There were no other problems. The neonate could eat and had stool normally.

The hematological and biochemical investigations were within normal limits. The CT scans revealed a great tumor with disorganized tissues, with calcifications and cystic spaces (Fig. 2). It invaded the retroperitoneum, compressed and dislocated all the abdominal organs.

Complete surgical excision was planned. The patient was brought to the operating theater, anesthetized and placed in the prone position. The operation started with a V-shaped skin incision with the tip on the sacrum. A tube was put into the rectum in order to identify that organ during the preparation of the tumor. En bloc excision was performed dissecting the tumor from gluteal muscles, levator ani muscles, rectum and abdominal organs (Fig. 3). The coccyx was transected at the junction of the sacrum and the middle sacral artery was ligated. Levator and gluteal muscles were reconstructed. The excess of skin was excised, and it was sutured carefully.
The tumor had 865 g and 15 x 17 x 12 cm (Fig. 4). The histological examination revealed a benign teratoma with no traces of malignancy. The postoperative recovery was uneventful. The child was discharged 5 days after the operation (Fig. 5). At 6 months follow-up, there are not pathological findings, except the scars over the buttocks.

Discussion

The word teratoma comes from classical Greek `teratos`, means a monster and during times was assigned to demons, sexual misconduct and abnormal fertilization and `-om` means tumor (10).

Prenatal diagnosis of sacrococcygeal teratoma is very important, because this tumor is associated with polyhydramnios, hydrops or placentomegaly. Repeated ultrasounds may show how large became the tumor (11). A large tumor may leads to tumor rupture during the vaginal delivery, with an important hemorrhage: in such cases the cesarian section is recommended (3). In our patient prenatal ultrasound diagnosed the big tumor in the 36th week of gestation. Consequently the cesarian section was performed, but in a hospital without a department of neonatology and pediatric surgery. Fortunately in this case the newborn and his mother had no complications, but the risk was high.

Complete excision of the tumor must be done, through a chevron shape incision. The dissection must be carefully, in order to preserve the muscles and rectum. Hemorrhage from middle sacral vessels and hypogastric arteries, injuries of the abdominal organs are the most common complications. En bloc excision of the tumor and the coccyx must be accomplished.

The operation was planed and performed carefully. The tumor was excised along with coccyx in toto. The failure to excise the coccyx, results in a recurrence rate of about 35- 40% (4). There were no abdominal organs injuries. The reconstruction of the muscles and skin led to a good functional and cosmetic result.

Alfa-fetoprotein levels should be determined immediately postoperatively and on discharge.

The tumor in the case presented here was a type III sacrococcygeal teratoma. Types I- III sacrococcygeal teratomas are seen as a variable mass at birth. According to Altman (12), type I is predominantly external with a small presacral component; type II has both a significant external and pelvic extension; type III, tumor is visible externally, but the predominant mass is pelvic and intraabdominal. Type IV teratomas are completely intrapelvic. If there are not diagnosed prenatally, they are difficult to diagnose and treat and more likely to be malignant. Patients with type IV teratomas tend to present later than those whose teratomas have an external component.

Every unexpected congenital abnormality represents a psychological problem for the parents, but such a huge tumor, which is a quarter of the newborn is a real shock. Despite these problems, the parents sustained all the time their baby.

The main differential diagnosis in this case is the meningocele. But the meningocele is placed cranial from the sacrum and one can feel the bulging of the fontanelle during the pressure of the tumor (4).

Follow-up should continue for at least 5 years: at monthly intervals for 3 month, at 3- monthly interval for a year, and thereafter once per year. Rectal examination and level of alfa-fetoprotein will be determined at each follow-up.

We followed-up the patient according to this protocol. There are no signs of recurrence after 6 months postoperatively. The child has no fecal or urinary problems. The cosmetic result of the operation is very good, one can barely see now the scars of the surgery.

References


