

Antenatal diagnosis of sacrococcygeal teratoma – two different case reports

Diagnostyka prenatalna potworniaka regionu krzyżowo-guzicznego – dwa przypadki o różnym przebiegu

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Abstract

Sacrococcygeal teratoma (SCT) is the most common fetal tumour. It may be readily diagnosed by two-dimensional ultrasonography in the early second trimester. Three-dimensional ultrasonography is recommended as a supplemental examination to obtain further detailed information for multidisciplinary management of SCT.

In the following article we present two cases of sacrococcygeal teratomas which exhibited two different courses.

In the first case, sacrococcygeal teratoma was diagnosed in the early second trimester. The condition of the fetus worsened during the follow-up and the parents opted for the termination of the pregnancy.

The other one presented at a later gestational age. The baby was born alive, immediately operated upon and did well until the age of 1, when we checked her condition for the last time.

Key words: **sacrococcygeal region / teratoma / prenatal diagnosis / fetus /**

Streszczenie

Potworniak regionu krzyżowo-guzicznego (SCT) jest najczęstszym guzem występującym u płodu. Może być łatwo zdiagnozowany za pomocą ultrasonografii dwuwymiarowej we wczesnym drugim trymestrze. Rekomenduje się wykonanie ultrasonografii trójwymiarowej jako uzupełniającego badania dostarczającego szczegółowych informacji ważnych dla wielodyscyplinarnego postępowania w przypadku SCT.

W bieżącym artykule przedstawiamy dwa przypadki potworniaka krzyżowo-guzicznego o różnym przebiegu.

W pierwszym przypadku guz został rozpoznany we wczesnym drugim trymestrze ciąży. Stan płodu ulegał stopniowemu pogorszeniu wraz z czasem trwania ciąży, dlatego rodzice zdecydowali się na zakończenie ciąży.

W drugim przypadku guz ujawnił się w późniejszym okresie ciąży. Noworodek urodził się żywy, został niezwłocznie zoperowany i rozwija się prawidłowo, co potwierdzono w trakcie wizyty kontrolnej po skończeniu przez dziecko pierwszego roku życia.

Słowa kluczowe: **region krzyżowo-guziczny / potworniak / diagnostyka prenatalna / płód /**

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Otrzymano: 30.12.2008
Zaakceptowano do druku: 20.04.2009

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Introduction

Sacrococcygeal teratoma (SCT) is the most common fetal tumour. It is a germ cell tumour, located in the presacral region, which arises from the pluripotent cells within Hensen's node [1]. These cells have the capability of differentiating into ectoderm, endoderm and mesoderm [2]. The incidence of SCT has been reported to be 1/35000- 1/40000 [3, 4]. There is gender preponderance, with females being 4 times more commonly affected than males [5-7]. Inheritance is generally sporadic, but familial forms have also been reported.

Case 1

A thirty-seven-year-old multipara patient was referred to our clinic at the 17th gestational week for amniocentesis due to advanced maternal age. Medical history was unremarkable except for the parents being consanguineous. Ultrasound examination revealed a 2cm cystic mass located in the sacral region with a solid intra-abdominal component occupying most of the abdomen. (Figure 1).

Colour Doppler examination revealed minimal vascularity of the tumour. The bony architecture of the spine and the intracranial structures were normal, neural tube defect was ruled out and the diagnosis of sacrococcygeal teratoma was made. Genetic sonogram was otherwise normal. The karyotype analysis was normal as well. The patient was recommended for a follow-up. At the 20th gestational week the sacrococcygeal mass measured 5cm and the intra-abdominal component of the teratoma enlarged and displaced the bladder to the left. (Figure 2).

Vascularity increased when comparing to the earlier ultrasound findings. Three-dimensional ultrasound images revealed a midaxial mass located in the sacral region. (Figure 3).

Fetal echocardiography was unremarkable for all structures. No signs of cardiac insufficiency and dilatation of the chambers were observed. The parents were informed about the follow-up, need for surgery and prognosis. They chose to terminate the pregnancy. Autopsy confirmed the ultrasound findings.

Case 2

A twenty-six year old primigravid patient was referred to our clinic at the 30th gestational week with a fetal mass. The patient had had no antenatal care before. On admission, the ultrasound showed 8cm, predominantly external, SCT with profound vascularity which was solid and cystic. Fetal echocardiography was normal. The amount of the amniotic fluid increased. Fetal blood sampling revealed a normal karyotype. Two days after the fetal blood sampling the patient was admitted to the antenatal clinic with premature rupture of the membranes. The mass enlarged when compared to the last examination and haemorrhage into the tumour and amnion was observed. A spontaneous rupture was suspected. A 2000 gram infant who had ruptured SCT was delivered by an emergent caesarean section. The infant underwent a surgery on the first postpartum day and SCT was removed together with the coccyx. During the surgery, the baby developed sudden cardiac arrest due to acute bleeding from teratoma.



Figure 1. Sacrococcygeal teratoma and its abdominal component first detected at the 17th week of gestation.



Figure 2. The external and the internal component of the sacrococcygeal teratoma at the 20th week of gestation.



Figure 3. The three dimensional presentation of sacrococcygeal teratoma in utero (3a) and after the termination (3b).

She was successfully resuscitated. The operation was withheld and the skin was closed two days later when the condition of the baby improved and she regained her strength.

Discussion

SCT may be classified as benign (mature), malignant or immature [8]. Mature teratomas constitute 90% of all SCT cases and they are usually solid, whereas immature teratomas, which have a higher malignant potential, are usually cystic. Associated malformations are present in 10-40% of the cases. The diagnosis of SCT is relatively easy with the use of a two-dimensional ultrasound. Three-dimensional ultrasound offers a chance to evaluate the tumour at various angles, with more visible sacral area and the spinal imaging. It is also possible to assess the tumour volume before the delivery. Based on current findings, three-dimensional ultrasound is recommended as a supplemental examination to give further detailed information on the multidisciplinary management of congenital tumours and masses [9]. Fetal MRI has been reported to be superior to sonography in assessing the intrapelvic and intraspinal extent of tumours and compression of pelvic organs by the tumour [10].

External tumours are easily recognized at birth as axial tumours located in the sacral region of the newborn. Tumours which are only located in the abdomen are not obvious at birth. Thus, they are usually diagnosed at a later time. The delay in proper diagnosis is often associated with the poor prognosis because malignant transformation is common.

Altman et al. classified the staging of SCT based on the amount of external and internal pelvic component in the American Academy of Pediatrics' surgical section (APPSS) survey [5]. Type I is predominantly external, with minimal intra-abdominal component, type II is external, with significant intrapelvic extension, type III is apparent externally but predominantly a pelvic mass extending into the abdomen is present and type IV is intra-abdominal, with no external presentation.

The classification by Altman is descriptive rather than prognostic. Benachi et al. classified fetal SCT into 3 prognostic groups: In group A, the diameter of SCT is less than 10cm and there is no or mild vascularity and slow growth. In group B, the diameter of the tumour is 10cm or greater and a pronounced vascularity, as well as fast growth, is present. Group C includes SCT with the diameter of 10cm or greater, but there is no or mild vascularity. They are not always fast-growing tumours and are predominantly cystic. The prognosis is good for groups A and C. However, for group B the prognosis is poor and maternal morbidity is high. Medical termination should be offered to this group. If this is not possible, weekly scan in a tertiary centre by a multidisciplinary team is necessary [11].

Vaginal delivery is possible for tumours smaller than 10cm. Prenatal cyst decompression may enable safe vaginal delivery or caesarean section through a transverse incision in case of cystic tumours bigger than 10cm. When the tumour is 10cm or larger, caesarean section through a vertical incision is recommended [12].

Mortality of prenatally diagnosed SCT is about 50%. It is attributed to complications such as malignant invasion, haemorrhage into the tumour, obstruction of the umbilical flow, high output cardiac failure, hydrops fetalis and bladder outlet obstruction [13, 14]. A subset of fetuses with large, rapidly growing, SCT may develop arteriovenous shunting through the tumour with evolution of high output cardiac failure, followed by placentomegaly, polyhydramnios, hydrops and subsequent fetal demise. Hydrops may also adversely affect maternal health and cause maternal mirror syndrome [15].

SCT may compress the bladder neck or obstruct the gastrointestinal system. While obstruction of the bladder neck causes oligohydramnios and pulmonary hypoplasia, the latter results in polihydramnios and preterm labour. To treat obstructive uropathy and preterm labour, a cyst-amniotic shunt may be placed or needle drainage can be performed [16, 17]. The disadvantage of needle drainage is that it requires multiple procedures and is often associated with the development of an infection [18].

SCT may be resected in utero. The aim of antenatal intervention is to halt or reverse the in utero physiological changes that are occurring. The first successful open fetal surgery to debulk the tumour was performed by Adzick et al. in 1997 on a 25-gestational-week-old fetus. Since then, a number of fetuses have been operated on [17]. As far as fetal surgery is concerned, premature rupture of the membranes and preterm labour, which are more commonly seen when fetal surgery is performed before the 23rd gestational week, remain the main obstacle [19]. High-output cardiac failure or maternal mirror syndrome are contraindications to fetal surgery. In such cases radiofrequency ablation is offered to patients as an alternative to fetal surgery. Haemorrhage into the tumour, intrauterine death, premature delivery, and complications of tissue necrosis have been reported with this method [16, 20].

Prenatal diagnosis improves the neonatal outcome by allowing an appropriate choice of mode, place and timing of the delivery. Obstetric management depends on the size, volume, vascularity, type and grading of the tumour, involvement of the adjacent organs and blood vessels. Close follow-up with repeated sonography is necessary to monitor the tumour size, extension into adjacent structures, tumour vascularity and evidence of cardiac failure. Serial measurements of Doppler blood flow for the early detection of cardiac failure is important [15]. Sudden death without any evidence of high output cardiac failure may also occur. Termination of the pregnancy may be considered in cases with poor prognostic signs such as cardiac failure, placentomegaly, marked hydrops or associated fetal anomalies [21].

The majorities of sacrococcygeal teratomas are benign and resectable after birth without significant morbidity, and are rarely associated with other malformations. Therefore, there is a chance for the child to lead a normal life without disability.

Acknowledgements

Written consent was obtained from the patients to submit these case reports.

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