EARLY DIAGNOSIS OF FETAL SACROCOCCYGEAL TERATOMA: A CASE REPORT

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Sacrococcygeal teratoma is a rare fetal neoplasm with an incidence of 1 in 40,000 births. Antenatal diagnosis is usually made after 22 weeks of gestation. Fetuses with this malformation are at risk of significant perinatal morbidity and mortality. Malignant components, coexisting with life-threatening anomalies, and chromosomal abnormalities are rare. Postulated causes of perinatal death include hydrops, dystocia, tumor rupture, preterm labor secondary to polyhydramnios, and anemia due either to hemorrhage or hemolysis within the tumor. Herein, we present a case of fetal sacrococcygeal teratoma diagnosed as early as 17 weeks of gestation.

Key Words: fetus, sonography, sacrococcygeal teratoma
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Sacrococcygeal teratoma is a rare congenital neoplasm in the fetus, occurring in 1 in 40,000 infants. These teratomas are derived from pluripotent cells in Hensen's node of the primitive streak and contain components arising from all three layers. Prenatal diagnosis by ultrasound may reveal a mass of either cystic, solid, or mixed appearance in the sacral area protruding through the perineum. Altman et al classified sacrococcygeal teratomas into four types according to their location [1]. Type 1 is the most common and is predominantly external, protruding from the perineal region with a minimal pre-sacral component. Type 2 appears externally with a significant intra-pelvic portion. Type 3 is usually visible and extends into the abdominal cavity. Type 4 is completely within the pre-sacral area and has no external component. This classification depends on the relationship between extra- and intra-pelvic parts and is considered descriptive with no prognostic value. Approximately 75% of affected infants are female. Most fetal sacrococcygeal teratomas are diagnosed between 22 and 34 weeks of gestation by prenatal sonography [2]. We present the case of a male fetus diagnosed at 17 weeks of gestation.

CASE PRESENTATION

A 28-year-old woman (gravida 4, para 2) was referred to our hospital at 17 weeks of gestation with the chief problem of a fetal sacral mass. There was no contributory factor in her obstetric or other medical history. Sonographically, the tumor mass was 3.5 x 4.0 cm with a complex cystic structure (Figure 1). No polyhydramnios, placentomegaly, abdominal wall edema, or pericardial effusion was found. After detailed discussion with the patient and her family, termination of pregnancy was accomplished through vaginal delivery by induction. A male infant weighing 250 g was delivered with Apgar scores of 0 and 0 at 1 and 5 minutes (Figure 2).

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The clinical course was smooth. Pathologic study of the tumor revealed an immature teratoma primarily composed of neuroectodermal elements, i.e. neuroepithelial rosettes.

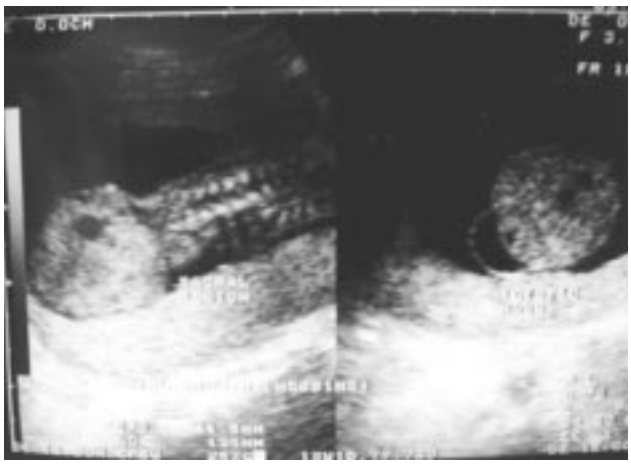

Figure 1. *A protruding tumor in the sacral region.*

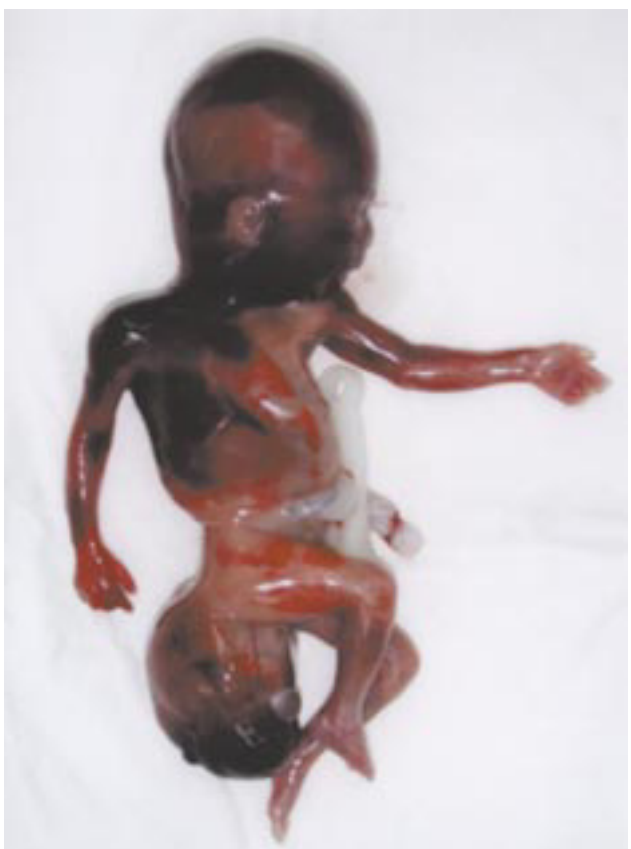

Figure 2. *Male infant with sacrococcygeal teratoma at 17 weeks of gestation.*

Discussion

The present case showed a complex cystic tumor in the sacral region of a male fetus which was diagnosed as a Type 1 sacrococcygeal teratoma. In general, a sacrococcygeal teratoma appears on the sonogram as a large mass attached to the fetal rump, projecting from the coccyx with polyhydramnios. The components may be purely solid, cystic, or a complex of both. The purely cystic form may be misdiagnosed as a myelomeningocele. The posterior spinal elements are preserved in sacrococcygeal teratoma, which distinguish it from myelomeningocele.

Sacrococcygeal teratomas are usually 7 to 8 cm in size. However, a sacrococcygeal teratoma of more than 25 cm has been reported [3]. Clinical features combined with hydrops and/or placentomegaly have great predictive value for prognosis. There were no such findings in the present case. Adjuvant imaging techniques such as magnetic resonance imaging (MRI) may be used to assess prognosis. However, although MRI is helpful for accurately defining the intra-pelvic extent of the sacrococcygeal teratoma, it cannot provide details of the extra-pelvic tumor, including the vascularity and differentiation of the tumor type. Ultra-fast T2 weighted imaging enables clear visualization of morphologic details of the fetus without motion artifacts [4].

Up to 18% of cases in a neonatal series have other abnormalities, for example, spina bifida or sacral agenesis [1]. However, karyotype study is usually normal. The differential diagnosis of a large mass projecting from the rump of a fetus includes lipoma, hemangioma, rhabdomyoma, sarcoma, dorsal midline hamartoma, and, if cystic, meningomyelocele. In a fetus with sacrococcygeal teratoma, obstetric complications include polyhydramnios, pre-eclampsia, hydrops, and preterm delivery. Among these, polyhydramnios can develop suddenly and contribute to the possibility of preterm labor. Placentomegaly can also be present with maternal pre-eclampsia. If sacrococcygeal teratoma causes venous compression, high-output cardiac failure and hydrops will result in a very poor prognosis for the fetus. However, if hydrops develops remote from term, fetal surgery may be an option for management. Langer et al reported a case of fetal surgery for sacrococcygeal teratoma performed at 21 weeks' gestation after development of hydrops, placentomegaly, and preterm labor [5]. Tumor

resection caused reversal of the hydrops, but intractable preterm labor developed. Nakayama suggested that fetal surgery may be indicated in patients with hydrops before 28 weeks' gestation [6]. This probably provides the best balance between the risks of fetal surgery and fetal death from hydrops.

The predictors of a poor outcome for a fetus with sacrococcygeal teratoma include diagnosis before 20 weeks' gestation, delivery before 30 weeks' gestation, and hydrops. Low birth weight and/or Apgar score less than 7 have also been regarded as predictors of poor prognosis [7]. In our present case, poor outcome for the fetus was expected. In a study of 29 cases, Kuhlmann et al reported an inverse correlation between tumor size and outcome [7].

Close sonographic (two- and three-dimensional) observation is the first step in the management of fetal sacrococcygeal teratoma, to monitor fetal growth and changes in tumor size and to assess the development of hydrops. In cases of symptomatic polyhydramnios and to prevent preterm delivery, volume reduction by amniocentesis and treatment with tocolysis are probably useful.

Barring the development of hydrops or maternal complications, delayed termination of pregnancy is suggested to allow fetal maturation. Gross et al recommended cesarean delivery in all fetuses with a sacrococcygeal teratoma of more than 5 cm [8]. An optimal uterine incision is important to avoid fetal injury and minimize the risk of tumor rupture. For this purpose, a vertical uterine incision can be used. In general, malignant changes in fetal sacrococcygeal teratomas are rare, including microscopic malignant foci, which may lead to the recurrence of a tumor that was initially thought to be benign. Therefore, it is strongly suggested that infants with sacrococcygeal teratoma should be followed closely for at least 3 years. We report this case to address not only its rarity, but also its diagnosis as early as 17 weeks of gestation.

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