Antenatal Diagnosis Of A Fetal Sacral Mass Containing A Pelvis And Limb Bones

Neil J. Murphy, MD, Colleen M. Murphy, MD, Frankie Lukey, RNC, ANP, Patrick J. Hanaway, MD, and Johannes H. Tan, MD

Sonographic examination during pregnancy has become increasingly common,1 though some authors have questioned the efficacy of sonography as a screening tool in low-risk women.2,3 Many family physicians use sonography in their own offices.4,5 Prenatal sonographic differentiation of a posterior fetal sacral mass can be difficult. The most common diagnoses include meningomyelocele and sacrococcygeal teratoma. If extrinsic fetal skeletal structures are seen, the diagnosis is more likely multiple gestation, acardiac monster, or conjoined twins.

We report a case in which a family physician and midlevel practitioner with level I sonography privileges in a rural hospital diagnosed a fetal sacral mass with four extrinsic limbs and a pelvis.

Case Report
A 33-year-old woman, gravida 5, para 4, at 10 weeks’ gestation sought initial prenatal care from a community health aide in her rural town of 1000 inhabitants. Her obstetric history was notable for four spontaneous vaginal deliveries. Her last delivery resulted in a 3269-g male infant, who died at 2 days of life as a result of total anomalous pulmonary vasculature. The patient had no other personal or family history of congenital anomalies.

The patient smoked one pack of cigarettes per day. She drank one to two drinks of alcohol per day until 10 weeks’ gestation but had no history of acetazolamide or other drug use.6 Her medical history was unremarkable. At 10 weeks’ gestation she had an Escherichia coli urinary tract infection that was treated with nitrofurantoin. Fetal heart tones were first documented at 16 weeks’ gestation by Doppler monitoring. Prenatal screening was unremarkable. A 1-hour glucose challenge test resulted in a glucose level of 132 mg/dL.

On routine examination the community health aide found the fundal height to be 23 cm at 24 weeks, but no heart tones were found by Doppler. The patient was referred to the 50-bed regional center 150 miles away in a town of 5000 inhabitants. A sonogram performed by a midlevel practitioner and family physician confirmed lack of cardiac motion. The slightly collapsed calvaria was compressed against the thorax. There was a 6.3- X 5.3- X 9.0-cm solid posterior mass with several hypoechoic areas or cysts. The mass had four linear calcifications consistent with nonaligned bones plus a curvilinear calcified structure consistent with a pelvis (Figure 1). The fetal vertebral column was intact, but visualization of the sacral area was difficult. There was no definite plane separating the mass from the lower fetal vertebrae. The biparietal diameter was 45 mm, and femur length on the main fetal body was 33 mm, consistent with 19.5 and 20 weeks’ gestation, respectively. There was no evidence of fetal hydrops. The amniotic fluid volume was normal. The placenta was posterior grade I.

The patient was referred to a tertiary care center 400 miles away in a town of 250,000 inhabitants. Sonographic examination confirmed the above findings with no additional relevant clinical information. Laminaria was used for cervical ripening, followed by a dinoprostone suppository (Prostin E2, Upjohn, Kalamazoo, MI) labor induction. The 350-g fetus was a normal phenotypic female with a midline sacrococcygeal mass consistent with an American Academy of Pediatrics Type I sacrococcygeal teratoma.7 No hydrops or gross anomalies were noted, except an imperforate fetal anus. The fetal heart was of normal size. The mass had two sets of two extrinsic legs and feet that measured 5 cm each. One set of legs had a full pelvis with no genitalia or anus. Microscopy revealed a mature teratoma with a variety of tissue types, e.g., fibrous, cartilage, and primitive neuroectoderm. The latter formed small cystic structures.

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From the Department of Obstetrics and Gynecology, Alaska Native Medical Center, Anchorage (NJM, CMM); Maternal Child Health, Yukon Kuskokwim Delta Regional Hospital, Bethel, (FL, PJH); and the Department of Pathology, Alaska Native Medical Center, Anchorage, (JHT), Alaska. Address reprint requests to Neil J. Murphy, MD, Department of Obstetrics and Gynecology, Alaska Native Medical Center, 255 Gambell, Anchorage, AK 99501.
Figure 1. Ultrasound of fetal sacral mass, right arrow; with linear calcified structure consistent with long bone, left arrow.

Discussion

There are no other antenatally diagnosed cases of sacrococcygeal teratoma with limb bones and a pelvis found on a MEDLINE computerized literature search. This case was diagnosed by a family physician and midlevel practitioner in a small rural hospital. The patient was appropriately referred to a tertiary care center.

Long bones have been described postnatally. Intrinsic calcification can be seen in 36 percent of sacrococcygeal teratomas. The prenatal diagnosis of sacrococcygeal teratoma is dependent on sonographic scanning, though amniotic alpha-fetoprotein and acetylcholinesterase levels can be elevated. Fetal karyotyping and echocardiography can help plan management.

Sacrococcygeal teratoma is the most common tumor in newborns, with an incidence of 1 in 40,000. Eighty percent of affected infants are female, and 85 percent of sacrococcygeal teratomas are solid. Rates of associated anomalies range from 5 to 25 percent. No pattern of anomalies has been described. In this case the relatively large size of the mass probably led to in utero death secondary to cardiovascular compromise.

Meningomyelocele is most often cystic, and the vertebral column is not intact. Our findings of a pelvis and four limbs suggested multiple gestation or poorly organized conjoined twins, e.g., pygopagus or ischiopagus, though no other organs were seen. An acardiac monster has well-developed lower limbs but would not appear affixed to the other twin. Other less common diagnoses for sacral masses include cystic hygroma, neurenteric cyst, sarcoma, chordoma, and ependymoma.

The rate of malignancy in sacrococcygeal teratoma can be 17 to 56 percent, and neither the size nor rate of growth predict malignancy. The imperforate anus raises the question of hereditary presacral teratoma. This autosomal dominant syndrome is associated with anorectal stenosis, as well as malignancy.

Though intrinsic calcifications are not unusual, we describe a sacrococcygeal teratoma with an extrinsic pelvis and long bones, which make the diagnosis more difficult. The antenatal recognition of sacrococcygeal teratoma could be especially important in view of the high rate of associated malignancy and as more aggressive prenatal management therapies develop, e.g., in utero surgery or induced abortion in the case of hereditary presacral teratoma.

Obstetric sonographic examination can be performed by family physicians with a high degree of accuracy and has been shown to be cost effective in the office setting. Congenital anomalies are infrequently encountered; hence, family physicians who perform obstetric sonography should be alert for anomalies and support their sonographic skills through continuing postgraduate medical education courses.

References

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