Brief Report

Dermoid Inclusion Cysts and Early Spinal Cord Tethering after Fetal Surgery for Myelomeningocele

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Myelomeningocele occurs in approximately 1 of every 2000 live births.[1] The prenatal incidence is higher, because many parents elect to terminate the pregnancy, once the diagnosis has been made.[2] Myelomeningocele often results in permanent impairment, including loss of sensorimotor function in the legs, hydrocephalus, incontinence of bladder and bowel, skeletal deformity, and type II Chiari malformation (cerebellar [vermis] and medullary herniation and inferior displacement below the level of the foramen magnum). Advances in ultrasonography and ultrafast fetal magnetic resonance imaging (MRI) have made it possible to diagnose myelomeningocele and other neural-tube defects early, but much remains to be learned about the optimal treatment of these disorders.

In recent years, in utero closure of myelomeningocele has been recommended to prevent damage to the spinal cord as a result of exposure to amniotic fluid through the open defect.[3-5] Approximately 220 in utero closures have been performed to date at four institutions. Although improvement in motor function has not been confirmed in children treated in utero, there has been a documented improvement in cerebellar herniation.[6] In addition, a comparison with historical controls suggests that children who have been treated in utero are less likely to require ventricular shunting for hydrocephalus, presumably because closure of the myelomeningocele sac provides a more normal pattern of spinal fluid circulation.[7,8] The amelioration of these sequelae may improve the quality of life for affected children.

However, these potential benefits must be weighed against the known postoperative complications of in utero repair, including induction of early labor, preterm delivery, infection, and death related to prematurity, as well as maternal morbidity. As more of these procedures are performed, other adverse sequelae may also become apparent. We describe three girls who had spinal cord tethering and large dermoid inclusion cysts in infancy. All three had undergone in utero closure of myelomeningocele at one institution.

CASE REPORTS

In Utero Closure

The preoperative evaluation and in utero treatment of the three patients have been described previously.[6] After written informed consent had been obtained from each mother, fetal MRI was performed with a single turbo spin–echo sequence on a 1.5-T unit, at 19 to 23 weeks of gestation. Surgical repair of the neural-tube defect was performed at 22 to 24 weeks of gestation. In each case, after general anesthesia had been administered to the mother, a hysterotomy was performed, and the lumbar area of the fetal spine was exposed. Primary dural closure was attempted with the use of standard techniques, and attempts were made to close the skin around the open defect. In each case, it was not possible to close the dura and skin, so a graft of acellular human dermis was used to cover the defect (AlloDerm, LifeCell). Amniotic fluid was replaced with warmed, sterile, lactated Ringer's solution. Tocolysis was initiated and maintained postoperatively to prevent preterm labor.

The mothers underwent MRI studies every three weeks postoperatively, in order to monitor fetal neural-tube development. The infants were delivered by elective cesarean section after lung maturity had been confirmed by the ratio of lecithin to sphingomyelin in the amniotic fluid at 30 to 36 weeks of gestation. Uncontrollable premature labor, which has precipitated early delivery in a number of other cases, did not occur in the three cases reported here. At birth, the three infants appeared to have nearly normal motor function in the legs, and no bladder dysfunction was detected clinically. Voiding cystometrograms were normal.

Subsequent Evaluation and Treatment

The three girls were followed postoperatively by local pediatric neurosurgeons. Loss of neurologic function occurred at an average age of 10 months (range, 9 to 11). Two of the girls had loss of motor function in both legs. In one of the two, decreased spontaneous movement of both legs was noted by the family, and radiographs of the lumbosacral spine showed progressive scoliosis. In the other infant, who had only slight plantar flexor weakness at birth, the motor function of the right leg was diminished and there was tenderness on palpation in the lumbosacral area. The third child presented with progressive bladder dysfunction. Whereas she had had normal voiding with a good stream and a normal cystometrogram after birth, a cystometrogram obtained at six months of age showed poor bladder compliance, with passive leakage at high intravesicular pressures. All three infants underwent MRI studies of the spine, without the administration of contrast material. The MRI scan from one of the infants is shown in Figure 1. Dermoid cysts and spinal cord tethering were present in all three infants.

Because of the findings on clinical examination and neuroimaging studies, surgical exploration was recommended for the three infants. The previous skin closures were reopened and extended in a cephalad direction. The dura was exposed and opened. Dura was adherent to the dermoid cysts. The capsules of the cysts were opened and

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