Congenital placental–cerebral adhesion: an unusual case of amniotic band sequence

Case report

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Amniotic band sequence is a disruption sequence having a broad spectrum of clinical manifestations ranging from partial amputations to major craniofacial and limb–body wall defects. Most reported cases of placental–cerebral adhesion pertain to patients with severe craniofacial defects who were either stillborn or who died a few hours after birth. The authors present a case of a male infant born with a placental–cerebral adhesion through a cranial defect. This adhesion was separated at birth, and duraplasty and primary scalp closure were performed. A detailed examination of the placenta revealed the presence of multiple amniotic bands. The case demonstrates that survival and normal early postnatal development are possible if the condition is treated promptly.

KEY WORDS • amniotic band sequence • placental adhesion • amniotic deformity • adhesion • mutilation complex • pediatric neurosurgery

A MNIOTIC band sequence is a condition in which aberrant amniotic bands, sheets, or strands adhere to, entwine, or constriict fetal parts, resulting in amputations or other deformities. Synonyms include amniotic deformity, adhesion, mutilation complex; amniotic band disruption complex; amniotic rupture sequence; amniotic adhesion malformation syndrome; and limb–body wall complex. It is a common sporadic condition having an estimated incidence of 1 in 1200 to 1 in 15,000 liveborn infants and 1 in 56 previable fetuses. We present an unusual case in which an infant boy was born with a placental–cerebral adhesion. Immediately afterwards, this adhesion was separated by duraplasty and primary scalp closure. A detailed examination of the placenta revealed the presence of amniotic bands.

Case Report

History. The mother of the patient was a 22-year-old gravida 1 woman who had experienced a single intrauterine pregnancy. She had a medical history remarkable for cervical intraepithelial neoplasia, which had been treated with a loop electrocautery excision procedure 2 years earlier.

There was no family history of congenital anomalies and no parental consanguinity. A routine fetal ultrasonography assessment performed at 19 weeks’ gestation revealed cranial abnormalities, including microcephaly, lemon sign, and mild ventriculomegaly. The ultrasonographic examination of the fetal head was deemed limited because the head remained in a suboptimal position throughout the study. A fetal MR imaging study performed at 22 weeks’ gestation confirmed the ultrasound findings and failed to demonstrate any additional significant abnormality. When the MR imaging study was reviewed retrospectively, the fetal head could be seen to be tapered toward the vertex and closely apposed to the placenta on all imaging sequences (Fig. 1). An amniocentesis revealed a normal 46, XY karyotype. No clear malformation syndrome was identified antenatally.

The mother presented to her local hospital with uterine contractions at 31 weeks’ gestation. She was transferred to a tertiary care hospital for treatment of threatened preterm labor. Nifedipine, penicillin G, and betamethasone were administered. Two days after admission, her contractions increased and were accompanied by vaginal bleeding and full dilation of the cervix. The baby was delivered by cesarean section without complication and required no resuscitation. At delivery, a scalp defect with placental adhesion was noted.
Examination. The baby weighed 1706 g and measured 41 cm in length, both normal for a baby born at 31 weeks gestation. His head circumference was below the fifth percentile for gestational age, measuring 25 cm, and his Apgar scores were 9 and 9 at 1 and 5 minutes, respectively.

A scalp defect over the vertex of the patient’s head measured approximately 12 cm². There was also a large skull defect and absence of most of the parietal bones bilaterally. A mass composed of the placenta, umbilical cord, and fetal membranes was adherent to the left parietal lobe (Fig. 2). The infant had mild facial asymmetry and hypoplasia of the left jaw. His right ear was overfolded, and vertical forehead creases were noted when he grimaced. His right hand had a transverse palmar crease and a short fifth finger with clinodactyly. His right foot had syndactyly of the second to fifth digits, and a strand of tissue on the sole between the fourth and fifth digits was interpreted as being a residual amniotic band. His left foot had partial second–third digit and fourth–fifth digit syndactyly and a transverse crease on the sole.

A computerized tomography scan obtained after delivery showed a large skull defect with absence of the osseous cranium over the entire vertex of the skull and multiple, interconnecting venous anomalies. Anomalous veins arising from the posterior fossa and from the deep venous system ascended and converged toward the vertex. These veins became confluent as they passed through the skull defect and joined a plexus of vessels in the placenta. No major abnormalities were evident in the cerebral hemispheres, ventricles, cerebellum, and vermis.

Operation. The patient was transferred to the operating room and general anesthesia was induced. The placenta was ligated with a silk tie approximately 3 cm from the surface of the scalp, and the bulk of the placental tissue was amputated. This maneuver allowed proper positioning and preparation before the definitive closure of the defect.

A skin incision was made in a coronal orientation starting at the edge of the scalp defect on the right side. The scalp was mobilized by subgaleal dissection, allowing the dura mater to be exposed circumferentially around the placental tissue. An SSS was not seen. The placenta adhered to the pia of the left parietal lobe through an opening in the dura that measured approximately 4 cm² (which was much smaller than the scalp defect). The placental tissue was dissected off the brain without complication. A duraplasty was performed using a polyester urethane graft, followed by primary closure of the defect.

Pathological Analysis. The amputated placenta was sub-

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Fig. 1. Coronal single-shot fast-spin echo MR image showing apposition of the fetal head to the placenta (P) above and a gap in the skull near the vertex (arrowheads), which was noted retrospectively following birth.

Fig. 2. Photographs showing the anatomical relationship between the placenta and the vertex of the head (left) and a closer view of the cranial defect and adherent placental tissue (right).
merged in water to assess it for amniotic bands. Multiple fragments of amniotic tissue measuring between 0.2 and 1.4 cm in length were noted at the base of the umbilical cord and at the periphery of the placental disc (Fig. 3).

A second specimen consisting of tissue from the site of the placental–cerebral adhesion was also obtained. Microscopically it consisted of fibrous tissue containing blood vessels of various sizes and marked acute and chronic inflammation. No neural or glial tissue was present.

**Postoperative Course.** A regimen of ampicillin and gentamicin, which was begun intraoperatively, was continued for 5 days. The baby recovered well and was transferred to his local hospital on postoperative Day 20.

One month postoperatively, a wound infection developed that required treatment with oral antibiotic agents. An MR imaging study performed when the patient was 7 weeks of age revealed an abnormally shaped head, a left parietal encephalomalacic cleft, and hypoplasia of the posterior corpus callosum (Fig. 4). With the exception of a thready midline anterior vein, the SSS was absent on flow-sensitive sequences (Fig. 5). At 2 months of age, the boy underwent surgical debridement and removal of the foreign duraplasty material. The granulation tissue and dural graft were removed without complication, and it was noted at the time of surgery that a fibrous layer had grown below the graft. The skin edges were closed, and the antibiotic regimen was continued to complete a 3-week course.

The boy was noted to have significant turricephaly, and when he was 3 months of age, a helmet orthosis was prescribed in an effort to correct the deformity of his head. He continued to grow, and his weight and length measurements remained roughly within the 50th percentile. His head circumference remained small, measuring less than the third percentile, but it continued to grow and its size remained parallel with this curve. His early development was appropriate for his corrected age, and he showed no evidence of seizures. He swallowed normally and muscle tone was equal in all limbs. He was last examined at 6 months of age and was well.

**Discussion**

Amniotic band sequence manifests as a broad spectrum of deformities and disruptions. Moerman, et al., divided these lesions into three types: 1) constrictive tissue bands, 2) amniotic adhesions, and 3) complex anomaly patterns, including the limb–body wall complex. Constrictive tissue bands are thin bands of amniotic epithelium that encircle one or more limbs and lead to loss of tissue distal to the entanglement. Amniotic adhesions are broad and involve the craniofacial area, leading to major defects such as severe craniofacial clefting and encephalocele. The limb–body wall complex consists of exencephaly or encephalocele with facial clefts, thoraco- and/or abdominoschisis, and limb defects.

As proposed by Torpin, the majority of cases of ABS are postulated to be caused by amnion rupture. The embryo forms within the amniotic cavity, which develops within the larger chorionic cavity. As the amnion grows, it presses against the chorion and obliterates the extraembryonic coelom. This cavity is obliterated by the 12th week of development. If this obliteration is not complete, the unsupported amnion can rupture, creating strands or sheets of amnion that entangle or adhere to the developing fetus. This proposed mechanism is most likely responsible for amputations, partial syndactyly, and constriction rings. It is not clear whether this process is responsible for more severe defects, such as craniofacial clefts and the limb–body wall complex. According to an alternative theory, first proposed by Streeter in 1930 and revised by Bamforth, a vascular germ disc disruption occurs earlier than 26 weeks' gesta-
Congenital placental–cerebral adhesion

Fig. 5. A postnatal MR venogram (coronal two-dimensional time-of-flight acquisition, three-dimensional reconstruction, right lateral projection). From the confluence of the straight and transverse sinuses (C) to the vertex, there is no evidence of an SSS. From the anterior edge of the skull defect (arrowhead), a thready segment of SSS extends anteriorly.

Congenital placental–cerebral adhesion is a rare occurrence that is associated with distal limb anomalies. In this case, the patient was either stillborn or died shortly after birth. It is probable that the severity of the associated craniofacial defects dictates the likelihood of survival from this type of ABS. In our case, the patient was microcephalic, and the venous drainage of the brain was abnormal with hypoplasia of the corpus callosum. The cerebrospinal fluid was otherwise well formed; thus, the patient’s early postnatal development was appropriate at 6 months of age. Despite preoperative imaging that indicated the presence of multiple vessels bridging the brain and placental tissue, little bleeding was encountered at surgery. Safe postnatal surgical separation of this unusual manifestation of ABS is possible in selected cases.

References


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