Case report

Medical Ultrasonography
2012, Vol. 14, no. 3, 254-256

Prenatal ultrasound diagnosis of open spinal dysraphism in the cervical verterbrae. Case report.

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Abstract

Spinal dysraphisms are lesions that can be identified at ultrasound screening examination from the second trimester of pregnancy, the majority being localized in the lumbosacral region. We present the case of prenatal ultrasound diagnosis of a 18 weeks fetus with cervical open dysraphism, a rare localization of open spinal dysraphisms. The alteration of cerebral normal anatomy was first identified; further examination showed the spinal defect with cervical location associated with a posterior cystic mass, the meningomyelocele.

Keywords: spinal dysraphism, Arnold-Chiari malformation, meningomyelocele

Introduction

The incidence for spina bifida is approximately 1 per 1,000 [1]. The term „spina bifida" is commonly used as a synonym of spinal dysraphism, although it properly refers to defective fusion of posterior spinal bony elements [2]. The terms „spina bifida aperta" or „cystica" were once used to refer to open spinal dysraphism and closed spinal dysraphism respectively [2]. Open spinal dysraphisms are located in the lumobsacral region in 90% of cases, in the thoracic region in 6-8%, and in the cervical vertebrae in 2-4% of cases [3]. Associated malformations include: Arnold-Chiari malformation, hydrocephalus, club feet, and, occasionally, hydronephrosis and heart defect [3]. At ultrasound in open spinal dysraphism, the lateral processes of the vertebral arch are separated, the neural canal is exposed posteriorly, and the posterior line and the overlying soft tissues are absent at this level [1]. When present, a surrounding meningomyelocele sac certainly aids the diagnosis of the spinal dysraphism. Ultrasound can predict the location and extent of the spinal defect with a high degree of accuracy [1]. We present the case of a 30 year old woman pregnant with a 18 weeks fetus with cervical open dysraphism.

Received 28.02.2012 Accepted 20.03.2012
Med Ultrason
2012, Vol. 14, No 3, 254-256
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Case report

A pregnant 30 year old woman was examined by ultrasound for her midtrimester ecography (18 gestational weeks). Cranial scans showed normal sized lateral ventricles and alterations of cerebral anatomy in form of "banana" sign and "lemon" sign, which are characteristic for Arnold-Chiari malformation. The careful examination of the spine, in multiple views (axial, coronal, sagittal, including 3D) found a cystic cervical mass associated with midline skin defect and minimal widening of the interpediculate distance of cervical vertebrae, an aspect consistent with open spinal dysraphism. No other anomalies were found with ultrasound. A Voluson 730 with convex, transvaginal and 3D transductors were utilized. At 22 gestational weeks, the patient decided to interrupt the

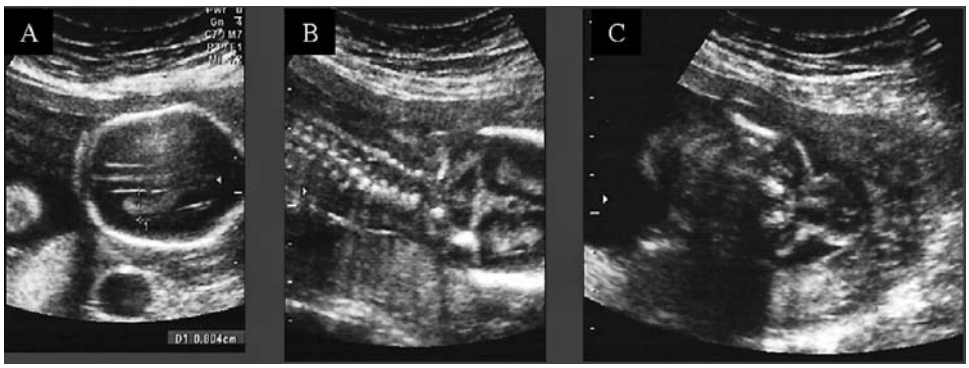


Fig 1. A: Cranial transverse scan showing bifrontal diformity of the calvaria, the lemon sign; the ventricles have normal diameter. B: Coronal section through the posterior fossa, the craniocervical junction and cervicothoracal spine, showing normal dimensions of the cerebellum, the obliteration of the cisterna magna, moderate splaying of the interpedicular distance in the lower cervical spine, moderate cervicothoracal scoliosis. C: Oblic coronal scan through the cervical spine showing the midline skin defect, the cystic appearance of the defect and only slightly splaying of the pedicles at the level of the defect.

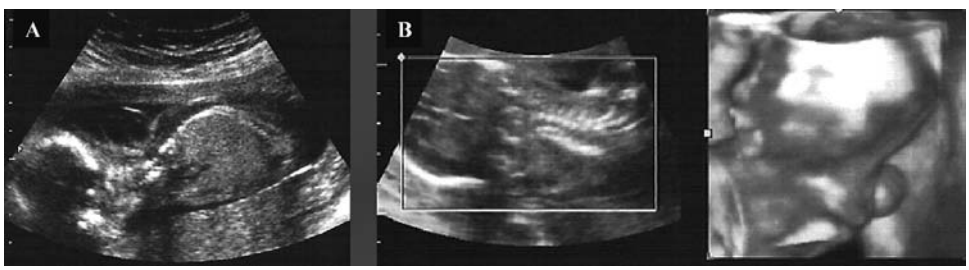


Fig 2. A: Parasagittal scan of the fetal spine showing the cystic cervical mass. B: 3D scan (surface rendering mode) of the posterior cervical region of the fetus showing the cervical mass.

course of pregnancy and the anatomopathological exam confirmed the anomaly.

Discussions

Cervical meningomyeloceles are extremely rare and consist of a fibroneurovascular stalk containing neurons, glia, and peripheral nerves emanating from a limited dorsal myeloschisis and penetrating through a narrow dorsal dural opening to fan out into the lining of a meningeal sac. In Pang`s et al series, the Arnold-Chiari malformation was present in only 44% of cases. [4].

Open spina bifida is associated with the Arnold- Chiari malformation [5]. The Arnold- Chiari malformation is characterized by a small posterior fossa with caudal displacement of the vermis, brainstem and fourth ventricle [6]. In the second trimester of pregnancy the manifestations of the Arnold-Chiari malformation are the "lemon" and "banana" signs [5]. At ultrasound the cisterna magna appears obliterated and the cerebellum is small size and abnormal shape and impacted deep into the posterior fossa; this condition is termed „banana" sign [1]. Frontal bossing, „lemon" sign, is also frequently present [1]. The lemon sign is found however in 1% of normal fetuses [6]. The observation that open spina bifida is associated with caudal displacement of the brain resulting in the „lemon" and the „banana" signs, shifted screening for this condition from maternal serum biochemistry to second trimester ultrasonography [5]. The cranial findings are more helpful for screening of spina bifida during the second trimester then the detection of the spinal defect itself [6]. The sensitivity of cranial signs in identifying spina bifida exceeds 99% [6]. The ventricular enlargement is present in a variable degree in virtually all cases of open spinal dysraphism at birth, but in less than 70% of cases in midtrimester; in most midtrimester cases is borderline or mild [1].

Our case is particular as the open spinal dysraphisms are only rarely localized in the cervical vertebrae. The ultrasound exam showed first the cranial defects that raised the suspicion of a vertebral defect. Careful further examination of the fetal spine showed the cystic posterior

mass with cervical location; the mass was only apparent when the fetus was far enough from the uterine wall to create a large liquidian interface between the fetal spine and the uterine wall that allowed the visualization of the mass. 3D views aided to the diagnosis. In our case, the splaying of the posterior arches of the vertebra was only minimal; this is characteristic for spinal defects with cervical location and makes the diagnosis more difficult in comparison with the larger bony defect with lumbosacral location which are easier to identify.

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