DIASTEMATOMYELIA IN PRENATAL AND POSTNATAL MAGNETIC RESONANCE IMAGING

DIASTEMATOMYELIE V PRENATÁLNÍM A POSTNATÁLNÍM OBRAZU MAGNETICKÉ REZONANCE

case report

Jaroslav Brichta[1]
Jarmila Skotáková[2]
Romana Gerychová[3]
David Horák[4]

[1]Clinic of Neurology, St. Anne Faculty Hospital, Brno
[2]Department of Pediatric Radiology, Brno Faculty Hospital, Brno
[3]Clinic of Obstetrics and Gynecology, Brno Faculty Hospital, Brno
[4]Department of Radiology, University Hospital Olomouc, Olomouc

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Adrresse for correspondence:
MUDr. Jaroslav Brichta
Clinic of Neurology, St. Anne Faculty Hospital Brno
Pekařská 53, 656 91 Brno, Czech Republic
e-mail: jaroslav.brichta@fnusa.cz

SUMMARY

Brichta J, Skotáková J, Gerychová R, Horák D. Diastematomyelia in prenatal and postnatal magnetic resonance imaging

Diastematomyelia or split cord malformation is a rare congenital anomaly. It represents a type of spinal dysraphism in children with the portion of spinal cord divided into two hemicords. In presented case Pang type I diastematomyelia with the hemicords divided by the bony septum is documented in prenatal magnetic resonance imaging (MRI), followed by postnatal MRI and computed tomography (CT) study. According to presented case, diastematomyelia without any other serious fetal anomaly may carry a good prognosis if early exactly diagnosed and treated with prenatal multidisciplinar counselling followed by proper postnatal treatment.
Key words: neural tube defects, prenatal magnetic resonance imaging, early diagnosis, prognosis.

SOUHRN

Brichta J, Skotáková J, Gerychová R, Horák D. Diastematomyelie v prenatálním a postnatálním obrazu magnetické rezonance

Diastematomyelie neboli rozštěpení míchy je vzácná vrozená anomálie. Představuje typ spinálního dysrafismu u dětí s rozdvojením části míchy. V uvedeném případě je prenatální magnetickou rezonancí (MRI) a následnou postnatální MRI a výpočetní tomografickou (CT) studií dokumentován případ diastematomyelie typu I dle Panga s míchou rozdělenou na dvě části kostním septem. Případ dokumentuje dobrou prognózu diastematomyelie sdružené s hemivertebra hrudní páteře bez neurologického deficitu v případě včasné a správné diagnostiky, multidisciplinárního přístupu a správného ošetření po porodu.
Klíčová slova: diastematomyelie, defekty neurální trubice, prenatální magnetická rezonance, včasná diagnostika, prognóza.

INTRODUCTION

Diastematomyelia or split cord malformation is a rare form of the spinal dysraphism in children characterized by complete or incomplete division of the spinal cord into two hemicords. The hemicords are sometimes divided by a septum of bone, cartilage or fibrous tissue. The pathology was first described by Cruvelhier in 1853. Most cases are associated with other anomalies of the spinal column such as hemivertebra, butterfly vertebra, spina bifida or kyphoscoliosis (1). Cutaneous stigmata on the back are present in more than 50% of patients, urologic dysfunction and progressive kyphoscoliosis can develop in older children. Diastematomyelia may be also associated with visceral anomalies such as horseshoe or ectopic kidney, ano-rectal or utero-ovarian malformations or became a part of severe craniorachischisis syndrome (2). According to

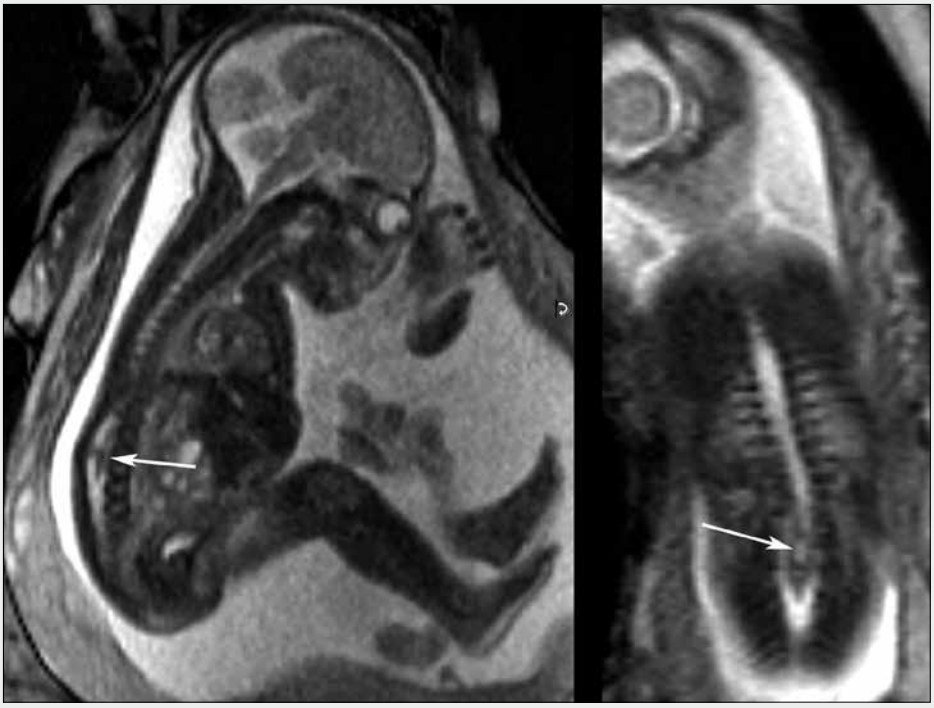

Fig. 1. ***Prenatal MRI showing diastematomyelia in L3 level***
Obr. 1. ***Prenatální MR obraz ukazující diastematomyelii v úrovni L3***

▲ *Obr. 1*

Pang (1), diastematomyelia is divided into 2 types. Pang type I is characterized by separate dural sac with arachnoid space surrounding each hemicord and osseous or fibrous spur. Pang type II features single dural sac and arachnoid space with no osseous spur, possible adherent fibrous bands tether the cord. The first visual signs of this anomaly can be visible even on the first-trimester ultrasound (US) screening, but usually it can be detected on the second-trimester US anomaly scan. The typical finding is the echogenic focus in the axial view of the spine. This finding has been detected in 0.06% of 10 070 prospective US examinations (3). For exact diagnostics of diastematomyelia type and exclusion of other fetal spinal abnormalities combining US and fetal MRI seems to be useful (4).

CASE REPORT

30-year-old pregnant women, gravida 2, para 1, was referred for the counselling, as she asked for the termination of the pregnancy. The pregnancy was after in vitro fertilisation/intracytoplasmic sperm injection (IVF/ICSI) procedure, parents refused the first-trimester screening for chromosomal abnormalities. Anomaly US scan at 23 weeks of gestation showed a widening of the lumbar spine in the coronal view with the hyperechogenic focus there and intact skin overlying the defect. There were also abnormal apperance of the fetal spinal curvature. The diagnosis of congenital cord anomaly was made and prenatal MRI was recommended. On MRI scans made 2 weeks later the symmetrical diastematomyelia in lumbar region and multiple hemivertebras in thoracolumbal region were identified (Fig. 1).Maternal serum α-protein levels were within normal range, female karyotype result 46XX was normal and no other fetal anomaly was detected. After neurosurgery specialist consultation, the parents were adviced to continue the pregnancy. At 36+0 weeks of gestation the female neonate was delivered via Caesarean section, with birth weight 2680 g, Apgar score 10/10/10 and without need of any special intensive care. The baby was well without any neurological symptoms except of hypertrichosis and mild dextroconvex scoliosis in thoracolumbar region. MRI and computed tomography (CT) was perforemd at the age of 2 months (Fig. 2 and 3). Defective thoracic spine formation and segmentation with T10–T12 hemivertebrae and ossified septum at L3 level were diagnosed. The infant underwent neurosurgery at the age of 4 months. During the surgery the bony septum was removed and the doubled spinal cord placed into a common dural sac. After the surgery the infant retained spontaneous mobility of her lower limbs and her sphincter functions remained intact.

DISCUSSION

Neural tube defects are possible to diagnose during the routinely performed prenatal US screening programme. Fetus US examination is usually done at the age from 19th to 20th week of the gestation age (GA). Diastematomyelia appears on US as hyperechoid structure clefting the fetal spinal structures. Nowadays three-dimensional US with multiplanar views can achieve better diagnostic accuracy. To verify the diagnosis prenatal MRI is the method of choice. Diastematomyelia is usually associated with other spinal anomalies: spina bifida, kyphoscoliosis, hemivertebras. Scoliosis appeares in more than 50% of patiens with diastematomyelia (5, 6), females are more often affected than males. There is no literature consensus regarding the consecutive surgery treatment. The surgery and bony septum removal should be performed as prophylactic measure (5, 7, 8), as prompt diagnosis and prompt prophylactic operations

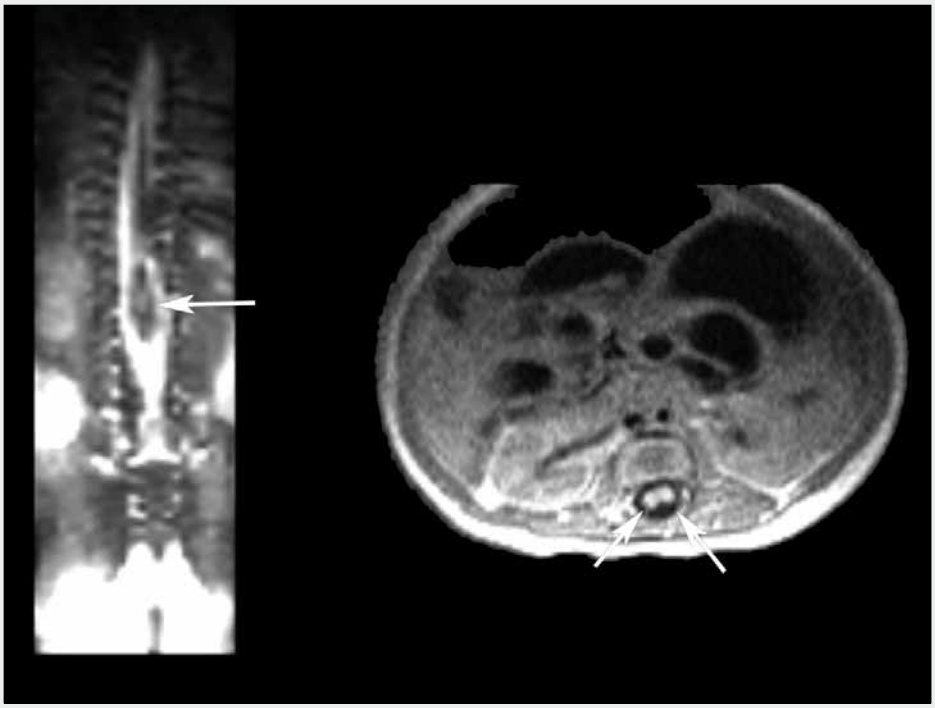

Fig. 2. ***Postnatal MRI with diastematomyelia finding***
Obr. 2. ***Postnatální MR obraz s nálezem diastematomyelie***

▲ *Obr. 2*

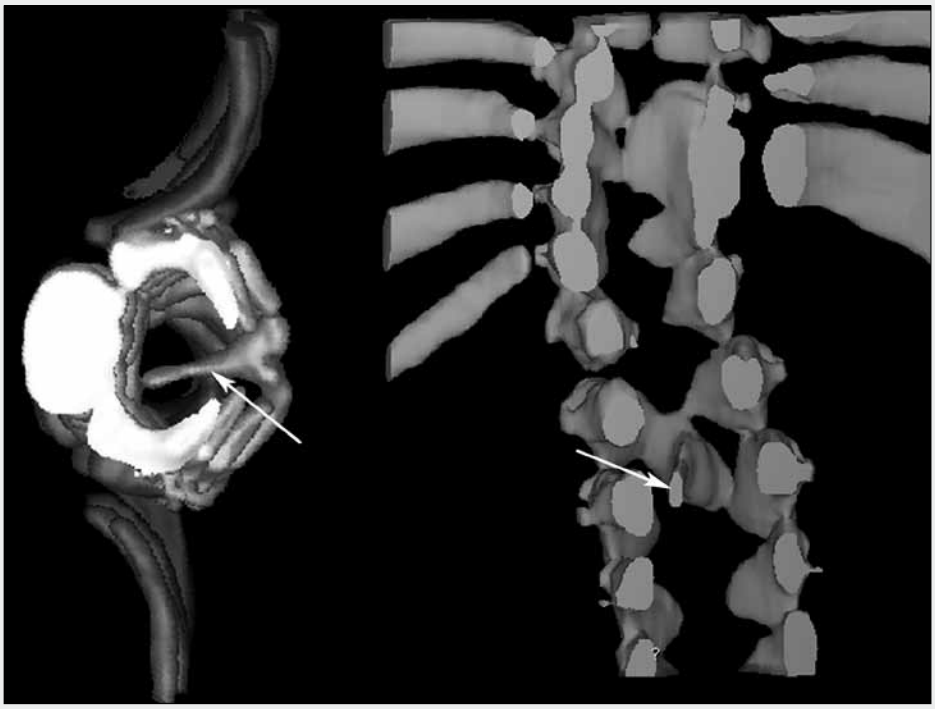

Fig. 3. ***CT 3D showing ossified septum at L3 level***
Obr. 3. ***Postnatální CT 3D obraz s osifikovaným septem na úrovni L3***

▲ *Obr. 3*

were associated with the best clinical outcome and good prognosis (9, 10). Other authors argue that the surgery should be performed only in case of progressive neurological symptoms to avoid possible complications and perioperative morbidity in still asymptomatic patiens (11). In our documented case, the surgery was performed to prevent the possibly irreversible neurological symptoms as surgery treatment usually cannot improve the neurological deficit preoperatively prominent (12, 13).

CONCLUSION

Based on presented case it seems that prenataly exactly US/MRI diagnosed diastematomyelia without any other serious fetal anomaly may carry a good prognosis, if treated with prenatal multidisciplinar counselling and followed by proper postnatal treatment.

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