ANNALES
UNIVERSITATIS MARIAE CURIE-SKŁODOWSKA
LUBLIN – POLONIA
VOL. LXII, N 1, 63 SECTIO D 2007

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Split cord malformation type I detected on multislice computed tomography

Split cord malformation (SCM) is a rare spinal defect. It is a congenital malformation where distal spinal canal is bisected by a septum. There are two types of SCM- SCM type I where two hemicords are separated by a spur at least partly ossified and SCM type II where doubling cord residue is in one dural tube and is separated by a non-rigid fibrous spur. Computed tomography (CT) and magnetic resonance (MR) (2, 8) play a major role in diagnostic process of these malformations. We present a case of an 11-year-old girl with bisected spinal canal diagnosed with multislice computed tomography (MSCT).

CASE REPORT

An 11-year-old girl with thoracic scoliosis was admitted to the Department of Radiology for assessment of a vertebral column. The examination was performed using an 8-row multislice computed tomography (MSCT). The scanning was conducted with 2.5 mm collimation, table feed 17.5 mm/s, rotation time 1 s, pitch 0.875. The analysis of pathological changes was performed with 2D multiplanar reconstructions (MPR) – frontal, sagittal, axial, oblique and curved, 3D volume rendering images and virtual endoscopy.

The examination revealed severe thoracic rotoscoliosis. CT scan of this patient showed the vertebral column deformation from Th 1 to Th 7 level. Vertebral bodies of these vertebrae were accreted and spinous processes were deformed (Fig. 5). Computed tomography studies demonstrated also that the 2nd, 3rd and 4th ribs on the left were accreted in the perivertebral region and the 3rd and 4th ribs were also accreted in the midscapular line.

CT scan showed an osteofibrous septum inside the spinal canal (Fig. 1, 2), a bony septum arising from the anterior surface of arch of the 4th thoracic vertebrae to the posterior wall of the 6th thoracic vertebral body (Fig. 4A). In the anterior part the bony septum was wider than in the posterior part (Fig. 4B). This septum divided the spinal canal into two symmetrical halves. Each part contained the separated dural tube with hemicord inside (Fig. 3). The separated spinal cord arising from the 1st thoracic vertebra level and joining together at the level of the 7th thoracic vertebra was visualized. CT scan also revealed the widened spinal canal at the level of splitted cord (Fig. 1).

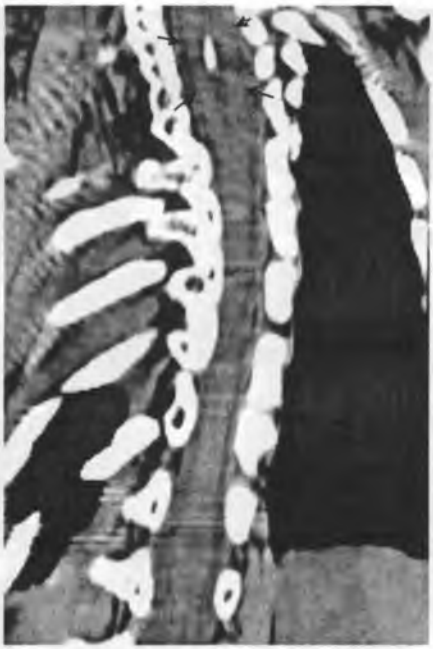

Fig. 1. Curved line reformation at the level of spinal canal. Osseous spur between splitted cord in the upper part of thoracic vertebra

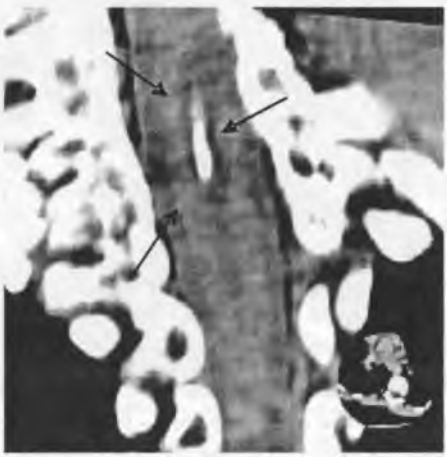

Fig. 2. Multiplanar reformation in the plane of split cord. CT image demonstrates bony spur and spinal cord cleft which extends from Th1 to Th7

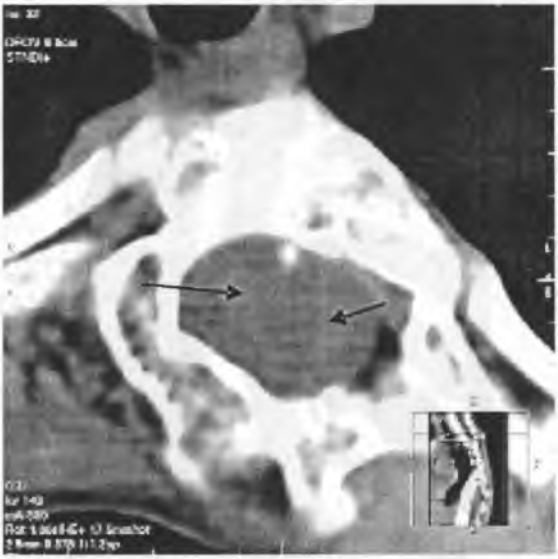

Fig. 3. Axial view in the plane below bony septum. This CT scan shows two separated hemicords and the lower part of the osseous septum

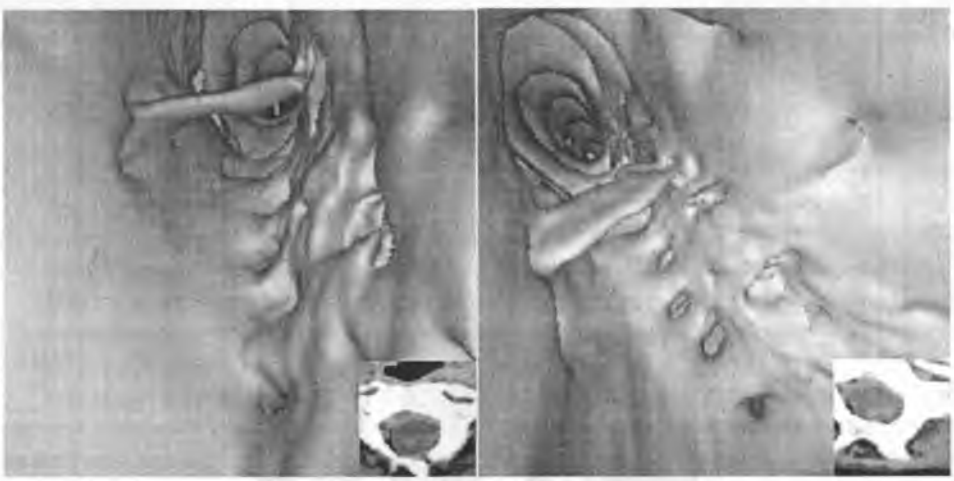

Fig. 4. Volume rendering virtual endoscopy view – reconstruction of vertebral canal. (A) superior view, (B) left lateral posterosuperior view

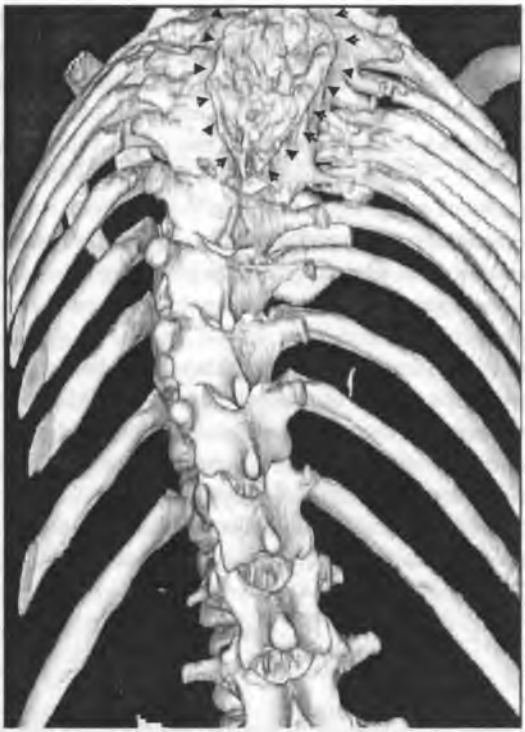

Fig. 5. Volume rendering view. 3D-reconstruction demonstrates deep lateral spinal scoliosis, anomalies of spinous processes and transverse processes in the upper part of the thoracic vertebral column

DISCUSSION

Split cord malformations are rare congenital anomalies. The theory explaining the embryological mechanism which causes SCM was proposed by Pang et al. (3). According to this theory SCM results from an ontogenic error occurring during the 3rd gestational week and is connected with existence and persistence of an accessory neurenteric canal (ANC). ANC connects the yolk sac with the amnion and that results in contact between the ectoderm and endoderm within the canal. This abnormal structure causes splitting of the regional notochord. The type of split cord depends on the nature of the septum and the state of the dural sac. So there are two types of SCM: type I – doubled cord resides in two dural sacs and is separated by an osseo-cartilaginous septum, type II – one dural sac is divided by a non-rigid fibrous septum (1, 3, 5). The case presented in the current report appears to belong to SCM type I.

Bony spur may divide the vertebral column and the cord into two symmetrical halves (when spur is strictly anterior-posterior). It can also divide the spinal canal asymmetrically with one normal and one hypoplastic hemicord. The most frequent location of the spur is at the lumbar level L3, L2, L4 and rare location of the spur is in the superior thoracic vertebrae and below S1 (3, 4, 5, 7).

SCM is very often associated with different bony anomalies such as scoliosis (like in our case), block- or hemivertebre, intersegmental fusion or *spina bifida* (8). Various cutaneous markers like hairy patch, subcutaneous lipoma, and cutaneous capillary hemangioma (6) are related with this condition.

Children with SCM at the birth may not present any neurological deficits. The neurological symptoms may appear with growth of the child. The main symptoms are connected with gait disorders and progressive spinal and foot deformities. Schijman reported 22 cases of SCM in children, 17 with SCM type I and 5 with SCM type II. Nine patiens had neuro-othopedic abnormalities (such as muscular atrophies and joint deformities in the lower limbs), 6 – with cutaneous stigmata without neurological abnormalities, 7 – with meningocele or meningomyelocele (7).

Sometimes first symptoms appear in the adult life. Adult patients suffer from dysesthetic pain in the legs and perineum and sensorimotor dysfunctions. Skalej et al. presented rare case of SCM diagnosed at a 48-year-old woman who suffered from an incomplete monoparesis of the left leg (8). It must be stressed that both types – SCM type I and SCM type II are equally likely to cause neurological abnormalities.

Some abnormalities in SCM can be detected on plain x-rays. MRI enables the study of the spinal cord and dural sac. However, the method of choice (especially in SCM type I) for study of the nature of the spur is multislice computed tomography and especially MPR and 3-D images, which allow its clear visualization to other specialists. CT enables correct localization and accurate measurements of the osseous spur which is very important for surgical procedures. CT also plays the major role in classification of SCM due to the possibility of detection calcified structure (SCM type I). The priority role for CT results also from the fact that SCM is frequently associated with different anomalies of bony structure (8).

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SUMMARY

Split cord malformations (SCMs) are rare spinal defects where the spinal canal and the cord are divided into two parts. All split cord malformations result from one pathology – arising an accessory neurenteric canal between yolk sac and amnion through the midline embryonic disc that split the regional notochord. Depending on the nature of the septum and the state of the dural sac SCM is divided into two types. We present a case of an 11-year-old girl, suffering from thoracic scoliosis, who was diagnosed with 8 -row multislice computed tomography as SCM type I. We stress the role of multislice computed tomography in the diagnostic process of SCMs and the role in classification

of SCM. Although MR is the method of choice for spinal cord anomalies, only computed tomography with multiplanar and 3D-reconstructions visualizes anomalies of bony structures or osseous spur, so important for future planning of surgical procedure, like in this case.

Rozszczep rdzenia kręgowego typu I wykryty za pomocą wielorzędowej tomografii komputerowej

Rozszczep rdzenia kręgowego jest rzadko występującą wrodzoną malformacją, w której kanał kręgowy i rdzeń podzielone są na dwie części. Wszystkie te wady powstają w wyniku błędu w rozwoju ontogenetycznym – co wiąże się z wytworzeniem dodatkowego kanału nerwowego pomiędzy woreczkiem żółtkowym a jamą owodni w linii pośrodkowej embrionu, powodując miejscowe rozszczepienie struny grzbietowej. W zależności od rodzaju przegrody i typu worka oponowego wyróżnia się dwa typy rozszczepu rdzenia kręgowego. W pracy przedstawiono przypadek 11-letniej dziewczynki ze skoliozą kręgosłupa piersiowego, u której zdiagnozowano rozszczep rdzenia kręgowego w badaniu wielorzędowej tomografii komputerowej. Autorzy podkreślają rolę MSCT w diagnostyce rozszczepu rdzenia i jego klasyfikacji. Pomimo że rezonans magnetyczny pozostaje badaniem z wyboru w diagnostyce wad wrodzonych rdzenia kręgowego, to jednak badanie MSCT z zastosowanie wtórnych rekonstrukcji multiplanarnych i opcji 3D pozwala zobrazować anomalie struktur kostnych kanału kręgowego i przegrody kostnej, co jest ważne w planowaniu przyszłego leczeniu operacyjnego, tak jak w tym przypadku.