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Clinical Manifestation of a Tethered Cord Syndrome at Children and Research Methods for Early Diagnosis of Disease

GALIMZHAN MAMIRBEKOVICH YELIKBAYEV and AIGERIM AITBAYEVNA TUTAYEVA

Department of Surgical diseases, International Kazakh-Turkish University K..A.Yassawi.

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ABSTRACT

Identification of the main clinical manifestation of a syndrome tethered cord at children and application modern visual research methods for early diagnosis of the disease. Are surveyed 30 children. The age patients was from 12 months to 18 years. Tethered cord syndrome in 21 supervision arose at pathological processes at the level of lumbar and sacral department of a backbone, at 5 patients the lumbar level, 3 in sacral and 1 patient on chest of vertebras. Skin manifestations at a tethered cord syndrome at 77% of children were shown hypertrichosis, a hypodermic lipoma, change of pigmentation of skin. Progressing of neurologic deficiency were shown by muscular weakness in 87% of cases and violation of sensitivity at 70% of patients. Impaired function of pelvic organs encountered in 90% of children. Main problem in diagnostics of a syndrome of the tethered cord at children at the present stage is that the few parents and practical doctors know about value of some skin manifestation, neurologic, orthopedic, urological violation which can signal about a back dizrafizm. The perspective direction is considered presurgical research of the neurologic and urological status of patient using an electromyography, a cistometrography and caused somatosensory potential.

Key words: Tethered cord syndrome, Kyphoscoliosis, Conus medullaris, Cistometrography, Children, Hypertrichosis.

INTRODUCTION

The tethered cord syndrome is the progressive form of neurologic deterioration arising because of violation of extent of a spinal cord various fixing back dizrafic changes[1, 2, 3].

Significant growth in number of patients, number of publications on early diagnostics syndrome of the tethered cord it is connected about introduction in practice of a magnetic and resonant tomography, use of monitoring of functions of the spinal structures involved in process (an electromyography, somatosensory and the sexual touch caused potentials) which increase efficiency of treatment[4,3,5]. Still, actual are questions of semi-illiteracy of parents and doctors on clinical manifestations a disease research methods for early diagnostics of tethered cord syndrome with children.

MATERIALS AND METHODS

Results of inspection of 30 patients of children's age with a tethered cord syndrome treated in neurosurgical depertment of regional children's hospital in Shymkent (Kazakhstan) from 2010 to 2015 are studied. The age of patients fluctuated from 12 months to 18 years. The main group was made by children of school age – 26 (52%) supervision. Ratio on a sex – 1:1,5 girls prevailed. Tethered cord syndrome in 21 supervision arose at pathological processes at the level of lumbar and sacral level of a backbone, at 5 patients at the

lumbar level, 3 in sacral and at 1 patient on chest of vertebras.

Diagnostics was reduced to comparison of clinical, the visual (CT, MRI), electrophysiological researches.

RESULTS

The clinical picture of the patient's admission to the hospital consisted of progressive neurologic deficit manifested by muscle weakness, violation of sensitivity, dysfunction of the pelvic organs. Orthopedic strain characterized by kyphoscoliosis, chest deformity, clubfoot, limb shortening with gait disturbance. Hypertrichosis, subcutaneous lipoma, skin pigmentation changes in skin manifestations were tethered cord syndrome. Hypertension-hydrocephalic syndrome was detected in 47% of cases. The defeat of the cranial nerves was noted in 27% of cases, and violation of static - in 23% of patients.

Pathological tethered cord syndrome led to vascular disorders because of its mechanical tension and displacement, causing the deterioration of the clinical picture of the disease.

In radiodiagnosis, 93% of children identified cleft arches spine, while 53% - more than four vertebrae. MRI and CT studies showed spina bifida occulta and deformity of the spine. In 14 (46%) of children progressing kyphoscoliosis accompanied by paresis of the lower limbs, combined with increasing bladder dysfunction. Myelography with contrast was performed in 20 (67%) children, where space is not enveloped subarachnoid space spinal cord and a ponytail on all sides. Cauda equina rootlets never left hemisphere posterior dural sac, indicating the likelihood of having counterfoil of the tethered cord syndrome.

MRI and CT examination of the spine and spinal cord performed in all cases. Along with this, 37% was made electroneuromyography lower extremities, ultrasound - 10%, in 6% of cases are caused somatosensory potentials.

Literature review and discussion

Causal factors of the syndrome may be trauma, tumors, malformations of the spine and spinal cord and cause mechanical tethered cord syndrome inelastic structure is located at the caudal end of the spinal cord, which prevents movement. It most often occurs in the lumbar-cross the area, but may be at any level of the spinal canal. Many studies have reported that the tethered cord syndrome is often found in girls[6, 7, 8], which is confirmed by our data.

Development of the syndrome in children and adolescents due to factors tethered cord syndrome and its components in the pathological focus in the continuing growth of the patients. All this leads to a mechanical stretching, the displacement of the spinal cord development in ischemic it further neuronal membranes rupture. The stretching of the spinal cord occurs in patients when the spine is growing faster than the spinal cord or when the spinal cord is exposed to the violent intervention of the release[9] and is found in 3-15% cases after surgery for myelomeningocele recovery[10, 4, 11].

Mechanical causes fixing terminal thread includes thickening filum terminal when the elongate spinal cord, as well as any non-elastic structure (fiber or fat tissue, tumors, lipoma, epidermoid tumors, myelomeningocele lipomyelomeningosele, scar formation) which are fixed to the rear portion of the spinal cord to the dura shell or bone septum[12, 13, 14]. These structures lack of movement, thereby causing the voltage at their fixation in the vertebral channel[9].

Clinic tethered cord syndrome clinically skin changes, vertebral abnormalities, orthopedic disorders (scoliosis and clubfoot), neurological deficits of the lower spinal cord, with a dysfunction of the colon and urinary bladder[15, 16, 17].

Skin changes

For cutaneous manifestations of cutaneous hemangiomas are tethered cord syndrome, hypertrichosis, dermal sinus, subcutaneous lipoma, and the appendage of the skin on the middle line in the lumbosacral region

and occurs in approximately 50% of patients with tethered cord syndrome[18]. These skin markers are often the key to the recognition of the basic dizrafic. Doctors should be a great alertness and suspicion when examining a child who has discovered a lipoma, dermal sinus hemangioma or dark spots on the middle line, hypertrichosis and asymmetric folds buttocks[19, 20, 21, 22]. Cutaneous manifestations of the syndrome of tethered cord syndrome in our study, 23 (77%) children were shown hypertrichosis, subcutaneous lipoma, changes in skin pigmentation.

Orthopedic changes

Neuroorthopedic syndromes include strain and muscle atrophy of the lower limbs, unsteady gait, pain in the limbs, scoliosis, kyphosis, clubfoot, congenital dislocation of the hips. According McLone D.G. et al. (1999) 75% of patients with tethered cord syndrome having orthopedic disorders[4]. In our study, orthopedic deformation occurred in 26 (87%) cases and manifested kyphoscoliosis, chest deformity, 24 (80%) children with clubfoot marked shortening of the limbs, and gait disturbance. The development of scoliosis and lordosis is the result of functional disorders of about vertebrate muscles, leading to curvature of the spine so that the spinal cord took the shortest course in the concave side of the spinal canal to reduce intramedullary tension. Deformation of the lower limbs and clubfoot apparently caused by the weakness of some muscles (due to the intramedullary lesions) who lose their balance Group antagonists muscles in the legs and feet. This imbalance in muscle strength similar to the changes arising from the ulnar nerve palsy[9].

Anomalies found bones of the spine in patients with tethered cord syndrome include spina bifida plate defects, semi vertebrae, the sacral aplasia, segmentation violation and can be detected in 95% of patients with tethered cord syndrom[18].

Neurological changes

progression of neurological deficits include pain in the lumbar region, weakness in the lower extremities, asymmetric hyporeflexia, increased muscle tone in the lower limbs, disturbance of sensitivity and occurs in 75% of patients with tethered cord syndrome[4]. In our study the progression of neurologic deficit manifested by muscle weakness in 26 (87%) cases, and violation of sensitivity in 21 (70%) patients.

Urological aspects

incontinence myelodysplasia common problem with which patients come to the urologist[23] The findings Tarcan T, Bauer S, Olmedo E. et. el. (2001) show that newborns with congenital malformations of the spine and spinal cord have a 32% risk of neural urological deterioration caused by the tethered cord syndrome[24]. Tethered cord syndrome after primary closure of the defect due to postoperative adhesion or infection are well recognized risk factors for neurologic impairment in patients with myelodysplasia. This condition can develop years after the surgery, but it is still unknown incidence. The results of urodynamic studies incontinence show that is often marked by hyper-reflex detrusor[25]. Impaired function of pelvic organs occurred in our study, 27 (90%) children.

Diagnostics

Diagnosis tethered cord syndrome easily put in the comparison of the clinical picture and the study of neuro-image. Clinical signs of spina bifida occulta, including skin changes, orthopedic disorders, vertebral anomalies, as well as progressive deterioration of neurological status and Urologic dysfunction associated with the conus medullaris give reason to suspect tethered cord syndrome. Typical features of neuro radiology as an elongated filament, the presence of a thick filum or tumor confirmed the diagnosis.

The bladder and sphincter of the rectum are the most likely areas of involvement in the pathological process and the changes in their functions[26, 27]. Therefore, urinary urodynamic studies are the primary diagnostic methods to assess whether the tethered cord syndrome. The change in urodynamic parameters, radiological signs of impairment and urinary incontinence should alert the urologist arisen about tethered cord syndrome.

In studies Tarkan T., Bauer S., Olmedo E. et. al. (2001) urodynamics with external sphincter electromyography has been a valuable method of early diagnosis of the tethered cord syndrome[24].

Radiological examination, MRI have a positive side for the diagnosis of tethered cord syndrome after a previous spinal surgery[28].

In the classic version tethered cord syndrome produce traction on the changed conus medullaris[1, 29, 2] however, the tethered cord syndrome has also been described in the "normal" state of conus medullaris[30, 18]. Conus medullaris reaches adult levels within 3 months after birth. The incidence of patients with clinical signs of tethered cord syndrome at normal position conus corresponds to the frequency of patients with altered conus, respectively [30, 18]. None of the patients with tethered cord syndrome in the normal position of the cone urological dysfunction was not the only pathology[18]. We, consider the tethered cord syndrome as the cause of neurogenic bladder reflex hyper even if the conus medullaris is located in a normal position without changing the terminal filum.

Location terminal thread is not essential for the diagnosis of tethered cord syndrome as the caudal end of the spinal cord has been positioned above the lines between the vertebra L1-2 in 18% of patients with tethered cord syndrome [30] and 50% higher than L2 - 3 for X-ray studies in adults[17]. Despite the irregular thickness of the terminal thread (> 2 mm), it is only a relative value, so when the thickness filum terminal diameter less than 2 mm, the diagnosis of the tethered cord syndrome is not excluded (for example, 1 mm thickness was included in the diagnosis tethered cord syndrome) , particularly adolescents and adult patients[17,9]. Inelastic thread during the operation and detected by histological examination of the fibers filum replaced ependymal fibers proves the presence of hard yarn, as a mechanical source of high tension within the spinal cord[17, 14].

MRI, CT and ultrasound in real time, a simple X-ray, electrophysiological studies to help determine the location, the level of conus medullaris and identify the fixing agent[28].

Important in the diagnosis of the syndrome of tethered cord syndrome cord is attached to the MRI study conducted in our study, 22 (73%) patients, presenting information about the anatomical structures and fixing the level of the location of the cone. Fat formation is easily identified in the sagittal and axial slices through high intensity on T1 fat slices. MRI diagnostic criterion in tethered cord syndrome was displacement of the spinal cord caudal (below L2 vertebra) and dorsally.

CONCLUSIONS

The main problem in the diagnosis of the syndrome of tethered cord syndrome in children at the present stage is that few parents and clinicians are aware of the importance of certain skin manifestations, neurological, orthopedic, urological disorders that may indicate spinal dizrafizm. Early detection of disease and timely surgical intervention to help halt or stabilize the progression of neurologic deficit.

A promising area is considered to be pre-operative study of neurological and urological status of the patient using electromyography, cistometrography and called somatic sensory potential. The need for further development in the field of neuro urological approach to solving problems.

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