Case study

WHAT IS CEREBRAL PALSY?

ABSTRACT

Aims

To present the case in support of thorough examination of spinal cord in cerebral palsy.

Presentation of case

We report the case of 4-year-old boy with lower paraplegia and speech retardation. The tonus was increased bilaterally in gastrocnemius muscles and thigh adductors. The muscle tonus was decreased in iliopsous. Electrophysiological examination revealed signs of decreased excitability of motoneurons at the level L 2 – S 2. MRI has confirmed lesions of spinal cord at that level in addition to injury at thoracic level and brain lesions. MRI spinal angiography has detected tortuous anterior spinal artery. The boy benefited from the electrophoresis with theophylllinum, applied on lower thoracic and first lumbar vertebrae with improvement of his legs motor skills.

Discussion and Conclusion

The first explorer of cerebral palsy, Dr. Little, blamed on both brain and spinal cord for clinical appearance of this disorder. However, his follower, Dr. Freud, diverted attention of neurologists from the spinal cord and limited it to the brain. Our presentation testifies to involvement of spinal cord and benefits from therapy applied on spine and, thus, confirms Little's point of view.

Keywords: *cerebral palsy, anterior spinal artery, spastic diplegia, Adamkiewicz artery*

INTRODUCTION

Executive Committee of International workshop in Bethesda, MD (USA) declared in 2007 that "in summary, after more than 150 years of debate we not yet have a universally accepted definition of CP; nor do we have an agreed method for classifying the impairment that has been shown to be robust in terms of validity and reliability"[1]. 15 years after, nowadays, the state of affairs did not changed. Still clinical findings are not consistent with either ex vivo MRI – CT examination of the brain, or postmortem study. We believe that the reason of the obscurity is our deviation from the position of the first explorer of Cerebral Palsy, Dr. Little. Dr. Little documented in his proceedings in 1853 and in 1861 [2], that he and many of his colleges had examined spinal cord postmortem thoroughly and almost in every case had found lesions. However, his great follower Freud, in 1893 and 1897 in his proceeding, devoted to cerebral palsy [3], rested exclusively on brain examinations, and consequently did not find correlation between clinical phenomena and postmortem findings. For explanation of this discrepancy, Freud suggested several tricky hypotheses, like paramedian injury, repair of initial brain lesion, psychological maternal disturbances, appearance of primitive reflexes and so on…Our contemporary view of cerebral palsy is based mostly on abovementioned hypotheses of Freud, because of his great influence on neurologic community, while concept of Little has been almost forgotten. In 1986 – 1989, as exception to this tendency, appeared reports of R. Clancy, J. Sladky, and L. Rorke [4, 5]. These authors detected spinal cord lesions in most of cases postmortem examinations of the newborn, expired from asphyxia. Examination of our patient with cerebral palsy revealed

structural and vascular spinal cord lesions besides brain injury, and thus, is consistent with reports of Dr. Little and Dr. Clancy et al. Animals studies also testify spinal cord involvement [6,7,8,9].Nowadays many noninvasive methods of spinal cord investigation are available, and we advocate their use in cerebral palsy.

CASE PRESENTATION

Case report of the patient V., boy, 4 y. o., whose parents complained on disturbance of speech and gait. It was the fifth pregnancy for mother at the age of 40 and the 4th labor. Three previous pregnancies had resulted in birth of the healthy children; one pregnancy had frozen on the 9th week. All children from the same father. This, 5th pregnancy, initially went unremarkable with normal Ultrasound and Cardiotocography. However, unexpectedly it resulted in premature labor on 29th week, with tocolysis. Eventual delivery occurred by means of urgent cesarean section due to tetanic uterine contraction with prolapsed arm and umbilical cord entanglement. The newborn had Apgar score 6/7, weight of 1460 gr, height 40 sm. For the periventricular hemorrhage, detected by ultrasound, and respiratory failure, he was treated in critical care nursery of the maternal hospital for 4 days and received CPAP respiratory support. On the 5th day he was delivered to the resuscitation unit of the pediatric hospital. After 10 days care, he was discharged into the neonatology division. At the age of 33 days, he was discharged home with the weight 2100 gr. Neurosonography revealed periventricular/interventricular hemorrhage, periventricular leukomalacia in cystic phase. Duplex sonography revealed decreased blood flow velocity in anterior cerebral arteries, venous flow being normal.

Motor development: holds his head from 2 months, rolls from back into tummy from 7 months, sits unsupported from 1,5 years, crawls from 11 months, crawls on his four from 1,5 years, rises and stands holding the prop from 1,5 years, walks holding the furniture from 1,7 years, rises himself and walks holding the baby stroller from 2,5 years, walks and runs with walkers Crocodille R82, from the 3 years, cannot stand alone. At the age of two years duplex sonography revealed decrease of the blood flow in the right vertebral artery when turning the head to the left. At the age of three years old duplex sonography revealed non-pathologic tortuosity of both common and internal carotid arteries and vertebral arteries in V 1 segment. Clinical examination at the age of 4 y. o.: Comprehends the speech and carries out all verbal commands. Expressive speech is represented by several dysarthric words. Pupils are equal, direct and consensual pupilloconstriction are normal. Extraocular movements are restricted in directions of gaze up and right. Rare nistagmoid vacillations of the globes when gazing laterally. The face is symmetrical. The tongue is not deviated, but undulates when sticked out. The oral automatism reflexes are revived. Upper tendon reflexes are decreased evenly. The patellar reflexes are revived, the Achilles reflexes increased up to clonus. Babinski sign is positive bilaterally. Motor findings – muscle bulk was slightly decreased in paravertebral muscles at Th 6 – 9. Tetraparesis much more pronounced in legs. Spastic paresis was observed in gastrocnemius muscles, adductors of thighs and in less extent in bicepses brachii. Flaccid paresis was observed in iliopsoas muscles. Sensory findings – pain sense was normal, vibration sense was diminished in lower extremities (22 second in legs while 11second in arms) No meningeal signs.

EMG at two years old included

1. Study of motor nerve conduction velocity (NC) and M – wave (CMAP) parameters during the stimulation of tibial and peroneal nerves from both sides.

2. Study of H – reflex from both sides

3. Needle EMG of the left tibialis anterior and of the left vastus lateralis

4. Interferential analysis

Stimulation of motor fibers of the left peroneal nerve has revealed DECREASED AMPLITUDE OF M – WAVE (CMAP) AND ITS FORM DEFORMED, WHILE THE MOTOR NERVE CONDUCTION VELOCITY BEING NORMAL. Stimulation of right peroneal and tibialis nerves revealed normal amplitude of both distal and proximal M wave (CMAP) and

normal motor nerve conduction velocity. H – reflex is registered from the left, being normal in amplitude, while is not registered from the right.

Needle EMG of the left anterior tibialis muscle and of the left vastus lateralis has not detected any spontaneous activity. Motor unit potentials are of normal duration and phase, while their amplitude being sometimes slightly decreased. Interferential curves are saturated and of normal amplitude.

Conclusion:

Signs of decreased excitability of motoneurons at the level L 2 – S 2. There were no signs of neither polyneuropathy nor spinal muscular atrophy nor myopathy (Figure 1)

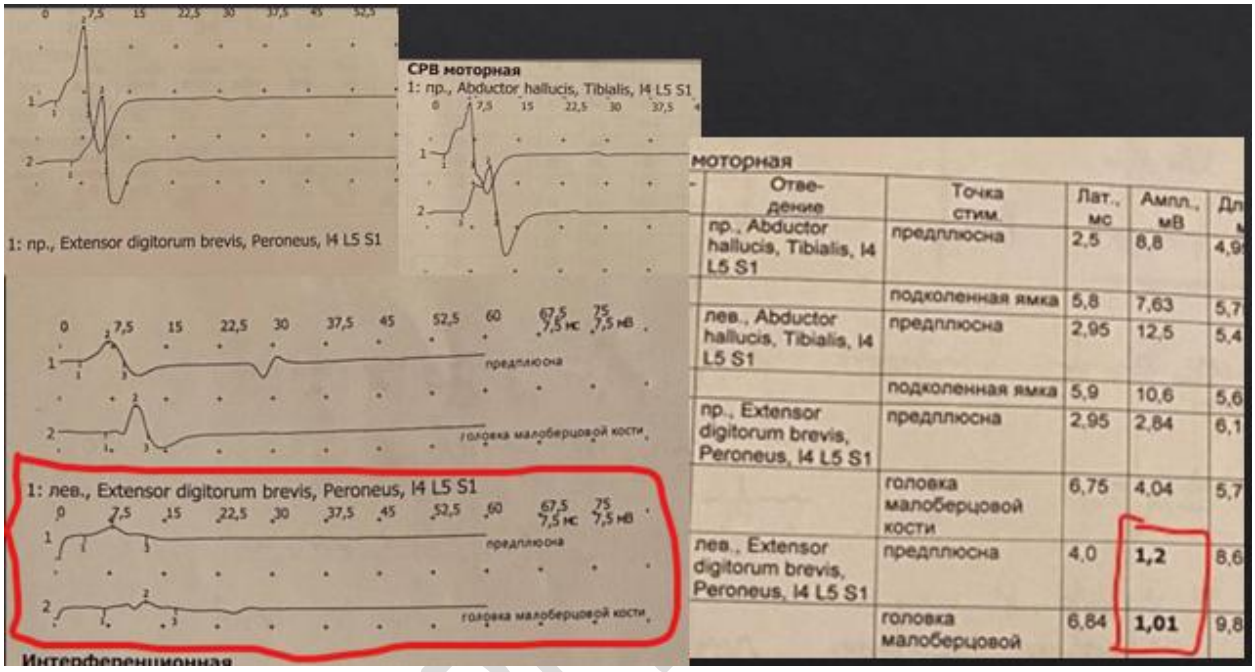

Figure 1. EMG of the patient V. Decreased and deformed M – wave (CMAP) is highlighted.

Brain and spine MRI with angiography at 4 years - old

(MR tomograph Siemens Skyra 3 T, Protocol T2 TRA, T2 FLAIR TRA FS, T1 SAG 3D, T2 COR, MDDW 20, Programm “Whole Spine)”

MRI features consistent with profound chronic posthypoxic and postischemic encephalopathy with the loss of white matter bulk and with periventricular leukopathy (areas of gliosis with cystic transformation). Symmetrical areas of hyperintensity in posterior parts of basal ganglia of residual posthypoxic origin. Both cortico – spinal tracts with the signs of Wallerian degeneration. Minor pineal cysts. Corpus callosum subatrophy. Hydrocephalus ex vacuo of lateral ventricles, cavum vergae. Spinal subatrophy at the Th 6 – Th 9 level with asymmetrical hyperintensity areas, more pronounced along the lateral columns. Diffuse weak heterogeneous increase of MRI – signal from the spinal cord below Th 6 vertebrae, along all the length of the cord, down to conus, apparently due to degeneration. Uncomplete fusion of laminae arcus vertebrae S 2 – S 3. Tortuosity of anterior spinal artery (Figure 2)

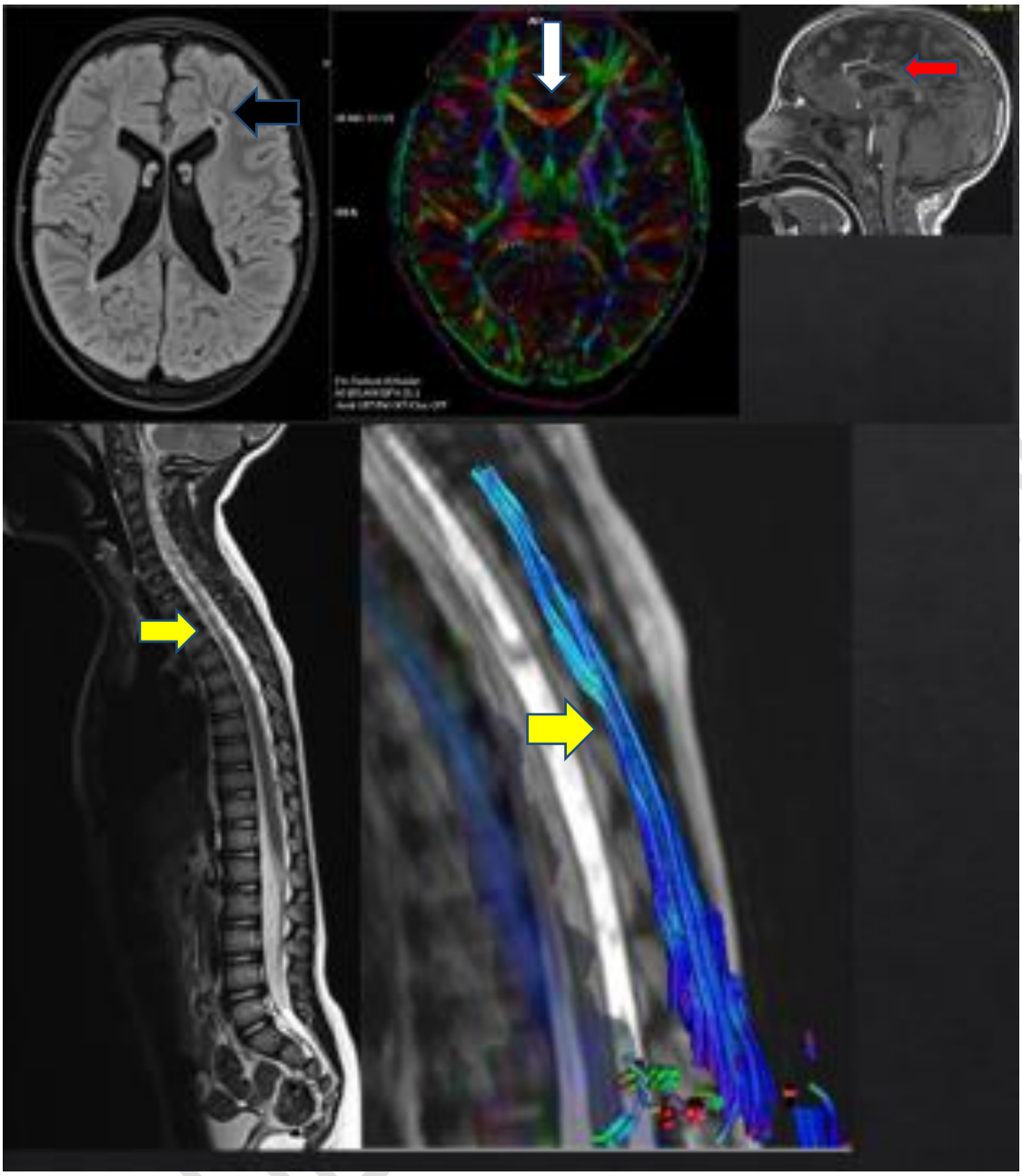

Figure 2. MRI with tractography of brain and spinal cord of patient V.

Black arrow – Periventricular areas of gliosis with cystic transformation, white matter depletion.

White arrow - Fibers of semioval centers pathways are depleted, cortico – spinal tracts with the signs of Wallerian degeneration.

Red arrow - Corpus callosum atrophy.

Yellow arrow - Depletion of spinal conducting pathways

The boy has benefited from electrophoresis with Theophylllinum, applied on lower thoracic and first lumbar vertebrae, followed by acupuncture (Figure 3), with improvement of his legs motor skills

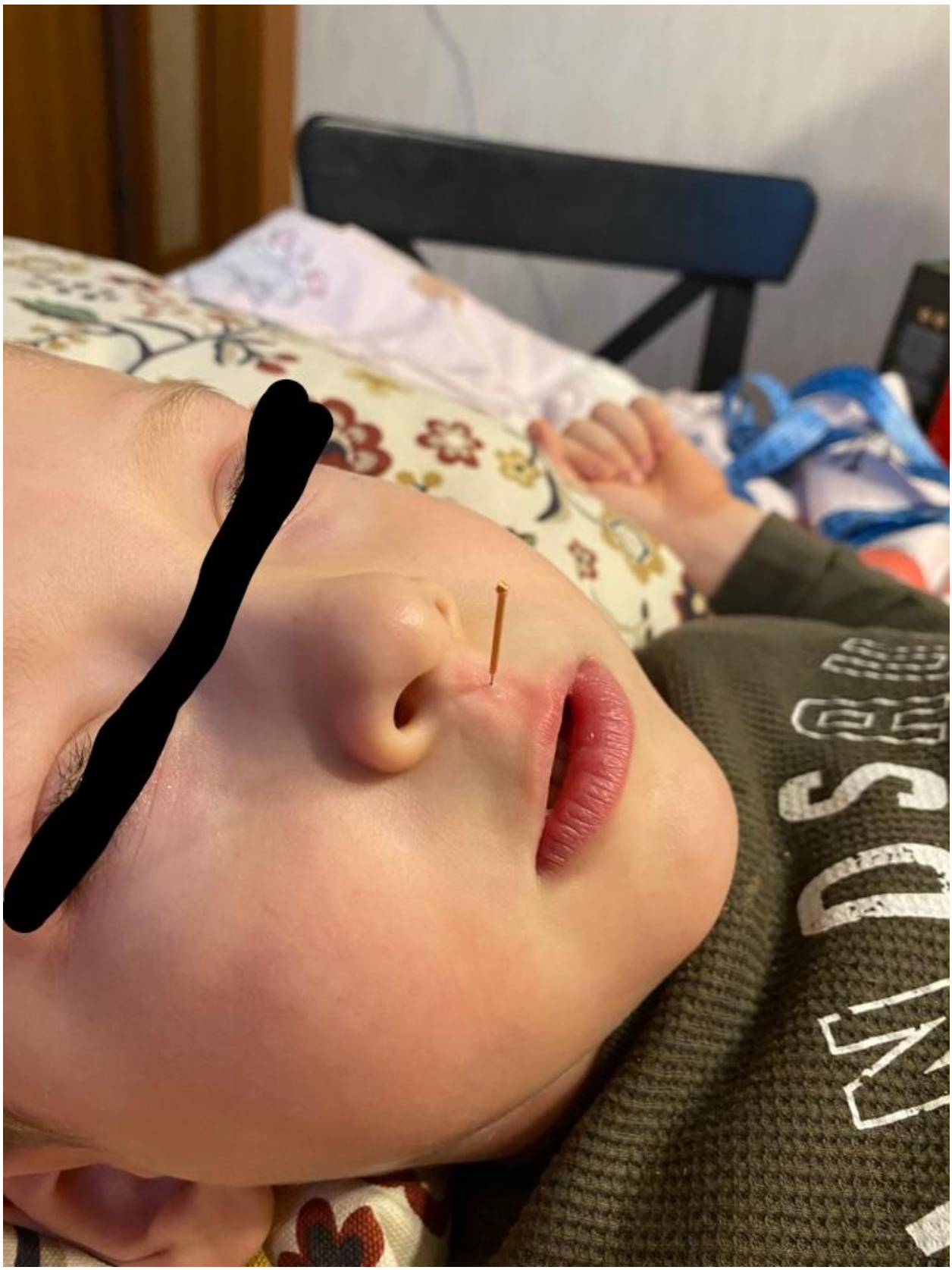

Figure 3. Acupancture treatment of patient V. with exposure on point Shui Gou.

DISCUSSION

Nobuyoshi Kawaharada in 2004 established usefulness of MRA in detection of Adamkiewicz artery (ARM) as well as 3 types of morphology of the anterior spinal artery (ASA) above the ARM junction [10]:

-Type A is noncontinuation of the ASA above the ARM junction.

-Type B is continuation of the ASA above and below the ARM junction.

-Type C is noncontinuation of the anterior spinal artery below ARM junction.

Our case is consistent with Type A according Kawaharada. (Figure 3). This type is more common though more vulnerable to ischemia

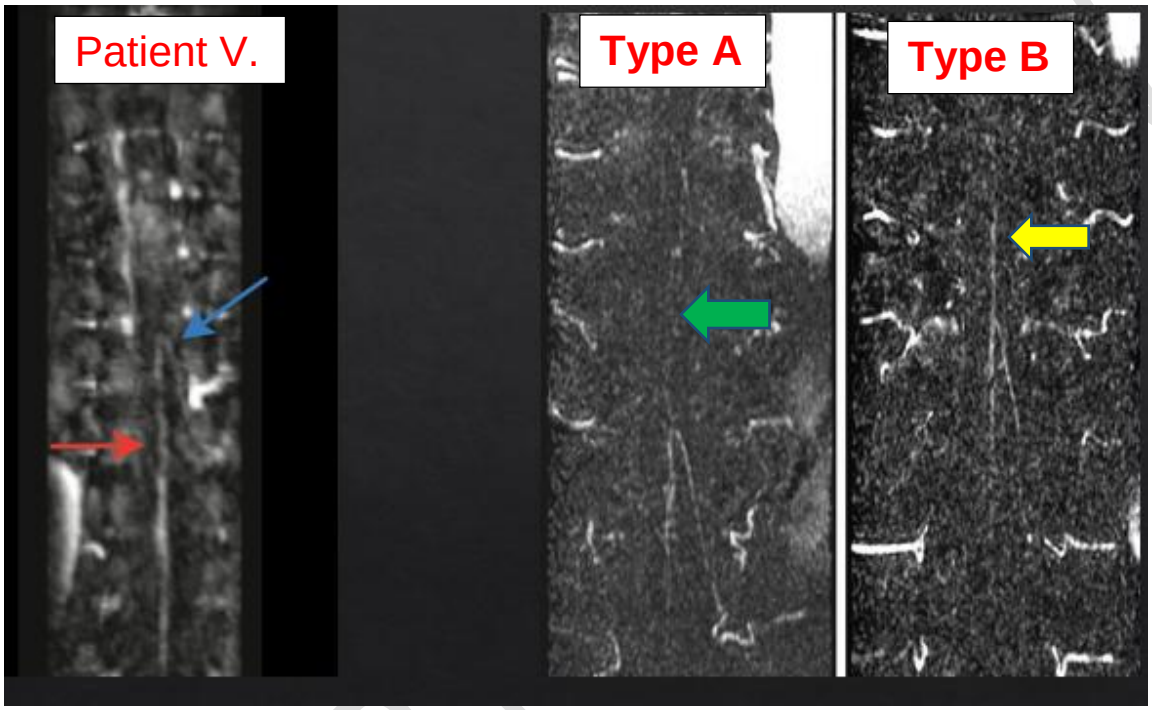

Figure 4. Comparison of spinal angiography of our patient V. (on the left) with established by Kawaharada two possible types of junction of Adamkiewicz artery with anterior spinal artery (on the right). Blue arrow – Adamkiewicz artery, Red arrow – tortuous anterior spinal artery of our patient V., green arrow – continuation of anterior spinal artery only down in type A junction, continuation of anterior spinal artery up and down in type B junction.

The first explorer of cerebral palsy, Dr. Little, reported his experience and of his colleges, when the spinal cord was always studied at autopsy, and lesions had been found in spinal cord almost in every section. The summary of his view of cerebral palsy could be expressed by the following: The injury in Cerebral palsy could result from birth trauma or hypoxia/ischemia or hemorrhage of both brain and spinal cord. Topical diagnosis is possible and imperatively required.

40 years later Dr. Freud, exploring cerebral palsy, came to opposite conclusion, when he neglected postmortem spinal cord examination [11]. His summery was the following: There is a poor correlation between clinical syndromes and neuropathologic lesions. Topical diagnosis of cerebral palsy is impossible. Neurological and psychiatric disorders cannot be firmly localized to a specific area of the cerebral cortex. The foundation was thus set for his broad and speculative explanations of cerebral palsy symptoms, as the sum of suffering of the parasagittal fetal brain area and its compensation process, and also based on the theory of psychoanalysis; a theory based on a theoretical construct of the mind

that had little correlation with the brain's anatomic pathology. Because of Dr. Freud's great authority, until now, we regard cerebral palsy from the Freud's point of view. The only exception made the group of scientists in 1986 – 1989, namely R. Clancy, J. Sladky, and L. Rorke. These authors detected spinal cord lesions in most of cases postmortem examinations of the newborn, expired from asphyxia.

Most cases of cerebral palsy arise because of hypoxic – ischemic encephalopathy (HIE). HIE in its turn, arises as response to change of systemic blood pressure, and accompanied by hemorrhagic insult, or ischemic insult, or periventricular leukomalacia, or parasagital lesions or combination of these forms. Preterm infants are known to be much more vulnerable to changes of blood pressure because of lack muscle layer in brain vessels. The lack of autoregulation in preterm infant is also true for the Adamkiewicz artery, the main blood supply to the lower spinal cord. When ischemia or hemorrhage takes place in brain, we just must expect the same event in spinal cord and include spinal cord in MRI study.

CONCLUSION

Many methods of noninvasive spinal cord investigation are available nowadays, and our case one more time advocates return to Dr. Little's view of cerebral palsy, and use of abovementioned diagnostic tools.

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