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MNiSW 1 pkt IndexCopernicus 3.22 wersja pierwotna

Rapidly progressing cord compression syndrome in a child

Szybko postępujący zespół ucisku rdzenia kręgowego u dziecka

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Streszczenie

Wstęp: Zwapnienia pajęczynówki współistniejące z tłuszczakiem kanału kręgowego zlokalizowanym zewnątrzoponowo, są rzadką przyczyną ucisku na rdzeń kręgowy, zwłaszcza u dzieci.

Opis przypadku: Prezentujemy 14–letniego chłopca, przyjętego z powodu postępującego niedowładu spastycznego kończyn dolnych, z ubytkami czucia oraz pęcherzem neurogennym. Od dzieciństwa obserwowano u niego niezgrabność i mniejszą sprawność ruchową. Zaburzenia chodu i kyfoskolioza nasilały się przez lata. Badanie rezonansu magnetycznego ujawniło tłuszczaka w odcinku piersiowym kręgosłupa. Z powodu gwałtownego nasilenia się zaburzeń neurologicznych, pacjenta operowano. Śródoperacyjnie oprócz tłuszczaka uwidoczniono pogrubiały worek oponowy. Badaniem histopatologicznym zweryfikowano obecne w nim zwapnienia pajęczynówki. Po zabiegu stan kliniczny pacjenta znacząco się poprawił.

Wnioski: Współistnienie różnorodnych zaburzeń prowadzących do zespołu ucisku rdzenia kręgowego, znacznie utrudnia proces diagnostyczny. Zwapnienia pajęczynówki są rzadkie, jednak powinny być brane pod uwagę, nawet jeśli wynik rezonansu magnetycznego (MRI) jest negatywny. Wczesna diagnoza prowadzi do leczenia operacyjnego, które daje zadawalające rezultaty.

Słowa kluczowe: zwapnienia pajęczynówki u dzieci, zespół ucisku rdzenia kręgowego, tłuszczak kanału kręgowego

Abstract

Background: Arachnoid calcifications coexisting with an intraspinal extradural lipoma are a rare cause of cord compression syndrome particularly in children.

Case report: We present a case of a 14–year–old boy who presented to our Department with rapidly progressing spastic paraparesis, sensation deficits, and neurogenic bladder. The first abnormal symptoms, namely decreased mobility and clumsiness had been visible since early childhood. Over the years kyphoscoliosis as well as gait disturbances developed. MRI of the spine revealed a lipoma of the thoracic spinal canal. Because the neurological symptoms were progressing extremely fast the child was scheduled for surgery. Intraoperatively, an extradural lipoma and a thickened dural sac were found. Histopathological examination detected arachnoid calcifications. After the surgery the patient made a satisfactory recovery.

Conclusions: Co–existence of various neurological disorders leading to the cord compression syndrome in children makes the diagnostic process challenging. Spinal arachnoid calcifications should always be considered even if magnetic resonance imaging (MRI) is negative. Early diagnosis leads to surgical treatment which gives satisfactory results.

key words: arachnoid calcifications in a child, cord compression syndrome, intraspinal extradural lipoma

Otrzymano: 20–05–2014 → **Zaakceptowano:** 30–06–2014 → **Opublikowano:** 17–07–2014

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Introduction

Two of the rarest causes of cord compression syndrome are arachnoid calcifications and intraspinal extradural lipoma both of which appeared simultaneously in a presented child. The originality of this case lies in a presentation of diagnostic difficulties caused by double pathology leading to the cord compression syndrome as well as in the possibilities of successful surgical treatment and rehabilitation.

Case report

History and physical examination

A 14–year–old boy was admitted to our department due to progressive spastic paraparesis of lower limbs, increasing motion difficulties and neurogenic bladder. The first abnormal symptoms, namely, decreased mobility and clumsiness of movements have been visible since the early childhood. For a few recent years gait disorders and significant kyphoscoliosis appeared and gradually deteriorated. Within the last 3–4 months a significant progressive lower limbs paraparesis occurred. Dysfunction of the urethral bladder and radicular pain in thoracic region have been reported by the patient.

Physical examination on admission revealed unstable gait. Walking on toes was difficult even with support. Walking on heels was impossible. Paraparesis of lower limbs was spastic with increased knee jerk reflexes with tendency to policlonus. Plantar response upwards and clonus were found bilaterally. Muscle atrophy of hypothenar eminence and positional tremor of thigh muscles were noticed. Abdominal skin reflexes were absent. Sensation was decreased below the level Th6.

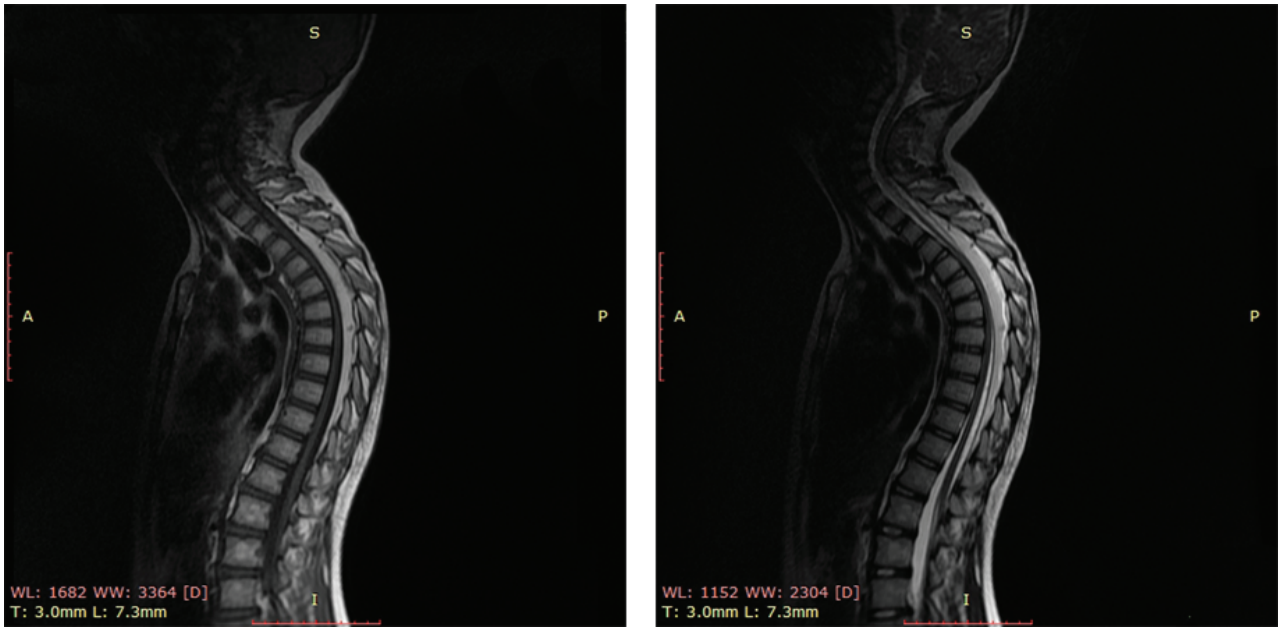

Fig. 1: Thoracic spine MRI sagittal plane in T1(left) and T2 (right) – weighted images showing exacerbation of physiological kyphosis. A lesion (most probably a lipoma) measuring 17,2 centimetres in length with maximal thickness of about 11 millimetres is noted at the dorsal part of vertebral canal. The lesion dislocates the dural sac and spinal cord forward. The cord is pressed against the anterior wall of the spinal canal in the Th3–Th6 region.

Tests and neuroimaging

Basic lab tests were within normal limits with the exception of slightly increased lactic acid level (2.57 mmol/l; norm: 0.6–2.1 mmol/l). Ophthalmological exam revealed hypermetropia. The electroneurography (ENG) showed decreased amplitude during stimulation of the sensory and motor fibers. Electromyography (EMG) was correct. Test for Lyme disease was negative. A result of spectrometry was not indicative of any congenital metabolic disorder. Thoracic spine magnetic resonance imaging (MRI) showed a large lesion reaching from Th2 to Th9, probably a lipoma. Oedema observed around the mass was probably due to blood or lymphatic stasis. There was no sign of anchoring of the cord. A signal of conus medullaris, dural sack and the structure of the cauda equina were normal. Results are presented in Fig. 1 and 2.

Surgery

Due to the dramatic progression of the spastic paraparesis, which made walking impossible, the child was transferred to the neurosurgical unit. The patient underwent Th2–Th9 laminectomy. An extradural lipoma was excised. During the procedure dural sac appeared to be thickened and tightened the spinal cord. Samples of excised tissue were send to histopathological laboratory. After decompression of the spinal cord, duroplasty with the use of dura mater substitute and Tachosil was performed. Postoperative course was uncomplicated. Spine stability was secured with a corset.

Final diagnosis

Histopathological examination revealed intraspinal extradural lipoma and arachnoid calcifications. Both of these conditions were diagnosed to be a cause of cord compressing syndrome in the presented patient.

Results of treatment

Patient's condition improved rapidly after the surgery. In spite of remaining paraparesis, the gait became independent. However, slight problems with heel walking still persisted. Plantar response was downwards bilaterally, the clonus was absent. The bladder regained its function. Kyphoscoliosis and muscle atrophy of hypothenar eminence and palms were visible as on admission. A slight paresis of 4th and 5th toe of the left foot occurred together with difficulties in lifting of outer edge of the left foot. Patient was transferred to the rehabilitation unit.

Follow–up

Currently the patient has slight paraparesis of lower limbs. Walking on longer distances and walking up the stairs are possible. Contractures are absent. Sphincters are under control.

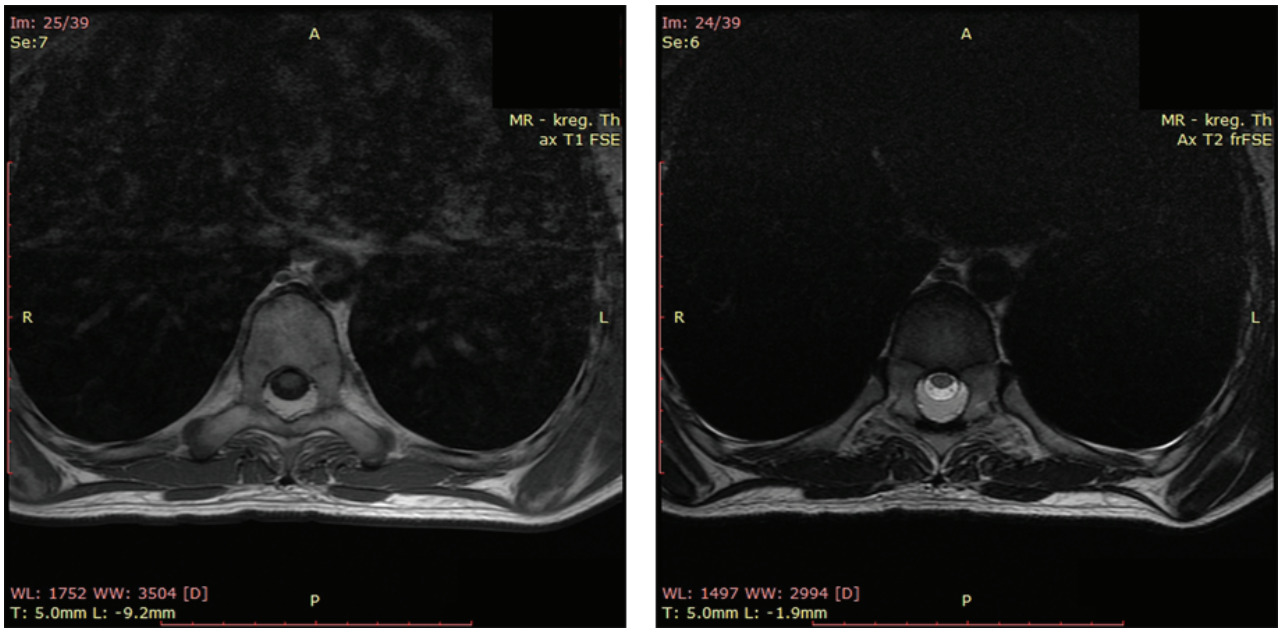

Fig. 2: Thoracic spine MRI axial plane in T1(left) and T2 (right) - weighted images. A lesion (most probably a lipoma) in the vertebral canal is seen. Fluid reserve is reduced. The lesion dislocates the dural sac and spinal cord forward. The cord is compressed.

Discussion

The occurrence of extradural lipomas without associated spinal dysraphism is very rare, accounting for 0.4–0.8% of all intraspinal tumors. These lipomas tend to grow slowly and almost imperceptibly over months or years and most typically cause progressive spastic paraparesis [1]. The same course was observed in our patient, whose walking difficulties were increasing over a period of several years. However, what remained unclear was a sudden deterioration of preexisting neurological deficits with new sensation defects and neurogenic bladder. MRI revealing only a lipoma, which seemed to compress the dural sac, what was not a convincing explanation of that surprisingly rapid progression.

Intraoperative discovery namely spinal arachnoid calcifications were an unexpected finding. Firstly, because of the fact that they were not visible in MRI, which is the first–line imaging for spinal disorders. Computed tomography (CT) was not performed in our patient. Secondly, because they are acknowledged as a rare and unpredictable cause of spinal canal stenosis and neurological deficits [2].

Spinal arachnoid calcification is the accumulation of calcium salts. It appears preferably in older population and is believed to be attributed to the previous injury such as: subarachnoid hemorrhage, meningitis, spinal surgery, trauma or spinal anesthesia [3], none of which was reported in our patient. Asymptomatic, diffuse calcifications may be caused by meningothelial degeneration, but they may occur in small foci of meningothelial proliferation as well. According to Wijdicks and Williams however [4], in most cases the pathogenesis remains unknown. It is true for the presented boy too, as his past medical history was insignificant and none of the performed tests including connective tissue disorder testing gave any explanation.

Increasing numbers of arachnoid cells are found in in the thoracic area. It is a probable explanation to the question why thoracic spine is the most commonly reported location of the described lesions. Also in our patient calcifications in the thoracic spine were noticed. In accordance to literature, calcifica-

tions in this region are usually described as cylindrical (surrounding the spinal cord) which was true for the presented boy as well [2]. His symptoms reflected typical signs of thoracic disease including back pain, regional and radiating pain, leg weakness and gait disorders, abnormal sensation feeling including tingling, burning and increased muscle tone in the lower extremities with associated excessive deep tendon reflexes. The severity of neurological deficits is believed to depend on the size, location, and morphology of the calcifications [5, 6].

As Toribatake et al. [6] claim, decompressive surgery may result in long–term improvement of neurological symptoms, which happened in the case of our patient as well. Complications associated with the procedure include neurogenic bowel or bladder dysfunction. Neurogenic bladder was present in our patients before the surgery, but there were no signs of neurogenic bowel in the postoperative course.

The question is, if calcifications were present in our patient at birth. It seems less possible, according to results of the study performed by Hassler [7], who looked for arachnoid calcifications in cadavers. The author examined three newborns and none of them presented with aforementioned pathology. On the other hand, calcifications compressing the cord since early childhood, would fully explain clumsiness and decreased motor functions observed in our patient for years. What should be considered, is a growing lipoma which could have accelerated the natural course of underlying disease.

Another possible explanation of our patient's history, is that the slowly growing lipoma was the initial pathology causing aforementioned gradually progressing symptoms. Arachnoid calcifications which occurred later on triggered acute compression cord syndrome. Typically they would remain clinically silent [2], but because of a lesion already present in the spinal canal, there was no longer any physiological reserve so the symptoms emerged.

After two surgeries and intensive physical therapy, our patient made an excellent recovery as in case described by Bagley et al. [8]. Although the reports of postoperative improvement in patients with cord compression syndrome caused by calcifications [8] alternate with papers describing significant neurological deterioration afterwards [9], the prognosis for our patient is good.

Conclusions

Co–existance of various neurological disorders leading to the cord compression syndrome in children makes the diagnostic process challenging. Spinal arachnoid calcifications should always be considered, even if MRI is negative. Early diagnosis leads to surgical treatment which gives satisfactory results.

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Wkład autorów/authors' contribution: Ewelina Kuźniar – analiza przypadku, redakcja artykułu, zebranie bibliografii; Agnieszka Stanuszek – analiza przypadku, redakcja artykułu, zebranie bibliografii; Aleksandra Gergont – analiza przypadku, redakcja artykułu, korekta artykułu; Marek Kaciński – analiza przypadku, korekta artykułu

Komentarz:

Jest to poprawnie napisany artykuł w którym autorzy dzielą się swoim doświadczeniem w leczeniu dość rzadko występujących tłuszczaków zewnątrzoponowych kanału kręgowego, które nie towarzyszą wadom dysraficznym kręgosłupa. Ciekawą jest dyskusja na temat stwierdzonych zwapnień wewnątrzoponowych, których obecność należy uznać jako zjawisko towarzyszące podstawowemu procesowi chorobowemu a nie jako niezależna patologia. Niewątpliwie zarówno guz jak i zwapnienia miały bezpośredni wpływ na postępujące uszkodzenie rdzenia kręgowego. Opisanie przypadku pokazuje jak powinna być przeprowadzona diagnostyka a następnie leczenie operacyjne.

prof. dr hab. Włodzimierz Jarmundowicz