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A 16-year-old boy with severe lumbar pain

Szesnastoletni chłopiec z silnymi bólami odcinka lędźwiowego kręgosłupa

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Abstract

A 16-year-old boy presented with acute lumbar pain and headache, and was hospitalised at a paediatric hospital. The symptoms appeared three days before his admission, the lumbar pain was strong and movement exacerbated it. On admission the patient had nuchal rigidity, bilaterally positive Kernig's sign and pain of the L4–S1 spinous processes. Blood tests revealed leukocytosis and positive IgM and negative IgG antibodies against *Borrelia spp*. Cerebrospinal fluid was xanthochromic with a high cell count. Culture of cerebrospinal fluid was negative. Magnetic resonance imaging of the lumbar spine revealed an oval tumour at the level of the L3 vertebra. The tumour was excised completely. Histopathological examination revealed myxopapillary ependymoma of the filum terminale. **Conclusion:** A sudden and unexplained pain of the lumbar spine appearing in children is an indication for in-depth diagnostic investigation, involving in particular magnetic resonance imaging, which allows one to visualise many pathologies, including spinal canal tumours.

Keywords: laminectomy, spinal tumour, neuroborreliosis, radiotherapy, paediatric neuro-oncology

Streszczenie

W pracy przedstawiono opis przypadku 16-letniego chłopca z silnym bólem okolicy lędźwiowej, nasilającym się przy pochylaniu i chodzeniu. Przy przyjęciu do szpitala stwierdzono sztywność karku i obustronnie dodatni objaw Kerniga oraz bolesność palpacyjną wyrostków kolczystych kręgów L4–S1. Badania krwi z odchyleń ujawniły leukocytozę oraz dodatnie przeciwciała IgM przy ujemnych przeciwciałach IgG przeciwko *Borrelia spp*. Wykonano punkcję lędźwiową, uzyskując ksantochromiczny płyn z wysoką cytozą, sugerujący ropne zapalenie opon mózgowo-rdzeniowych, jednak posiew wykluczył zakażenie bakteryjne. Badanie rezonansu magnetycznego kręgosłupa lędźwiowego uwidoczniło zmianę w kanale kręgowym na wysokości kręgu L3, którą usunięto operacyjnie. Histopatologicznie potwierdzono rozpoznanie wyściółczaka śluzowo-brodawkowatego nici końcowej, rzadkiego nowotworu dzieci, który może dawać objawy radikulopatii lędźwiowej, do paraparezy kończyn dolnych włącznie. **Wnioski:** Nagły, niewyjaśniony ból kręgosłupa lędźwiowego u dzieci jest wskazaniem do diagnostyki obrazowej, w szczególności rezonansu magnetycznego, pozwalającego uwidocznić wiele patologii, w tym guzy kanału rdzenia kręgowego.

Słowa kluczowe: laminektomia, guz rdzenia kręgowego, neuroborelioza, radioterapia, neuroonkologia dziecięca

CASE REPORT

A 16-year-old boy reported with his parents to the admissions department of the Independent Public Children's Hospital in Warsaw in June 2018 due to severe lumbar pain and temporal headache lasting 3 days. During interview the boy reported that the spinal pain appeared in the evening after a few minutes' walk and sitting on concrete. The patient described the pain as severe, radiating towards the legs and exacerbating during walking and bending down. The pain rendered everyday activities impossible. Past medical history, including chronic diseases, medication taken, allergies, family history and vaccinations was not revealing. In September 2017 the patient was treated in hospital for aseptic meningitis, the aetiology of which was not determined at the time.

The general condition of the patient on admission was rated as medium. The boy was suffering, but was able to fully engage in meaningful communication. Vital signs (temperature, heart rate, respiratory rate and capillary refill time) were normal. The boy's blood pressure was 125/65 mm Hg. A remarkable observation was that the patient assumed

a forced position with spinal hyperextension (opisthotonus). On physical examination, apart from low-grade fever, no significant abnormalities were found in the skin, pharynx, ears and respiratory, gastrointestinal, genitourinary and cardiovascular systems. Neurological examination revealed nuchal rigidity, bilaterally positive Kernig's sign and tenderness on palpation of the L4–S1 spinous processes. Tendon reflexes were preserved, but were slightly asymmetrical, stronger on the right. Laboratory tests revealed a normal complete blood count [haemoglobin (Hgb) 15.3 g/dL, haematocrit (HCT) 44.1%, red blood cells (RBC) 5.16×10^{12}/L, white blood cells (WBC) 11.06×10^{9}/L, platelets 228×10^{9}/L]. On the day of admission a computed tomography (CT) scan of the lumbar and sacral spine without contrast was performed. The scan did not reveal any abnormalities. During further hospitalisation an enzyme-linked immunosorbent assay (ELISA) was performed in order to check for Lyme disease; both IgM and IgG antibodies were found to be positive. A Western blot confirmation test was also conducted, which indicated early infection (positive for IgM antibodies, negative for IgG antibodies). As part of a diagnostic workup for neuroinfection, lumbar puncture was performed and xanthochromic fluid was obtained (Fig. 1) with a high cell count ($1{,}153 \times 10^{6}$ cells/L), increased level of lactic acid (5.7 mmol/L) and protein (250 mg/dL) and decreased glucose concentration (<0.55 mmol/L); the fluid's presentation was consistent with pyogenic meningitis. Cerebrospinal fluid culture was negative; however, antibodies against *Borrelia* spp. were detected (ELISA positive for IgM and IgG). Based on nerve root symptoms, inflammatory changes in the cerebrospinal fluid and positive *Borrelia* spp. infection tests, neuroborreliosis was suspected and antibiotic therapy was started (ceftriaxone). Due to significant diagnostic doubts, a CT scan was performed the next day, which revealed only a site of bone loss in the sacral bone at the level of S1, interpreted as a developmental variant. Subsequently, an magnetic resonance imaging (MRI) scan of the lumbar spine without contrast revealed an oval mass of $29 \times 19 \times 11$ mm in the spinal canal lumen at the level of L3. The mass filled the whole spinal canal lumen and displaced spinal roots posteriorly. FLAIR MRI of the head revealed only fine areas of elevated signal, which were probably post-traumatic. Based on consultations with a neurologist and a neurosurgeon a decision was made to treat the proliferative mass of the spinal canal surgically.

On day 6 after admission complete laminectomy was performed on L3 and partial laminectomy on L2 and L4 with complete excision of an intradural cauda equina tumour. No macroscopic spread of the neoplasm was found during the operation. Histopathological examination of the material removed during surgery revealed World Health Organization G1 myxopapillary ependymoma (MPE) with massive extravasation. During the postoperative period clinical improvement was observed: the patient started to move normally with no paralysis, including no micturition disorders.

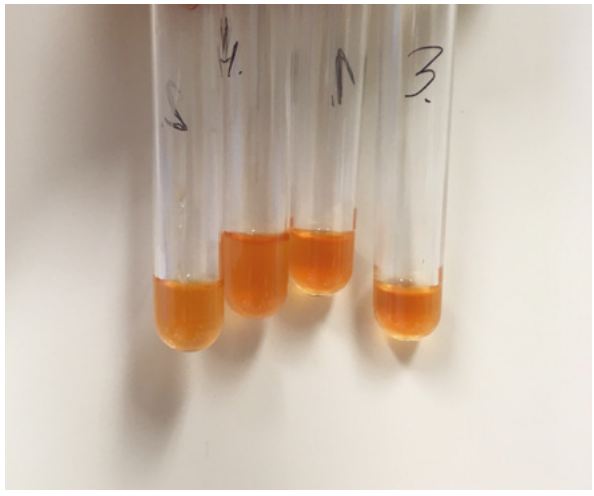

Fig. 1. Xanthochromic cerebrospinal fluid collected from the patient

Due to the possibility of primary fluid spread, the boy was transferred to the care of an oncology clinic.

DISCUSSION

Myxopapillary ependymoma of the filum terminale of the spinal cord is an extremely rare neoplasm in children; it is most common in young adults and is usually diagnosed between 35 and 37 years of age. The most common locations for MPE are filum terminale of the spinal cord and cauda equina. The tumour presents with non-specific lumbar or sacral pain; it can also produce symptoms of lumbar radiculopathy. In advanced cases, paraparesis may occur. Apart from lumbar pain, the reported patient had positive meningeal signs, which are not characteristic for the clinical presentation of this type of neoplasm. Despite that, thanks to careful examination of the patient and analysis of the clinical presentation and the results of additional tests, the diagnosis was made already on day 3 of hospitalisation and causal treatment was applied on day 6. Such swift action made it possible to avoid permanent neurological damage and remove the tumour completely with no macroscopic spread. What was also important was the fact that the patient reported to hospital early due to a sudden onset of acute pain. Such a beginning of disease is characteristic for the paediatric population: due to a narrower spinal canal and faster tumour development, the period of prodromal symptoms is short and a sudden onset of acute symptoms is typical. As a result, MPE diagnosis is determined quicker in children than in adults, in whom the disease is diagnosed at later stages due to slowly developing symptoms[1,2].

In the present case, justified doubts regarding the diagnosis of neuroborreliosis were key to determining the actual disease: a xanthochromic cerebrospinal fluid, which indicated past bleeding, and a high pleocytosis with elevated protein concentration, which was consistent with pyogenic

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meningitis, did not correspond with the cerebrospinal fluid presentation typical for neuroborreliosis, in which pleocytosis is usually between a few dozen to a few hundred cells and mononuclear cells clearly dominate the microscopic image.

A similar inconsistency in the cerebrospinal fluid presentation was described in the literature for a 33-year-old patient with similar cerebrospinal fluid parameters, in whom subarachnoid haemorrhage was suspected. After the verification of the preliminary diagnosis, a spinal canal tumour at L1–L2 was detected[3]. Therefore, one needs to bear in mind that xanthochromic cerebrospinal fluid may be a sign of not only meningeal vessel damage, but also of a spinal canal tumour.

Although CT imaging did not reveal any abnormalities, severe pain and clear focal neurological symptoms indicated a possible lumbar lesion. The only element that could have accelerated the diagnosis and shorten the time of diagnostic workup for infection as the suspected cause of the patient's complaints was MRI of the spine performed first, instead of a CT scan. However, MRI, despite its numerous assets, is a poorly available procedure, especially for unscheduled admissions; in addition, CT allows one to detect fresh bleeding and as such, it usually turns out to be sufficient during preliminary diagnostic investigation. However, it is important to remember that normal CT findings do not exclude the presence of focal lesions in the spinal canal.

The final diagnosis accounted for the atypical cerebrospinal fluid presentation. The massive extravasation into the tumour found on histopathological examination explains the xanthochromic cerebrospinal fluid and the pyogenic meningitis presentation on microscopic smear that is inconsistent with the negative culture. Due to the lack of clinical symptoms, a positive IgM antibodies test for Lyme disease alone was not sufficient to diagnose it. IgM antibodies often produce false positive results. Full recovery following surgical treatment indicates a lack of any additional causes of the patient's clinical condition. An interesting element of the patient's medical history is viral meningitis a year before: it is possible that it could have been the first sign of the neoplasm already developing at the time. Changes in the cerebrospinal fluid could have been the effect of an inflammatory reaction accompanying the tumour, especially since the aetiological factor was not identified.

The treatment of choice for MPE in children is complete excision of the tumour. Despite the fact that MPE is usually a benign neoplasm, it is characterised by frequent recurrences (30%)[4], especially in individuals below 35 years of age, in whom the risk of relapse is increased[5]. Apart from age, risk factors for MPE recurrence include incomplete tumour excision and atypical cells found on histopathological examination of the tumour. Research is underway on the optimal treatment of MPE to reduce the risk of recurrence. Currently, despite the proven efficacy of postoperative radiotherapy, it is not routinely used in children because of too many complications occurring in this age group[6–9].

The indications for postoperative radiotherapy in children are incomplete tumour excision and a multifocal type of MPE.

In conclusion, in the case of unexplained and persistent lumbar pain in children, one should remain vigilant and perform MRI[10] in order to exclude spinal canal tumours that may not be evident on CT.

Due to a high recurrence rate of MPE, frequent follow-up visits and diagnostic imaging procedures are very important in postoperative care. Such management allows one to detect possible disease recurrence early and treat it effectively.

Conflict of interest

The authors do not report any financial or personal affiliations to persons or organisations that could adversely affect the content of or claim to have rights to this publication.

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