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Case report

Acute opercular syndrome in 4-year-old boy

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ABSTRACT

Acute opercular syndrome is a rare described syndrome caused by a sudden damage of the cerebral cortex and subcortical white matter, located around the insula. A rare cause of this syndrome can be an infectious agent, particularly herpes simplex virus. Quick diagnosis and immediate initiation of treatment significantly reduce the risk of neurological consequences and mortality. We present a case of encephalitis of unknown etiology and severe course, with the symptoms of acute opercular syndrome in 4-year-old boy.

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1. Introduction

The incidence of encephalitis in developed countries is estimated about 2.2–7.4 cases/100,000 persons/year, in pediatric population is higher 10.5–13.8 cases/100,000 [6]. It can be caused by infections (especially viral, but also bacterial, parasitic, fungal), immunological response (acute disseminated encephalomyelitis, acute hemorrhagic leukoencephalopathy, acute necrotizing encephalitis) and factors not related to infection: vascular, cancer, metabolic, toxic, drug-induced. Herpes simplex virus (HSV) is the most common infectious agent causing encephalitis. Herpes simplex encephalitis (HSE) in 90% of cases is caused by type 1 HSV, in 10% by type 2. This is primary infection in 30%, recurrent in 70%. The most common symptoms are: fever, seizures, focal neurological symptoms, personality changes, vomiting [3,7].

Acute opercular syndrome (facio-labio-glosso-pharyngo-laryngo-brachial paralysis) is a rare described syndrome caused by usually bilateral lesions of the cerebral cortex and subcortical white matter, located around the insula. The most common causes of this syndrome are incidents of ischemic stroke (thrombotic, embolic, defect of blood vessel), developmental abnormalities (brain tissue dysplasia), infectious agents, especially HSE, multiple sclerosis, head injury, tumor. The typical symptoms of opercular syndrome, resulting from

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Abbreviations: ADEM, acute disseminated encephalomyelitis; CRP, C-reactive protein; CSF, cerebrospinal fluid; CT, computed tomography; HSE, herpes simplex encephalitis; HSV, herpes simplex virus; IgG, immunoglobulin G; IgM, immunoglobulin M; MRI, magnetic resonance imaging; PCR, polymerase chain reaction.

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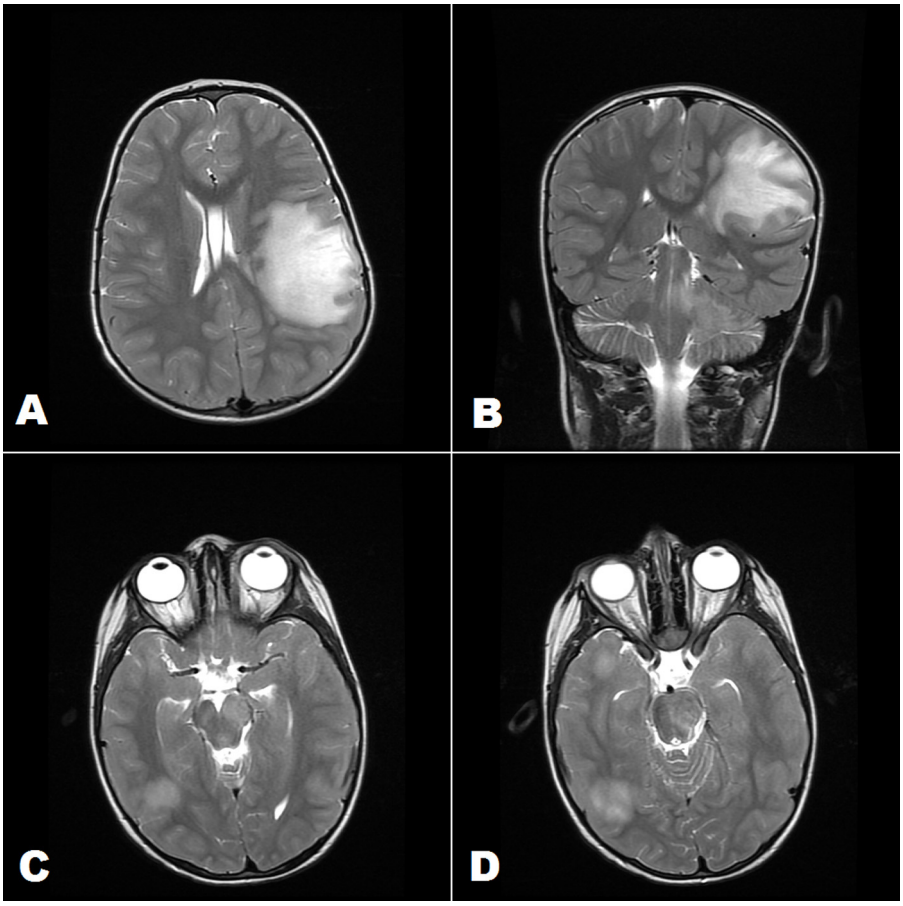

Fig. 1 – (A–D) The first day of hospitalization. (A and B) T2-weighted axial MRI scan, the biggest lesion located on the border of left parietal, temporal and frontal lobe, with edema, mass effect and the shift of central structures of the brain.

damage the frontal and parietal motor tracts, include swallowing disorders, motor aphasia, paralysis of facial muscles and masseter. Differential diagnoses of acute opercular syndrome in children include congenital abnormalities (bilateral opercular polymicrogyria), neurodegenerative disorders, potential causes of bulbar palsy, among other things, myasthenia gravis, botulism, Brown-Vialetto-Van Laere syndrome, as well as the recurring form in patients with epilepsy.

We present a case of encephalitis of severe course and unknown etiology in 4-year-old boy.

2. Case report

4-year-old boy was transferred from a district hospital with suspected meningitis. Previously, he had a fever since 2 days. There was pharyngitis diagnosed, the boy received amoxicillin with clavulanic acid. On the second day of the disease there were found progressive right-sided hemiparesis, speech disorders and abnormal eye movements. Lumbar puncture was performed, but the result of cerebrospinal fluid (CSF) analysis was not diagnostic (technical error), fluid culture – negative. After 6 h, due to the rapid deterioration of the general condition, the child was transferred to our hospital. On admission, patient's general condition was assessment as severe, with a disturbance of consciousness, deep right-sided hemiparesis, abnormal eye movements, bradycardia 50–60 beats/min, the presence of meningeal signs (nuchal rigidity, Kernig's sign, Brudzinski's neck sign). Blood tests were normal, except from a slightly elevated CRP (7 mg/l). Because of the severe general condition of the child, unclear clinical picture and the presence of focal neurological symptoms, the performance of lumbar puncture was temporarily postponed, there was given acyclovir (500 mg/m^2 body surface area/8 h), mannitol (0.25 g/kg/8 h), dexamethasone (0.6 mg/kg/24 h). Cerebral magnetic resonance imaging (MRI) demonstrated the presence of extensive, multifocal lesions in both hemispheres of the brain, spinal cord and cerebellum, with increased signal intensity on T2-weighted images, with edema and mass effect accompanying the biggest lesion located on the border of left parietal, temporal and frontal lobe, with the displacement of central structures of the brain (encephalitis? ADEM? gliomatosis?) [Fig. 1]. The imaging of the chest and abdomen showed no pathological changes. There were conducted multidisciplinary medical consultations, including neurological, neurosurgical, oncological. In the differential diagnosis there were considered infectious, neoplastic, demyelinating and vascular causes of changes. It was found that an infectious etiology of lesions is most likely. There were continued intravenous acyclovir (500 mg/m^2 of body surface

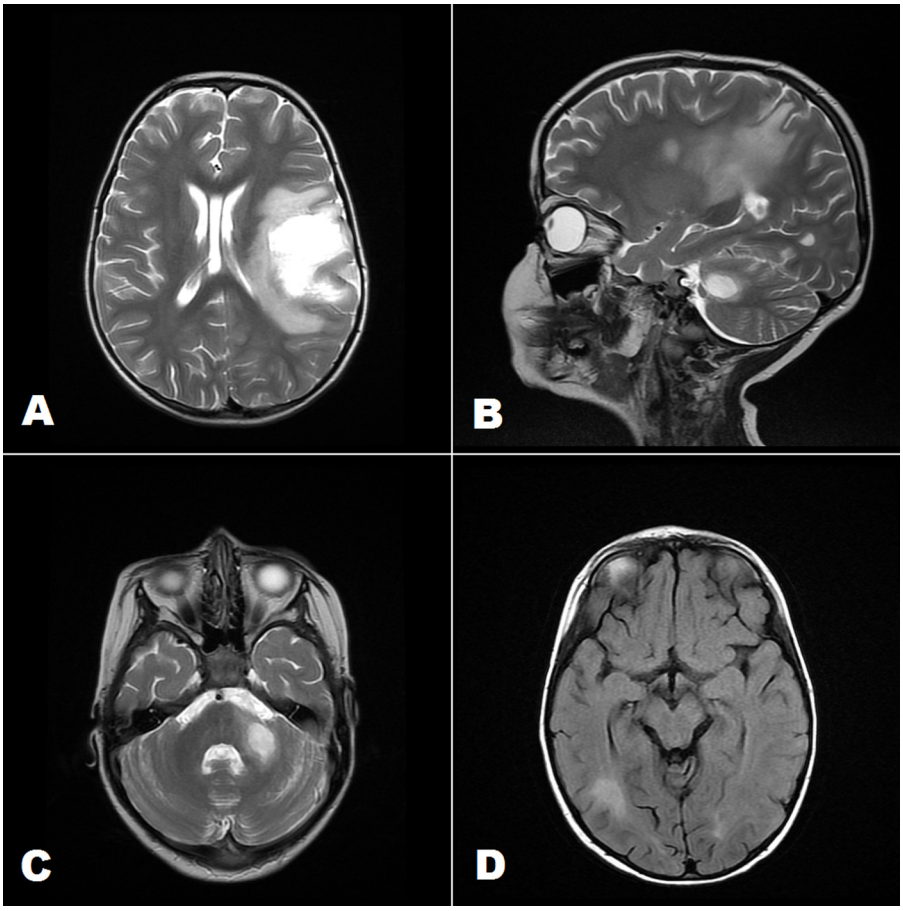

Fig. 2 – The 8th day of treatment. (A–C) Encephalomalacia in the largest changes.

area/8 h), dexamethasone (0.6 mg/kg/24 h). The boy also received intravenous human immunoglobulin (total dose of 2.0 g/kg).

As a result of treatment the general condition of the boy gradually became better. From the fifth day of treatment the boy was conscious, with a present response to voice and touch, but reduced mimic facial movements, severe aphasia, lack of pharyngeal reflexes, dysphagia resulting in the need to use of a stomach tube, the right-sided hemiparesis and bilateral Babinski's sign were found in neurological examination. We also observed several incidents of subfebrile body temperature, in the absence of clinical and laboratory signs of infection. The control MRI (including magnetic resonance spectroscopy) performed on the 8th day of treatment demonstrated high dynamics of earlier described lesions, with the signs of encephalomalacia in the largest changes [Fig. 2]. The smaller lesions showed weaker signal intensity compared to the first examination. The displacement of central structures of the brain was still present. The nature of the changes was still unclear, however there was no characteristic of proliferative lesions and acute demyelination contrast enhancement. After the improvement of the general condition of the child and disappearance of edema and mass effect on MRI, lumbar puncture was performed. The result of CSF analysis was normal. Additional blood and cerebrospinal fluid tests (microbiological, serological and molecular) did not give a confirmation of the presence of an infectious agent (including HSV) as a potential etiological agent of encephalitis. Treatment with intravenously acyclovir was continued for 28 days, the dose of dexamethasone was gradually reduced until discontinuation of treatment in 21st day of treatment. The boy had a speech therapy and was rehabilitated physically. Gradual improvement of general condition and significant reduction in the severity of neurological symptoms was achieved. Boy can walk without support, with discreet right-handed hemiparesis, he has not imbalances. Swallowing disorders withdrawn completely, while a slight speech disorder persisted – a boy speaks vague a bit and has difficulty pronouncing some words. In control MRI, performed 9 weeks and then 6 months after starting treatment, a marked regression of changes was found – reducing the size of the largest lesions and disappearance or significant reduction in signal intensity of the other [Fig. 3].

3. Discussion

Etiology of encephalitis in this patient is unknown. According to the guidelines of the Association of British Neurologists and British Paediatric Allergy, Immunology and Infection Group, patients with suspected encephalitis should have immediately performed a lumbar puncture. In the case of presence of

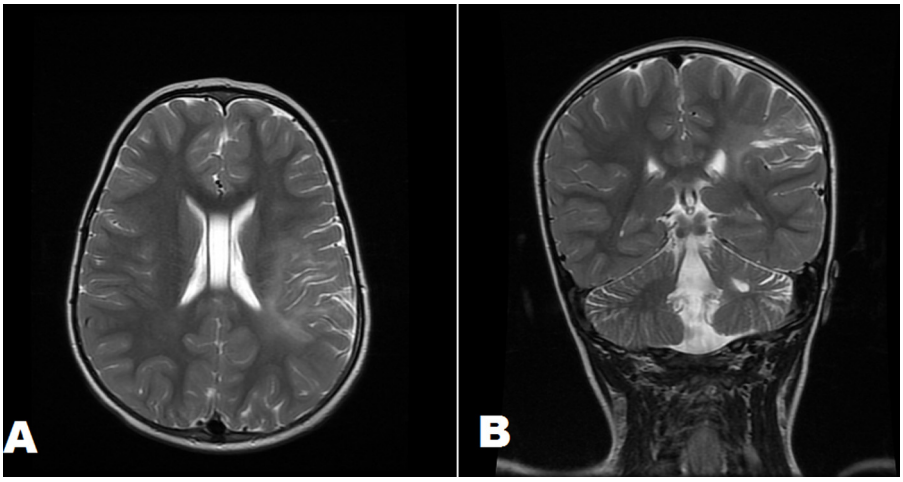

Fig. 3 – Control MRI performed 6 months after starting treatment. T2-weighted axial MRI scan, a marked regression of changes.

clinical contraindications to puncture it is urgent to perform CT or MRI of the brain and lumbar puncture perform immediately after the exclusion of radiological contraindications [7]. CSF PCR test for HSV is considered to be the gold standard in the diagnosis of HSE. The sensitivity of this test in immunocompetent adults is estimated at 96%, specificity 99% [2,7]. The test result may be negative in the first days and above than tenth day of the disease [2,4,7,10]. The detection of HSV-specific IgG antibodies in CSF may be helpful in the diagnosis, especially when the test is repeating after 10–14 days. The presence of HSV-specific IgM antibodies, fourfold increase in CSF HSV-IgG level with an interval of 10–14 days and a higher level of IgG in the CSF than in blood serum is considered to be diagnostic. In the case of negative test results diagnosis of HSE is questionable, however, in the case of early initiation of antiviral therapy or in patients with immunodeficiency intrathecal synthesis of HSV-specific IgG may be reduced or absent. It seems that in described case HSV encephalitis cannot be excluded, because of a late performance CSF tests, resulting from pre-existing contraindications to lumbar puncture (clinical and radiological), as well as early initiation of treatment. In addition, the sensitivity of diagnostic methods (CSF PCR for HSV and CSF HSV-IgG) in children is 70–76%, differently to 96% in adults [2–4]. The MRI images of the brain seem to confirm the diagnosis of HSE. Typical changes in the early phase of HSE include hypodense lesions in low-medial parts of the temporal lobe, orbitofrontal cortex and limbic structures (including the hippocampus), with increased signal intensity on T2 weighted images, and in the later period of the disease – necro-hemorrhagic changes [4,9]. However, there are reports of unusual location of lesions in the course of HSV-encephalitis, especially in children – increasingly in frontal and parietal lobe and turn the insula [3,4]. In described case, initially extensive, multifocal hypodense changes with increased signal intensity on T2-weighted images were found, subsequently – necrotic. Quick improvement of general condition after initiating antiviral therapy and almost complete disappearance of neurological symptoms also seem to confirm the infectious etiology of the disease. ADEM is the second diagnosis that should be taken into account. However, there was also no characteristic of acute demyelination contrast enhancement in MRI. The boy received intravenous human immunoglobulin and glucocorticoids, but the type of drug and its dosage did not conform to those used in acute demyelination, though the condition of the boy underwent a rapid improvement.

There was no description of acute opercular syndrome in the course of HSE in child in Poland; there are a few reports in the international literature. Kocak et al. and De Kleermaeker et al. described the cases of children who have experienced acute opercular syndrome in the course of HSE, confirmed by positive result of CSF PCR for HSV and changes in MRI [1,8]. García-Ribes et al. described four children with suspected HSE, with symptoms of acute opercular syndrome, with the typical changes in the MRI, but only one child had positive result of CSF PCR for HSV [5]. In the case of our patient HSE was not confirmed, however, the result of cerebral MRI, clinical course of the disease and quick improvement after initiating antiviral therapy make this diagnosis possible.

Acute opercular syndrome in the course of HSV-encephalitis in children is very rarely recognized. Due to its severe course and serious clinical consequences it seems to be very important considering this syndrome in the differential diagnosis of encephalitis in children. Quick diagnosis and immediate initiation of treatment significantly reduce the risk of neurological consequences and mortality.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

REFERENCES

[1] De Kleermaeker FG, Bouwmans AE, Nicolai J, Klinkenberg S. Anterior opercular syndrome as a first presentation of herpes simplex encephalitis. J Child Neurol 2013.

[2] De Tiège X, Heron B, Lebon P, Ponsot G, Rozenberg F. Limits of early diagnosis of herpes simplex encephalitis in children: a retrospective study of 38 cases. Clin Infect Dis 2003;36(10):1335–9.

[3] De Tiège X, Rozenberg F, Héron B. The spectrum of herpes simplex encephalitis in children. Eur J Paediatr Neurol 2008;12(March (2)):72–81.

[4] Elbers JM, Bitnun A, Richardson SE, Ford-Jones EL, Tellier R, Wald RM, et al. A 12-year prospective study of childhood herpes simplex encephalitis: is there a broader spectrum of disease? Pediatrics 2007;119(February (2)):e399–407.

[5] García-Ribes A, Martínez-González MJ, Prats-Viñas JM. Suspected herpes encephalitis and opercular syndrome in childhood. Pediatr Neurol 2007;36:202–6.

[6] Jmor F, Emsley HCA, Fischer M, Solomon T, Lewthwaite P. The incidence of acute encephalitis syndrome in Western industrialized and tropical countries. Virol J 2008;5:134.

[7] Kneen R, Michael BD, Menson E, Mehta B, Easton A, Hemingway C, et al. National Encephalitis Guidelines Development and Stakeholder Groups. Management of suspected viral encephalitis in children – Association of British Neurologists and British Paediatric Allergy, Immunology and Infection Group national guidelines. J Infect 2012;64(May (5)):449–77.

[8] Kocak O, Yarar C, Yakut A, Ekici A, Yimenicioglu S, Saylisoy S. Akathisia in association with herpes simplex encephalitis relapse and opercular syndrome in children. Brain Dev 2014;36(February (2)):167–70.

[9] Tunkel AR, Glaser CA, Bloch KC, Sejvar JJ, Marra CM, Roos KL, et al. The management of encephalitis: clinical practice guidelines by the Infectious Diseases Society of America. Clin Infect Dis 2008;47:303–27.

[10] Weil AA, Glaser CA, Amad Z, Forghani B. Patients with suspected herpes simplex encephalitis: rethinking an initial negative polymerase chain reaction result. Clin Infect Dis 2002;34:1154–7.