

ISSUES IN THE DIAGNOSTIC MANAGEMENT OF SANDIFER'S SYNDROME IN A 3-YEAR-OLD MALE CHILD WITH ADENOTONSILLAR PATHOLOGY: A CLINICAL CASE

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ABSTRACT — Chronic adenoiditis has a high prevalence among children, specifically in preschool-age population and in midchildhood. Chronic adenoiditis often occurs against the background of somatic comorbidities. Severe hypertrophy of pharyngeal and palatine tonsils causes hypoxic ventilatory response and, subsequently, leads to the development of obstructive sleep apnoea syndrome.

KEYWORDS — children, Chronic adenoiditis, obstructive sleep apnoea/hypopnea syndrome, Sandifer's syndrome, interdisciplinary follow-up, management.

INTRODUCTION

In clinical practice, one patient might suffer from several diseases simultaneously. They differ in progression and degrees of impact on each other. Chronic adenoiditis is one of most prevalent diseases in preschool-age population [1]. Chronic intermittent hypoxia against the background of chronic adenoiditis is formed due to the syndrome of sleep apnoea/hypopnea. It is known that respiration impairment during nocturnal sleep is revealed in children of any age.

Prevalence of obstructive sleep apnoea/hypopnea syndrome (OSAHS) in children in overall population ranges from 0.0% to 5.7% [2]. These indices vary according to the age. At the age of two-six years, prevalence of snoring amounts to 10-14% [3], while prevalence of OSAHS reaches 3% [4]. Adenotonsillar pathology in preschool-age children may induce different nervous system pathologies including such impairments as learning disability, behaviour decline, the syndrome of central nervous system (CNS) hyperactivity, the attention deficit and hyperactivity disorder (ADHD), etc. [5]. A rare form of nocturnal hypoxemia against the background of OSAHS is impairment of the extrapyramidal system.

Early diagnostics of cases related to comorbidity between chronic adenoiditis and Sandifer's syndrome is of critical significance for correct diagnosis, symp-



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omatic treatment and disease prevention, taking into account that Sandifer's syndrome represents a multi-disciplinary diagnostic and therapeutic challenge [6,7].

The purpose

of the present study is to present a clinical case of Sandifer's syndrome in a child with adenotonsillar pathology complicated by OSAHS.

A CLINICAL CASE

A male preschool child Fyodor aged 3 was first examined by a neurologist of the Neurological Centre of Epileptology, Neurogenetics and Brain Research of the University Clinic (UCNC) in October 2016. The child suffered from stereotypically proceeding abrupt awakenings during nocturnal sleep with anxiety and crying; disorder of nasal breathing during nocturnal sleep, snoring and nocturnal enuresis; occasionally, dystonic head tilt with version and rotation to the left was registered as well as dystonic muscle tension in extremities and the neck during and after awakening; regular long-lasting release of mucus from nasal cavity. Prenatal period was not burdened.

ANAMNESIS VITAE: the child was born at the gestational age of 40 weeks. At the age of six months, dystonic seizures were first diagnosed in his trunk and limb muscles. An examination to determine their origin was not performed. At the age of one year, seizures became more frequent and the child was hospitalised. Video-EEG-monitoring was conducted which verified the diagnosis of epilepsy. The child underwent antiepileptic therapy for one year. No epileptiform activity was revealed during subsequent video-EEG-monitoring. However, hyperexcitability, excessive salivation, nocturnal snoring dystonic tension in neck and limb muscles remained. That served as the reason to refer the parents to an epileptologist and an otolaryngologist.

NEUROLOGICAL STATUS: the child's state was satisfactory, consciousness was clear, psychomotor and speech development corresponded to age norms. The child was hyperexcitable and impulsive. Attention deficit was observed. Cranial nerves: without lesions. No lesions have been revealed in motor, coordination and sensorial spheres. The patient had no meningeal signs and was continent.

At the otolaryngologic consultation, the patient's mother reported regular long-lasting release of mucus from nasal cavity, frequent episodes of acute respiratory viral infections up to one time per month; a new qualitative sign was occasional manifestation of eructation and musty smelling breath.

OBJECTIVE STATUS: nasal breathing was difficult. Excessive mucus secretion was observed.

Examination by means of a zero-degree inflexible endoscope (endoscope tube diameter equalling 1.7mm) after topical anaesthesia and anaemisation was used. It revealed that the nasal mucosa was cyanochrous, rears of inferior nasal conchae were enlarged, choanal lumens were blocked by 2/3 by hypertrophied tissue of pharyngeal tonsil; pharyngeal openings of auditory tubes were partially blocked by lymphoid tissue. The mouth was open during breathing; high-arched palate was observed; posterior pharyngeal wall mucosa was moderately hyperaemic; lateral pharyngeal bands were enlarged; palatine tonsils have first-degree hypertrophy; tonsillar lacunae were clean. Maximum sizes of posterior mandibular, submandibular, anterior and posterior cervical lymph nodes were 7.0/10.0/5.0 mm (over three groups) on both sides. Lymph nodes were elastic, non-tender and were not matted together.

In order to conduct differential diagnosis of Sandifer's syndrome, focal epilepsy and dyskinesia of non-kinesiogenic origin, the child underwent additional examination. Magnetic resonance imaging (MRI) of the brain (1.5 Tesla) showed that lymphoid tissue widened nasopharynx lumen by 2/3, the nasopharynx dome was blocked. No structural pathology of the brain was revealed; no cerebrospinal fluid dynamics impairment was found. Cardiorespiratory monitoring (for 8 hours) revealed four respiratory anomalies and two episodes of obstructive apnoea with maximum duration of 15 seconds. Two episodes of hypopnoea with maximum duration of 40 seconds were also observed; the apnoea-hypopnoea index (AHI) was 2.0 per hour (the normal value for children is 1 per hour). Desaturation index was 3 episodes per hour. Snoring was registered with the index of 30%. Mean oxygen saturation was 96% with the minimum of 88% which is a prognostic criterion for unfavourable course of the disease. Mean heart rate totalled 85 bpm, with the minimum of 51 bpm and the maximum of 196 bpm.

EPICRISIS: ronchopathy, OSAHS of medium severity; pronounced respiratory sinus arrhythmia; transient nocturnal hypoxemia of medium severity. Nocturnal video-EEG-monitoring shoed that the structure of nocturnal sleep was defragmented; deep stages of slow-wave sleep were not reached, sleeping was of superficial character. Ultrasonography (US) of the stomach with an aqueous-siphon test (intake of 200 ml of water). Trendelenburg positioning for five minutes did not show the presence of gastroesophageal reflux (GR). However, signs of GR complicated by esophagitis with spontaneous reopening of esophageal hiatus were revealed during a fibrogastroduodenoscopy (EGD).

In general, it is noteworthy that onset of increased respiratory disease frequency in absence of confirmed

contacts in this child was registered simultaneously with the period of active motor performance and the onset of reflux manifestations forming neurological symptoms and the risk of aspiration.

On the basis of neurological, otolaryngologic and gastroenterological examination a diagnosis was verified: Sandifer's syndrome with mainly nocturnal abrupt dystonic attacks with involvement of limb, trunk, face and neck muscles; reactive (GR-associated) chronic adenoiditis; II degree hypertrophy of the pharyngeal tonsil, I degree hypertrophy of palatine tonsils; widespread hypertrophy of neck group lymph nodes on both sides of combined (reactive post-inflammatory) genesis.

COMPLICATION: ronchopathy, OSAHS of medium severity, pronounced respiratory sinus arrhythmia; transient nocturnal hypoxemia of medium severity.

A case conference with the participation of a neurologist, an otolaryngologist and a gastroenterologist was held and a collective decision of the otolaryngologist and the neurologist to perform a cold-plasma adenoidectomy as a salvage surgery was rendered.

ONE-YEAR CATAMNESIS: the child is registered with dispensary observation by a neurologist, an otolaryngologist and a gastroenterologist of the UCNC. General state of the child is satisfactory. Nocturnal sleep is normalised. No dystonic attacks or enuresis during sleep are registered. Pronounced syndrome of attention deficit and hyperactivity disorder has decreased significantly, socialisation of the child is not compromised at the moment. Nasal breathing is free. No relapses of neurological, gastroenterological or ENT-pathologies are observed within the framework of the interdisciplinary follow-up. Antiepileptic preparations were withdrawn more than one year ago.

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