CLINICAL CASE

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CLINICAL CASE OF CYSTIC FIBROSIS WITH THE DEVELOPMENT OF CYSTIC FIBROSIS-RELATED DIABETES IN A 14 YEARS OLD BOY

ABSTRACT

The paper presents a clinical case of cystic fibrosis mixed form, severe, combined with cystic fibrosis-related diabetes, in a fourteen-year old boy.

Objective: demonstration of the clinical case of cystic fibrosis, combined with cystic fibrosis-related diabetes, a boy of 14 years.

Materials and methods

The analysis of the cystic fibrosis-related diabetes patient's observations. The patient is the oldest patient among those diagnosed with cystic fibrosis.

Conclusion

Thus, the feature of the case of cystic fibrosis (CF), a boy of 14 years, is a complicated course with chronic Pseudomonas aeruginosa infection and persistent connection cystic fibrosis-related diabetes.

Keywords: cystic fibrosis, North, children.

INTRODUCTION

Cystic fibrosis (CF) (Cystic Fibrosis) - the most common genetic abnormality, caused by a mutation of the gene CFTR (cystic fibrosis transmembrane conductance regulator), causing a violation of transportation of chlorine ions, sodium and bicarbonate in the epithelial cells, which causes progressive damage to the exocrine glands vital organs [1] . CF frequency varies among Europeoids from 1: 600 to 1: 17,000 births. The RF frequency of cystic fibrosis is according FGBU Medical Genetic Research Center 1: 10,000 newborns [2]. According to the Ministry of Health YARMIATS of Sakha (Yakutia) is registered in 13 patients with cystic fibrosis. Cystic fibrosis-related diabetes prevalence varies depending on diagnostic criteria and screening. In Minnesota found 9% of 5-9-year-olds, 26% of 10-20 year olds and 50% of 30 years and above [5]. In recent years, the life expectancy of CF patients has been steadily increasing, at the same time increases the frequency of complications in these patients, such as diabetes mellitus (DM), accounting for a variety of data from 2.5 to 32%. In clinical practice cystic fibrosis-related diabetes (MZSD) is often diagnosed only when the manifestation of carbohydrate metabolism disorders [3]. Diabetes in cystic fibrosis (CFRD - cystic fibrosis-related diabetes) affects approximately 20% of adolescents and 40-50% of adults with cystic fibrosis. Since adolescence, the incidence is 3%, with a certain predominance of the female gender. According to Russian scientists, impaired glucose tolerance (IGT) occurs in 50-75% of adult patients with cystic fibrosis and clinical diabetes mellitus (DM) manifests in 5-15% of cases. In 2006, at the Russian Centre of cystic fibrosis IGT was found in 53% of patients (aged from 2 months to 18 years), and insulin-dependent diabetes (type cystic fibrosis-related diabetes) as manifest forms of endocrine lesions of the pancreas, - 1% of patients [4].

Objective: demonstration of the clinical case of cystic fibrosis, combined with cystic fibrosis-related diabetes, a boy of 14 years.

MATERIALS AND METHODS

The analysis of the cystic fibrosis-related diabetes patient's observations. The patient is the oldest patient among those diagnosed with cystic fibrosis.

THE RESULTS AND DISCUSSION

Child is from the 2nd pregnancy, with toxicosis at the first half, the 2nd delivery. Age of mother during pregnancy 23 years. Self- delivery, in time. He was born with a weight - 2820 growth - 51 cm. Breastfeeding - up to 1 month. Psychomotor development with a lag: the child began to keep a head in 4 months, to sit - from 9 months, to go - from 1 year 9 months. From the first days of life there was frequent (5-6 times a day), mushy stool. From 3 months frequent colds, there is a constant nasal congestion, at 6 and 9 months, he suffered pneumonia. From the day of birth body mass gain is poor. In 2002, cystic fibrosis was diagnosed. He was examined in the Center of cystic fibrosis (Moscow) in 2003, the diagnosis was confirmed. Basic therapy: Creon 75 thousand units x 4 times a day, while snacking addition to 400 thousand units per day, Pulmozyme 2.5 mL x 2 times daily. The patient is a carrier of a chronic Pseudomonas aeruginosa infection; 4 times a year is admitted to the Department of Pediatric Pulmonology center RBN№1 - NCM routinely for antimicrobial therapy. Consistently receives Pulmozyme, inhaled colistin. Last admission from 4 to 18 July 2016.

Complaints on admission: cough with muco-purulent sputum and wheezing, nasal stuffiness.

When having an objective medical examination his height was 162 cm, the body mass was 41 kg. The condition of the patient was very severe and he had a hyposthenic physique. The cutaneous integument and mucous membrane had a natural colour, moderate humidity. The peripheral lymph nodes were multiple, matted and painless, the size is up to 0.8 cm. The fingers were like «drumsticks», the nail coatings were like «hour glasses». The shape of the chest was correct. The frequency of respiratory movements was 28 in a minute and there was a mixed pattern of breathing. The percussion sound on the lungs was bandbox. When auscultation in lungs, the breathing was tough, mosaically weakened, carried out in all area, humid small- and medium bubbly sounds on both sides, more on the right. There was dry humming wheezing when

forced expiration. The nasal breath is moderately complicated, the discharge was mucous. Heart rate was 116 in 1 min. The tones were clear, rhythmical. The tongue was moist laid over by a whitish-yellow coating.

The stomach was painless. The liver performed at 1 cm from under the costal arch, the pancreas was not palpable. The boy urinated freely. There was a formed stool twice a day.

Here are data of laboratory instrumental studies: monocytosis (14,3%)- general blood test dated from 12.01.16, monocytosis remained on the 21.01.16 (12,6%) , subcompensated respiratory acidosis (pH 7,279, pCO2 52,4, pO2 32,1, sO2 51,9, FCOHb 0,3, FO2Hb 51,3, FMetHb 0,8, cBase -2,0 mmol/L, cHCO3 19,8 mmol/L, K+ 4,4 mmol/L, Na+ 137 mmol/L, Ca2+ 0,64 mmol/L, Cl 105 mmol/L) was in the parameters of the venous blood acid-basic state dated from 12.01.16 . A urine screen test was within normal limits dated from 21.01.16: 80.0, the colour was light yellow, transparent, specific gravity was 1025, urine was acidic, no protein, sugar +++, acetone was negative. Biochemical blood test dated from 12.01.16 shows reduction of urea level (0.4 mmol/L) and growing of alkaline phosphatase (922.9 u/l), atherogenic index was 2,19. In the second medical examination dated from 21.01.16 it was found a hypoglycemia (18.3 mmol/L). Antistreptolysin O 196,9 IU / ml (Russia index) was negative.

Immunosorbent assay of hepatitis antigen dated from 18.01.16 was negative. On direct microscopic sputum examination dated from 22.12.15 there was a massive growth of Pseudomonas aeruginosa, sensitive to ciprofloxacin, metallo-beta-lactamases, resistant to amikacin, gentamicin, piperacillin, ticarcillin, Candida albicans. Sweat test dated from 13.01.16 is 119,74 meq / l. Urine amylase dated from 12.01.16 was 61 u/l. Helico test is Hp (-) dated from 18.01.16 . Blood on glycated hemoglobin dated from 25.01.16 was 9%. Here are data of immunosorbent assay of hepatitis antigen dated from 18.01.16: HBsAg was not found, Anti-HBcor was not found, anti-HCV result was uncertain 1,24, HCV IgG/ IgM was not confirmed, HCV-cor IgG/ IgM п/т, HCV-ns IgG/ IgM п/т – were not found. Glycemic profile dated from 23.01.16: at 7 a.m - 8,7 mmol/L, at11 a.m - 11,98, at 4 p.m. - 19,1, at 20p.m. - 13,4, at 03 a.m. - 19,6 , at 7 a.m - 13,2. There is a daily urine dated from 25.01.16: protein was 0,04 g/l and acetone, glucose were negative. There is a microscopy of sputum dated from 20.01.16: yeast-like fungu was positive. ECG dated from 12.01.16 had a sinus rhythm with heart rate 80 beats per minute. Electrical axis of heart had normal position. Incomplete right bundle branch block. Voltage of left ventricular complex was raised .There is the bronchoscopy dated from 13.01.16: 2-sided catarrhal bronchitis. Fiberoptic gastroduodenoscopy dated from 14.01.16: incompetence of cardia. Duodeno gastric reflex. P esophagitis. Congestive expressed gastroduodenopathy. A chest computer tomography (20.01.16): uneven broaden on both sides, deformations of bronchi.

The walls of the bronchi are thick. Part of clearance was filled with discharge. Earlier spot changes are not defined. Lung interstices were sealed diffusely irregularly. Effusions in pleural was not defined. Increasing signs of intrathoracic lymph nodes were not noted. There was an abdominal and retroperitoneal space computer tomography dated from 20.01.16: liver and pancreas have grown up moderately, contours were equal. The structure of parenchyma was homogeneous, indices were within the normal range. Intrahepatic bile ducts were not expanded. The gallbladder was reduced, there were hyperdensive concretions in the body cavity. The lymph nodes remain in a gastrohepatic omentum of a porta of hepar. Pancreas was with signs of atrophy of the parenchyma, tail and body density of pancreas was reduced. The kidneys were located typically, not enlarged, contours were equal, renal cortex was sealed. Pelvis of kidneys were not expanded. Enlarged mesenteric lymph nodes with small calcifications were remained. Effusions in abdominal was not defined. Medical report: lipophanerosis of pancreas. There were gallstones in the bladder. It was noted the increasing of lymph nodes of abdomen.

There is a medical report of a Color Flow Doppler Mapping echocardiography dated from 21.01.16: Ectopic mount MK chords with minimal regurgitation. Regurgitation was on tricuspid valve 0 - 1 st. The cavities of the heart were not expanded. Ejection fraction - 68%. He was consulted by a gastroenterologist, and an endocrinologist.

Clinical diagnosis: Cystic Fibrosis, mixed form, severe. Chronic purulent bronchitis.

Bilateral mixed bronchiectasis of lower lobes. Chronic persistent pseudomonas infection. Chronic respiratory failure.

Chronic pancreatic insufficiency. Cholelithiasis. Chronic maxillary sinusitis. Chronic superficial gastritis common, active stage. Duodeno-gastric reflux with reflux esophagitis 1st class distal to the deficiency of the cardia rosette. Diabetes, first identified. A deviated septum of the nose to the right, without breaking the nasal breathing.

Treatment: sulperazon, ciprofloxacin, cefepime, inhalation of colistin, pulmozim, creon, ursodez, acetylcysteine, flomax, flukorus, linex, omeprazole, motilium, fluconazole.

Physiotherapy: ultraviolet irradiation № 7 on the chest, therapeutic physical training, bioptron № 4 on the chest. Replacement insulinotherapy was given: Apidra- breakfast – 3 units, lunch – 3 units, dinner – 3 units (for glucose above 14 mmol / l 1 U). Lantus: 10 p.m.- 7 units. Discharged 27.01.16.

CONCLUSION

Thus, the feature of the case of cystic fibrosis (CF) in a 14 year old boy is a complicated course with chronic *PSEUDOMONAS AERUGINOSA* infection and persistent connection of Cystic Fibrosis-related diabetes.

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EXPERIENCE EXCHANGE

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THE DYNAMICS OF THE LIVER FUNCTIONAL STATUS IN PATIENTS WITH ODONTOGENIC PURULENT-INFLAMMATORY DISEASES OF THE MAXILLOFACIAL REGION, COMPLICATED BY SEPSIS

ABSTRACT

To assess the degree of hepatic function disorder in patients with widespread phlegmons of the maxillofacial area, complicated by sepsis, the researchers studied blood levels of cytolytic enzymes alanine aminotransferase and aspartate aminotransferase. It is established that at the widespread phlegmons of maxillofacial area complicated by a sepsis the concentration augmentation in blood serum of transaminases is observed, and degree of their augmentation corresponds to the standard criteria of expression of pathological process. Including in a complex of medical actions of intravenous laser radiation of blood in a combination with sodium hypochlorite, especially a combination of ultra-violet radiation of a blood with a double irrigation of wounds by sodium hypochlorite, contributes to normalization of cytolytic enzymes indicators by 6-7 days from treatment initiation.

Keywords: odontogenic pyoinflammatory diseases, sepsis, maxilla-facial phlegmon, odontogenic infection, sodium hypochlorite, ultra-violet radiation of a blood.

At patients with acute odontogenous pyoinflammatory processes of maxillofacial area complicated by a sepsis often functions of internals and their systems, including a liver [2,4,5,8] are broken that often leads to serious disorders of a regulation of metabolic processes and change of a metabolism in an organism in general [7,9,10,12]. These circumstances dictate need of carrying out complex assessment of the general and local disorders, and also development of

Table 1

Distribution of patients on gender and age, persons

Group of a research		Age						total
		Young 18-44 years		Average 45-59 years		Elderly and senile 60 and more years		
		M	W	M	W	M	W	
Control group		5	5	5	5	5	5	30
Primary group	1 subgroup of a research	2	5	4	3	1	2	17
	2 subgroup	5	2	3	4	3	2	19
	3 subgroup	3	2	1	3	-	-	9
	4 subgroup	3	3	4	3	-	1	14