УДК 616.8-009.86-036.2-053.2

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ENCEPHALOTRIGEMINAL ANGIOMATOSIS OF STURGE-WEBER-KRABBE-DIMITRI. THE CASE FROM CLINICAL PRACTICE

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Summary. This paper presents data about rare congenital pathology from the group of the phakomatoses due to violation of neuronal proliferation — encephalotrigeminal angiomatosis. There were considered modern clinical, diagnostic and curative aspects of this pathology. It was described nonclassical clinical case of the patient during examination and treatment in the department of children's psychoneurology of the Institute of Pediatrics, Obstetrics and Gynecology of the National Academy of Medical Sciences of Ukraine.

Keywords: encephalotrigeminal angiomatosis, phakomatoses, violations of the neuronal proliferation, diagnosis, treatment, children.

Encephalotrigeminal angiomatosis of Sturge—Weber—Krabbe—Dimitri is classified as rare hereditary sporadic disease from phacomatosis group, characterized by presence of angiomatosis of brain lining vessels, face and eye capillaries with frequency 1 for 50 000 pregnancies; at that no differences by gender are noted [44, 52]. Phacomatoses are the group of hereditary progressing diseases with combined nervous system, skin, eyes and internal organs affection due to disturbances in neuronal proliferation processes during neurogenesis in foetus. The most frequent phacomatoses with nervous system affection are the following: Recklinghausen neurofibromatosis, Bourneville tuberous sclerosis (Bourneville—Pringle disease), encephalotrigeminal angiomatosis (disease of Sturge—Weber—Krabbe—Dimitri), cerebro-retinal angiomatosis (Hippel—Linday disease), ataxia-telangiectasia (Louis—Bar syndrome), cortical-meningeal diffuse angiomatosis of van-Bogaert-Divry, and some other forms [2, 19, 54]. We should remember that during neurogenesis certain stages of nervous system formation are singled out (dorsal and ventral induction, neuronal proliferation, migration, organization and myelinisation), and each development abnormality (malformation) of brain is connected with these stages. At neuronal proliferation stage (pregnancy month 2–5) neurons and glia proliferation in periventricular zones takes place with neurons further migration and formation of cortical and sub-cortical structures. In case of neuronal proliferation disturbances phacomatoses develop, in particular, encephalotrigeminal angiomatosis; microcephaly; megaloencephaly, etc. Angiomes are formed from the remains of vascular plexus, which remains in nerve tube head zone [10, 11, 44]. Neuronal proliferation process disturbances are caused both by hereditary factors (genetic, chromosomal, teratogens), and sporadic disease factors with new spontaneous genetic mutations. In recent publication from group of authors (Shirley M. D., Tang H., Gallione C. J. et al., May 2013), the information is presented about certain mutation isolation in gene GNAQ, which may cause encephalotrigeminal angiomatosis [16, 17, 48].

Encephalotrigeminal angiomatosis was first described in 1879 by W.A. Sturge (Sturge W.A. A case of partial epilepsy, apparently due to a lesion of one of the vasomotor centres of the brain. Transactions of the Clinical Society of London. — 1879. — 12:162), who supervised the patient with facial skin angioma, glaucoma, partial epileptic seizures, and hemiparesis. In 1922 F.P. Weber discovered intracranial petrifactions on the X-ray of patient with similar clinical manifestations (Weber F.P. Right-sided hemi-hypertrophy resulting from right-sided congenital spastic hemiplegia, with a morbid condition of the left side of the brain, revealed by radiograms. Journal of Neurology and Psychopathology (London). — 1922. — 3: 134–139). The similar radiologic changes in this pathology were reported by V.Dimitri in 1923, and K.Krabbe described morphological changes in this disease. In 1937 van der Hoeve included encephalotrigeminal angiomatosis into phacomatoses group, which was reflected in his work (van der Hoeve J., Mahoney W. A fourth type of phakomatosis: Sturge-Weber Syndrome. Verh K Akad Wet Amst 36:1937) [3, 44].

Encephalotrigeminal angiomatosis is characterized by classis symptoms triad (facial angiomatosis, brain lining angiomatosis, glaucoma and convulsions). However, this classic symptoms triad is met in about 20% of cases; more frequently are noted mono- and bi-symptom forms of this pathology [7, 21, 24, 36].

According to E.S. Roach classification [42], there are 3 basic forms of encephalotrigeminal angiomatosis:

I type: facial angiomas and leptomeningeal angiomas, sometimes with glaucoma;

II type: only facial angiomas, without central nervous system affection, sometimes with glaucoma;

III type: isolated leptomeningeal angiomas, usually without glaucoma.

We should mention that apart from these 3 basic forms of encephalotrigeminal angiomatosis there are also its atypical forms as individual syndromes:

— Jahnke syndrome (1930), — at that glaucoma is absent;

— Schirmer syndrome (1860), — at that glaucoma and hydrophthalmus appear early;

— Lawford syndrome (1884), — at that glaucoma appears lately, eye balls volume does not increase;

— Milles syndrome (1884), — its characteristics are combination of choroid tunic angioma without eye balls volume increase [3, 44].

Nevus angiomatodes in case of encephalotrigeminal angiomatosis manifests itself already during the first months of life, and is typically situated in innervation zone of Ist and IInd branches of trifacial nerve (ocular, maxillary) from one side in 70 % of cases, or on two sides in 30 % of cases as «burning spot». At birth the nevus is of pale-pink colour, and later becomes brighter and darker («port wine colour»). In 40 % of cases nevus angiomatodes is situated on trunk and extremities [1, 38, 45]. Neurological manifestations character

depends on the degree and position of angiomatous transformation of vascular and pia matter (leptomemgial angiomatosis), which is more often situated in parietal and occipital lobes, and may be accompanied by hemiparesis, hemianopsia, mental retardation, behavioural disorders, transitory disturbances in brain circulation and epileptic seizures. There is no certain link between skin angiomatosis expansion and epileptic seizures pronouncement; however, there is a connection between epileptic seizures frequency and mental retardation degree and severe psychic disturbances. Convulsions may manifest already during the first year of life at first from the side opposite to angioma, later they become generalized and increase further with age. During the first year of life epileptic seizures happen in 75% of such patients, and by the age of 5 years their frequency increases to 95 %, and about 60 % of such seizures become resistant to anticonvulsive therapy, demanding neurosurgical intervention. In some cases there is a combination of encephalotrigeminal angiomatosis with microgyria, which considerably worsens prognosis [14, 19, 29, 49, 50]. Arrested development and mental retardation happen in 50–60 % of cases, hemiparesis — in 25–56 % of cases, constituting in general 33% [26, 43]. In case of disease progression may develop vessels calcification, cerebral atrophy, intracranial hypertension and hydrocephaly (due to drainage disturbances). Some patients manifest severe headache attacks with vomiting — migraine-like attacks [27, 34, 39]. Eye affection in this pathology is manifested as iris angiomatosis, angiomatosis of ciliary body and the proper vascular membrane. In 50–70 % of cases takes place glaucoma, which may be present from birth (in 60% of cases), or is formed with age (in 40 %). There are described cases of hydrophthalmus, retinal epithelium degeneration, retinal detachment and vision loss [15, 47, 50, 51].

Among important methods of this pathology diagnostics we should name skull X-ray examination (which allows revealing cortical calcification in the form of winding band-like strips, which repeat brain surface contours («tram way» symptom)), computer tomography, magnetic resonance tomography (MRT) and MRT in angiographic regimen (which reveals angiomatosis of vascular and pia matter and specifies its intracranial location). In the majority of cases brain angiomas are situated on the same side that facial angiomas with gradual calcification and brain atrophy in affected zone. Also the method of diffuse tensor magnetic resonance tomography is used to assess conduction tracts and affection degree of brain white matter in this pathology at microstructural level [5, 6, 20, 25, 32, 35, 40, 46, 53]. Method SPECT (single-photon emission computed tomography) discovers hyperperfusion foci at early disease stages, and hypoperfusion foci — at later stages; method PET (positron tomography), in particular, with isotope 18-fluorodeoxyglucose (18- FDG PET), reveals hypo-metabolism foci [4, 9, 30, 33]. Magnetic resonance spectroscopy (MRS), discovered decrease in NAA (N-acetyl aspartate) level in brain in encephalotrigeminal angiomatosis cases. N-acetyl aspartate is acetic groups' donor in the process of myelin synthesis by glial cells, and its decrease is considered to be the reliable indicator of neuronal dysfunction and neuronal death [13, 18, 31, 37].

This pathology differentiated diagnostics should be made for other diseases from phacomatosis group and for other congenital abnormalities of central nervous system development.

Encephalotrigeminal angiomatosis treatment is symptomatic, using anticonvulsive, neuroprotecting preparations, and medicines that decrease intracranial and intraocular pressure. By individual indications laser-therapy and roentgen therapy methods may be used. In case of epileptic seizures resistance neurosurgical interventions are applied, such as: hemispherectomy, focal cortical resection, callosotomy, or nervus vagus stimulation [8, 12, 22, 23, 28, 41].

Below is described clinical case of girl patient O., 12 years old, who underwent examination and treatment in Children Psychoneurology Department of State Institution «Institute of Pediatry, Obstetrics and Gynaecology of the National Academy of Medical Sciences of Ukraine» with the diagnosis: «Symptomatic epilepsy, frequent polymorphous seizures (myoclonal, complex fractional seizures). Sturge-Weber disease. Hypoplasia of right temporal lobe pole and angiomatosis of brain lining in right parietal zone (according to MRT results)».

The girl was admitted into hospital with complaints on seizures of polymorphous character (myoclonal, complex fractional seizures), more frequently as myoclonus during falling asleep. Myoclonus had the form of single myoclonal convulsions almost every day or every other day, and could have duration for 20–30 minutes.

Medical history data — the girl from the second pregnancy (from the first pregnancy — the boy, now aged 19 years, healthy). The second pregnancy went on with abortion threat during the second trimester. Delivery took place after 38–39 weeks; it was rapid natural labour. Body weight at birth was 3800 g, body length 54 cm, Apgar scale assessment 6/6 points, umbilical cord was wrapped around the neck (not tightly). The girl stayed for treatment in intensive therapy department for new-borns at home with diagnosis: «Hypotoxic-traumatic CNS affection. Brain oedema. Erb paresis on the right». Further development was uncomplicated. Parents negate hereditary diseases. At the age of 1.5 years the girl manifested seizures — as hands myoclonus. She received Phenobarbital for one year (with insufficient effect). Later, after she was prescribed Carbamazepine the seizures frequency increased. Further the girl received Depakine, Topamax (with insufficient effect), and recently before admitting into the department she started taking Levetiracetam.

Neurological status — craniocerebral nerves functions — unremarkable. The girl is oriented in time and space. Her consciousness is not disturbed. She answers questions correctly. Tendon reflexes, D=S. Abdominal reflexes positive. She does not present focal symptoms, pathological reflexes, or coordination and sensitive disturbances.

Brain magnetic resonance tomography (MRT) — conclusion: brain lining angiomatosis in the right parietal zone (Sturge—Weber syndrome), pole hypoplasia of the right temporal lobe.

Electro-encephalogram (EEG) — conclusion: moderate diffuse changes of regulatory character due to diffuse slowdown, pronounced alpha-rhythm dysrhythmia, splashes of acute theta-asynchrony in parietal-temporal zones, predominantly on the left.

Neurosurgeon consultation, — at the moment of examination neurosurgical treatment is not indicated.

Logopedist-psychologist consultation — conclusion: psychological deviations are not established.

Ophthalmologist consultation — conclusion: vis od/os-1.0, optic media and eye fundus are normal.

During treatment process (neuroprotective, anticonvulsive, metabolic therapy) the anticonvulsive therapy was corrected and Levetiracetam dose increased up to 60 mg/kg of body weight. During treatment the girl manifested 2 seizures as single myoclonic twitching in a hand during several seconds. The girl was discharged in satisfactory condition for neurologist and paediatrician supervision at home.

This case illustrated non-classical course of Sturge—Weber—Krabbe—Dimitri syndrome without facial angiomatosis

and glaucoma, but with brain lining angiomatosis, hypoplasia of right temporal lobe pole and frequent polymorphous seizures. This pathology is classified as «neuronal proliferation disturbance» with specific structural changes in central nervous system and various clinical manifestations. Therefore, it stresses importance and need for timely application of neurovisualization methods for confirmation and differential diagnostics of such pathological condition, because the timely diagnosis, accompanied by early medical-genetic examination determines further management of patients with this nosology.

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