CLINICAL DESCRIPTION OF PATIENTS WITH RECIDIVOUS VENOUS BRAIN INFARCTIONS AND WITH FACTOR II (PROTROMBIN) - MUTATION G/A 20210 AND WITH ERYTEMA NODOZUM

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ABSTRACT

The aim of the current investigation was to describe the clinical history of two patients; treated at the Clinic of Neurology of UMHAT „D-r G. Sranski" – Pleven. They were admitted from the emergency department with recidivous venous brain infarctions.

Material and methods: The patients were admitted from the emergency department of the University Hospital – Pleven in the Clinic „ Neurology and Neurosurgery". They were at severe condition, at epileptic status. Complete anamnestic data were taken. Double brain MRT and CT scan, ECG, Ultra sound of the abdomen and heart, X-ray of the lungs, EEG, biochemical, microbiological, immunological, serologic and genetic tests, lumbar puncture were applied. The patients were discussed with neurologist, neurosurgeon, cardiologist, ophthalmologist, anesthesiologist and infectionist. Coagulation tests were complied. The fibrinolythic and antiepileptic treatment was continued successfully. The patients were discharged in good condition without motor deficit and epileptic symptoms.

Results: The analysis of the received results and data of the laboratory investigations revealed challenges and alternatives concerning the clinical interpretations, diagnosis and differential diagnosis.

Conclusions: It's necessary to be provided more investigations in the Genetic and Molecular pathology laboratories for the correct final diagnosis.

Key words: *venous brain infarctions, risk of thrombophylia, factor II (Protrombin) – mutation G/A 20210, erytema nodozum, differential diagnosis.*

Introduction:

The thrombosis of the dural sinuses and of the brain venous vessels are rare forms of stroke – 0,5 – 1 % of whole stroke cases. According to the biggest investigation [International Study on Cerebral Venous and Dural Sinuses Thrombosis (ISCVT)] – 78% of patients are under 50 years. There are no population investigations.[3]

Risk factors are acquired: trauma, surgery interventions, pregnancy, puerperium, antiphospholipid syndrome, hormon therapy, cancer **and genetic -** (hereditary thrombophylias). [1]

The clinical diagnosis depends on the typical clinical symptoms, neuroimaging confirmation and doctor's experience. The clinical picture is a result of the elevated intracranial pressure and of the focal brain injury from venous infarction or haemorrhage. The headache is the most frequent symptom – at 90% of the patients. The headache is diffuse, becomes progressively more severe for days to weeks. At approximately 25% of patients, it might be **isolated headache (idiopathic intracranial hypertension) without** focal deficit and papiledema. These cases are a diagnostic challenge and clinical exam for the qualification of the neurologist. The clinical symptoms depend on the localization of the cerebral venous or dural sinuses thrombosis. Most frequently superior saggital sinus is affected, which leads to headache, elevated intracranial pressure, papiledema. Thrombosis of the transversal sinus most frequently is associated with inflammation and infection of the internal ear.

Thrombosis of the deep venous vessels – 16% of the patients could be associated with infarctions in the thalamus and basal ganglia. Approximately 40% of the patients are with epileptic seizures. Bilateral injures of the brain structures are common in cerebral venous or dural sinuses thrombosis. Slowly development of the symptoms is characteristic for cerebral venous and dural sinuses thrombosis. At 58% of cases the diagnosis is established > 48 hours to 30 days from the initiation of the symptoms. The CT with contrast shows the classic „empty delta sign" – central hypointensity, because of very slow or even lack of blood flow in the sinus and contrast enhancement around it. MRT is with higher sensitivity than the CT. During the first week the cerebral venous or dural thrombosis might be isointensive at T1 and hypointensive at T2, because of elevated deoxyhemoglobin. After the second week the lesion is hyperintensive at T1 and T2, because of degradation of the methemoglobin. The negative CT and MRT findings don't reject the diagnosis. In these cases angiography should be applied. [4]

The aim of the current investigation was to describe the clinical history of two patients, treated at the Clinic of Neurology of UMHAT „D-r G. Sranski" – Pleven. They were admitted from the emergency department with recidivous venous brain infarctions.

Material and methods:

The patients were presented at the Clinic Neurology and Neurosurgery from the emergency department of the University Hospital – Pleven. They were in severe condition with epileptic status. Complete anamnestic data were taken. The classic clinical triad of headache, focal neurologic deficit and elevated intracranial pressure was diagnosed. The headache was constant, sharp, hemicranial or generalized, and became progressively more severe, refractery to therapy. They were with focal neurologic deficit including light central hemiparesis, dysarthria, disturbed consciousness, visual disturbances, mental dysfunctions and epileptic status.

Investigations and Results:

Double brain MRT and CT scan, ECG, Ultra sound of the abdomen and heart, X-ray and CT of the lungs, EEG, biochemical, microbiological, immunological, serologic and genetic tests, lumbar puncture were applied. The patients were discussed with neurologist, neurosurgeon, cardiologist, ophthalmologist, anesthesiologist and infectionist.

MRT of the brain, native and post contrast: **Patient № 1 -** N.R.K. (with erytema nodozum). The described findings might be interpreted as parenchimal changes associated with venous thrombosis of the superficial venous vessels parieto-occipitaly at the left side.

MRT of the brain - control investigation after 30 days: Normal MRT brain investigation showing normal structures and signal density.

MRT of the brain, native and post contrast: Patient № 2 – M.V.B. **(confirmed heterozygote type possessing of the gen coding factor II (Protrombin) – mutation G/A 20210).**

She was operated 4 years ago for subcortical glyosis. The described findings might be interpreted as changes associated with partial thrombosis of the sinus sagitalis superior at the middle third with recidivous venous infarctions at the area of Gyrus calcarinus dex.

MRT of the brain - control investigation after 30 days: status after osteoplastic trepanation of the left parietal area with perifocal cortical and subcortical glyosis. A little encephalomalatic lesion at the irrigation area of the a.cerebri post.dex. – chronic brain infarction. A little area of subcortical glyosis at the left frontal lobe. Without other pathologic changes of brain parenchima.

EEG of the patient № 1: Focal activity with complexes pick-wave from the parieto-temporal area at the left side with marked secondary bilateral synchronisation. Fone activity – alpha rhytm 8-9 Hz, good modulation and saved somatotopic distribution.

EEG of the patient № 2: Focal activity - wide left-sided distribution of groups with high voltage and slow waves. Fone activity – alpha rhythm 9-10 Hz, bilateral posterior maximum, wide anterior distribution. The analysis of the received results and data of the laboratory investigations revealed challenges and alternatives concerning the clinical interpretations, diagnosis and differential diagnosis.

Differential diagnosis: It may concern bacterial meningitis, brain abcess, acute disseminated encephalomyelitis (ADEM), subdural empyema, viral meningoencephalitis, primary and metastatic brain tumors, cerebral infarction or hematoma. These are all conditions that might include the classic clinical triad of headache, elevated intracranial pressure and focal neurologic deficit.

The treatment of the cerebral venous and dural sinuses thromboses includes: heparine or low-molecular heparine in full anticoagulant doses, followed by indirect anticoagulants, acetazolamide in the cases with elevated intracranial pressure, anticonvulsant therapy, antibiotics, LP, shunt, monitoring of the visual functions, decompressive hemicranectomia.[2] The patients were discharged in good condition without motor deficit, seizures or other symptoms. Complications from the other organs and systems of the body were not established.

Conclusions:

1. It's necessary to be provided more investigations and tests in the Genetic and Molecular pathology laboratories for the correct final diagnosis.
2. Because of new neuroimaging techniques and improvement of surgical and antibiotic treatment, the mortality rate because of venous and dural sinuses thromboses has significantly declined.
3. The diagnosis and treatment of the cerebral venous or dural sinuses thrombosis, even now, are of therapeutic challenge. The treatment must be enough in duration and with adequate anticoagulation screening and observation.

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