Relato de Caso

Thrombotic Thrombocytopenic Purpura after Ischemic Stroke: Case report

Púrpura Trombocitopênica Trombótica após Acidente Vascular Cerebral Isquêmico: Relato de Caso

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RESUMO

Introdução e proposta: Púrpura Trombocitopênica Trombótica é uma doença rara cuja causa é desconhecida. Além da trombocitopenia, ela é caracterizada por anemia hemolítica, sintomas neurológicos, febre e anormalidades renais. A maioria desses achados são causados por formação de trombos em capilares e arteríolas pelo corpo. Esse artigo reporta um caso de TTP após acidente vascular cerebral e sua proposta é descrever as características clínicas e patofisiológicas da TTP. Relatamos um mulher de 52 anos que apresentou três episódios de acidente vascular cerebral. O terceiro episódio foi complicado pela TTP. Anemia hemolítica e trombocitopenia foram identificadas.

Unitermos: Púrpura Trombocitopênica Trombótica, Acidente Vascular Cerebral.

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SUMMARY

Background and purpose: Thrombotic thrombocytopenic purpura (TTP) is a rare disorder, the exact cause of which is unknown. In addition to thrombocytopenia, it is characterized by hemolytic anemia, changing neurological symptoms, fever, and renal abnormalities. Most of these findings are caused by the formation of platelet thrombi in capillaries and arterioles throughout the body. This article reports a case of TTP after Ischemic Stroke and its purpose was to describe the clinical and pathophysiologic features of TTP. We describe a 52-year-old woman who presented three onset of ischemic stroke. A third episode of ischemic stroke was complicated by TTP. Hemolytic anemia and thrombocytopenia were identified.

Keywords: Thrombotic Thrombocytopenic Purpura, Stroke.

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CASE REPORT

A 52-year-old woman, who presented with three onset of ischemic stroke, with interval of four months between the first one and almost one month in the last one. After the first onset she had paresthesia in right hand, paresis in right superior limb, and motor dysphasia.

Neurological examination on the third admission showed disturbance of consciousness, worsened of the muscular power, right hemiparesis and dysphasia.

Blood parameters revealed in the third onset severe hemolytic anemia (hemoglobin 7.8g%, erythrocyte 2,360,000 $10^3/mm^3$, hematocrit 23.4%, reticulocyte count

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9.5%), severe thrombocytopenia (platelet count 14.000/ μl). Anisocytosis, macrocyte, polychromatophilia, poikilocytosis. Results of prothrombin time, activated partial thromboplastin time were normal. Unconjugated bilirubin concentrations were elevated. There was mildleukocytosis with normal representation of the granulocyte and lymphocyte lines. Hematuria and proteinuria.

CT showed reduction of brain hemisphere volume. Parietal, right posterior fossa, and left superficial frontal area hypodensity.

The tests for underlying diseases that known to complicate disseminated intravascular coagulation (DIC) were negative. A diagnosis of thrombotic thrombocytopenic purpura was made.

Laboratorial data in episode before were within normal values.

THROMBOTIC THROMBOCYTOPENIC PURPURA

In 1924, Dr. Eli Moschcowitz published the first case study of young woman with "an acute febrile pleomorphic anemia with hyaline thrombosis of the terminal arterioles and capillaries". Fever, bleending, neurologic and renal abnormalities, and a microangiopathic hemolytic anemia characterized her illness. This syndrome subsequently became know as Moschcowitz's disease, or "thrombotic thrombocytopenic purpura (TTP)"[1].

There are two types of TTP: the single acute episode and chronic relapsing TTP.

TTP is characterized by the typical findings of severe thrombocytopenia (usually less than 10,000 platelets/ mm^3 in acute episode), hemolytic anemia, schistocytosis, and polychromasia[2]. Central nervous system ischemia and neuropathy may range in severity from transient bizarre mentation and behavior to sensory motor deficits, aphasia, seizures or coma, fever, which occurs in only a minority of patients, and renal abnormalities, with hematuria and/or proteinuria. Renal failure is rare. Symptoms of ischemia in gastrointestinal circulation, especially abdominal pain[3], have been recognized with increasing frequency.

Pathophysiology

The basic pathologic processes of TTP have been proposed to involve endothelial cell injury and the subsequent releas of platelet-aggregating substances that result in the formation of thrombotic lesions in terminal arterioles and capillaries[4]. The specific underlying cause of these thrombi has not been established.

The lesions of TTP are seen most commonly in the microvasculature of the brain, kidney, pancreas, heart, spleen, and adrenal glands, but can be found throughout the body, though less commonly in the lung or liver. The partial and complete occlusions in many organs, leading to organ failure, along with the consumptive thrombocytopenia, result in the clinical manifestation of TTP.

Recently, investigators showed that a deficiency in a specific plasma protease responsible for cleaving vWF plays a crucial role in the pathogenesis of congenital and acquired idiopathic trombotic thrombocytopenic purpura[5]. The von Willebrand factor-cleaving protease has now been identified as a new member of the ADAMTS family of metalloproteases, designated ADAMTS 13 [6].

CONCLUSIONS

It is generall assumed that endothelial cell injury is the primary event in the pathogenesis of TTP. Our patient developed TTP after ischemic stroke. Romani et al suggest that endothelial cell activation is not the primary event leading to TTP. Vascular perturbation seems to be a consequence, rather that a cause, of the disease[7]. Investigation of vascular injury and mechanisms of microvascular thrombosis remain the frontiers of investigation in thrombotic microangiopathy.

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ERRATA

Na Revista Neurociências, edição "Volume 13 nº 01", no artigo "Qualidade de sono de pacientes com esclerose lateral amiotrófica: análise dos instrumentos de avaliação", página 24, considerar na Tabela 1 "depressão ≥ 20" e "STAI ≥45"; na página 26 onde se lê "masii" o correto é "mais" e o último parágrafo termina "com ELA". No artigo "Procedimentos fisioterápicos para disfunção vésico-esfincteriana de pacientes com traumatismo raquimedular – revisão narrativa", página 36, deve-se ler o Quadro 1 com suas divisões corretas:

Quadro 1. *Literatura sobre tratamento fisioterapêutico da bexiga neurogênica após TRM.*

AUTORES	TIPO/ESTUDO	Nº pts	COMENTÁRIOS
Eustice et al, 2002	revisão sistemática	355	verificaram a importância dos efeitos do esvaziamento vesical cronometrado e incitado
Cheng et al, 1998	Ensaio clínico randomizado	80	avaliar a acupuntura em bexiga neurogênica por lesão da medula espinal – relatório preliminar relatando os benefícios com a técnica.
Yamanishi et al, 2000	ensaio clínico randomizado	68	ensaio clínico randomizado duplo-cego de eletroestimulação para incontinência urinária por hiperreflexia do detrusor
Gomes, 1998	estudo de casos e controles	20	realizar trabalho muscular isométrico no tratamento da reeducação vesical em portadores de bexiga neurogênica flácida decorrente ao choque medular pós-traumatico verificando 70% de melhora dos pacientes
Hongo et al, 2000	estudo de casos e controles	13	acupuntura para os sintomas clínicos e mensurações hemodinâmicas em pacientes com lesão da medula espinal com hiperreflexia do detrusor.
Weiss, 2001	estudo de casos e controles	52	técnica miofacial em pacientes com urgência e aumento da freqüência urinária característica da bexiga hiperativa
Amarenco et al, 2003	estudo de casos e controles	44	comparando as cistometrias antes e após a eentp. 21 pacientes ocorreram aumento em 50% ou mais no volume no momento da contração não inibida, 13 aumentaram mais de 100 ml o volume da 1ª. contração, 15 melhoraram mais de 50% da ccm e 1 melhorou a ccm sem aumentar o volume na contração não inibida.
Fakacs et al, 1992	revisão de literatura		reabilitação das bexigas neurogênicas
Bo et al, 2000	revisão de literatura		tratamento não farmacológico para bexiga hiperativa- exercícios do assoalho pélvico
Payne, 2000	revisão de literatura		terapia comportamental para bexiga hiperativa
Vapnek, 2001	revisão de literatura		tratamento das disfunções urinárias
Roe et al, 2002	revisão de literatura		treino vesical em pacientes adultos com incontinência urinária
Sakakibara et al, 2001	revisão de literatura		crise de hiperreflexia autonômica em pacientes com bexiga neurogênica
Magaldi et al, 2002	revisão de literatura		fisioterapia uroginecológica: incontinência urinária e prolapso