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BENIGN ANGIOPATHY OF THE CENTRAL NERVOUS SYSTEM OR REVERSIBLE CEREBRAL VASOCONSTRICTION SYNDROME

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SUMMARY – Benign angiopathy of the central nervous system is a subset of primary angiitis of the central nervous system characterized by "benign" course. It means that changes of cerebral vessels are reversible after treatment with corticosteroids and calcium channel blockers, so these abnormalities are believed to reflect vasospasm rather than true vasculitis. The diagnosis is made on the basis of clinical presentation, brain magnetic resonance imaging and cerebral angiography. We present a young man with acute onset of headache and neurologic impairment secondary to ischemic stroke with intracerebral and subarachnoid hemorrhage. Cerebral angiography showed characteristic findings of diffuse vasculitis but good response to treatment with corticosteroids and calcium channel blockers distinguish this benign angiopathy from the more aggressive form of the central nervous system vasculitis.

Key words: *Cerebrovascular disorders – diagnosis; Cerebrovascular disorders – drug therapy; Cerebral arterial diseases – complications; Benign angiopathy; Vasoconstriction; Cerebral angiography; Corticosteroids; Calcium channel blockers; Case Ceport*

Introduction

Primary angiitis of the central nervous system (PACNS) is a rare disease with potentially harmful or even fatal outcome that often affects young adults and can be distinguished from secondary vasculitis associated with infectious disorders, connective tissue diseases, malignancies and toxic drug effects[1]. The angiitis is focal and segmental in distribution, involving small and medium-sized leptomeningeal and intracranial vessels[2]. The spectrum of neurologic symptoms is multifaceted; the patient may have intense headache, confusion, decreased cognitive function, changes in consciousness, epileptic attacks and symptoms resembling multiple sclerosis[3]. The diagnosis is frequently made on the basis of clinical presentation, brain magnetic resonance imaging (MRI) and cerebral angiography without pathologic confirmation[4], but definitive diagnosis can only be made on the grounds of biopsy from leptmeninges and parenchyma[5,6]. Benign angiopathy of the central nervous system (BACNS) is a subset of PACNS characterized by acute presentation (most commonly headache), normal to mildly abnormal cerebrospinal fluid (CSF) findings, female predominance and highly abnormal cerebral angiography (reversible after treatment), requiring less intensive treatment than has been traditionally used[7,8]. The term "benign" may be misleading, however, because strokes and significant neurologic dysfunction sometimes occur. BACNS could respond favorably to treatment with corticosteroids alone or in combination with calcium channel blockers, so it is very important to distinguish it from more aggressive PACNS[9].

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Case Report

A 27-year-old male was admitted to Intensive Care Unit for sudden onset of severe headache, nausea, vomiting, confusion, aphasia and development of right hemiplegia with supranuclear facial palsy. Previous medical history did not reveal any disease except for bronchial asthma.

On admission, the patient underwent emergency extended neurological examination that showed deviation of the eyes to the left side, right supranuclear palsy, aphasia, right hemiplegia with sensory extinction, and positive ipsilateral Babinski sign.

He was cardiopulmonary compensated with arterial hypertension 250/150 mm Hg and immediately treated with an antihypertensive, analgesics and antiemetic. Blood pressure was monitored, routine hematologic and biochemical investigations, electrocardiography (ECG), funduscopy, chest x-ray and brain computed tomography (CT) were done.

Laboratory findings were normal except for high ESR 34, leukocytes 14.8x10, platelets 436, CRP 7.2 mg/L, AST 79 U/L, ALT 337U/L, GGT287 U/L, cholesterol 5.9 mmol/L, LDL cholesterol 4.1 mmol/L; ECG, cardiac ultrasound, funduscopy, chest x-ray, CSF, immunologic tests (CH50, C3, C4, ANF, ENA, aCL, ANCA, CD4, CD8, LAc, cryoglobulins aCI-IgG, aCI-IgM, ACE), serologic tests (HCV, AgHBs, HAV IgM, AcHIV, CMV IgM) were normal, herpes IgG was positive. CT scan of his brain showed hemorrhagic lesion of the brain stem with discrete subarachnoid hemorrhage (SAH) and extensive left parieto-occipital ischemic lesion (Fig. 1).

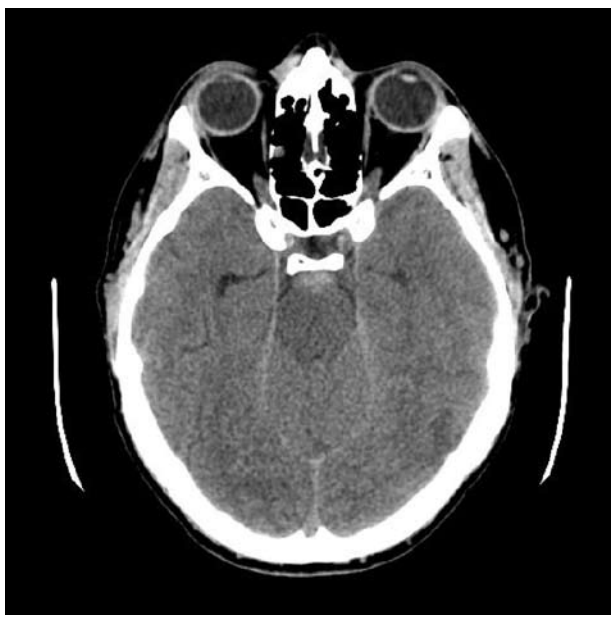

Fig. 1. Multislice computed tomography: hemorrhagic lesion of the brain stem with discrete subarachnoid hemorrhage and left ischemic lesion.

As the patient was a young person very suspect of vasculitis, he was transferred to clinical department where diagnostic procedures were continued. MRI showed hemorrhagic lesion of the brain stem with discrete SAH and extensive left ischemic lesion, while digital subtraction angiography (DSA) revealed multifocal segmental stenosis of intracranial vessels (ACM, ACA, ACP and AB). He was treated with corticosteroids and calcium channel blockers, then started early with passive physical rehabilitation, which resulted in partial recovery of neurologic disabilities with reduction of right hemiplegia and speech disturbances. One month later, he was transferred to the Center of Physical Rehabilitation and after three months, he was able to walk without assistance and had mild residual right spastic hemiparesis with motor dysphasia. Control magnetic resonance angiography (MRA) was normal.

Discussion and Conclusion

This report describes a young male patient with serious neurologic disease characterized by acute presentation (headache, confusion, aphasia and right hemiplegia). CSF analysis was normal, other systemic diseases associated with cerebral involvement were excluded with immunologic and serologic tests, and brain angiogram showed changes typical of vasculitis (segmental narrowing, ectasia and beading in multiple vascular territories). He was treated with corticosteroids and calcium channel blocker, with good response, so his course could be considered "benign" in spite of stroke and significant neurologic dysfunction. Control MRA was normal, therefore unnecessary and prolonged treatment with immunosuppressants was avoided.

Our patient had intracerebral and subarachnoid hemorrhage in a setting of vasoconstriction and high blood pressure. It is a rare finding in the setting of BACNS and may represent a new feature in some patients with BACNS[8].

The most distinctive angiographic feature of BACNS is that abnormalities are completely reversible, usually within 4 to 8 weeks. Thus, these abnormalities are believed to reflect vasospasm rather than true vasculitis[10].

Corticosteroids alone are appropriate therapy for patients with BACNS and can be tapered off over 3 to 6 months. Calcium channel blockers have been added in an attempt to reduce vasospasm. Repeated transcranial Doppler ultrasonography can be a reliable noninvasive investigation to monitor the effect of treatment and demonstrate reversibility of the vasoconstriction[11].

It is very important to recognize and distinguish benign angiopathy from the more aggressive form of CNS vasculitis because the treatment and response vary between these two entities[9].

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Sažetak

BENIGNA ANGIOPATIJA SREDIŠNJEG ŽIVČANOG SUSTAVA ILI REVERZIBILNI MOŽDANI VAZOKONSTRIKCIJSKI SINDROM

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Benigna angiopatija središnjeg živčanog sustava je podtip primarne upale krvnih žila središnjeg živčanog sustava (PACNS) koju karakterizira „benigni“tijek. To znači da su promjene na krvnim žilama mozga reverzibilne nakon liječenja kortikosteroidima i blokatorima kalcijevih kanala pa se pretpostavlja da su promjene prije odraz vazospazma nego prave upale. Dijagnoza se postavlja na osnovi kliničke prezentacije, magnetske rezonance mozga i moždane angiografije. Prikazujemo mladića s naglo nastalom glavoboljom i neurološkim poremećajem na podlozi ishemijskog moždanog udara s intracerebralnim i subarahnoidnim krvarenjem. Cerebralna angiografija je pokazala promjene tipične za difuznu upalu krvnih žila, ali dobar odgovor na liječenje kortikosteroidima i blokatorima kalcijevih kanala razlikuje ovu benignu angiopatiju od agresivnijeg oblika vaskulitisa središnjeg živčanog sustava.

Ključne riječi: *Cerebrovaskularne bolesti – dijagnostika; Cerebrovaskularne bolesti – farmakoterapija; Moždane arterijske bolesti – komplikacije; Dobroćudna angiopatija; Vazokonstrikcija; Moždana angiografija; Kortikosteroidi; Blokatori kalcijevih kanala; Prikaz slučaja*