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PRIMARY CEREBRAL VASCULITIS AS CAUSE OF ISCHEMIC STROKE CASE REPORT

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Abstract

Primary vasculitis of the central nervous system (PACNS) is a rare disorder of the blood vessels of the brain and the spinal cord without any evidence of systemic vasculitis. The clinical course of cerebral vasculitis ranges from fulminant to indolent and may be marked by fluctuations in clinical signs.

We present the case of a 45 years old male patient who was admitted to our hospital due to exhibiting symptoms consistent with a frontal lobe syndrome. Clinically, he manifested alterations in behavior, apathy, and loss of sphincter control. The initial symptoms he had one month prior to the admission to our hospital were left hemiparesis, confusion and disorientation. Onset at a young age and absence of risk factors indicated the presence of an uncommon cause of stroke.

To elucidate the underlying cause, a series of examinations were conducted, including three instances of brain MRI scans, computed tomography angiography, carotid Doppler ultrasound, and an electroencephalogram.The finding of morphologically altered blood vessels, with thickening of the wall of the right internal carotid artery, with reduced and irregular flow, reduced flow speeds and initial vascular resistance along the entire length up to the intracranial entrance does not rule out that it can be one of the forms of vasculitis.

Keywords: vasculitis,angiitis, stroke, brain,magnetic resonance imaging

Introduction

The term cerebral vasculitis (arteritis,angiitis) encompasses several inflammatory vasculitides that lead to stenosis, occlusion, or rupture of an artery, capillary, or venule in the central nervous system [1].

The exact etiology and pathogenesis of PACNS are unknown, but some infectious agents and connective tissue disorders are postulated as triggers. Studies which describe the exact epidemiology of this rare disorder have not been done, and the available data shows an annual incidence rate of 2,4 cases per 1 million person per year. The disorder has been found to have an equal distribution among both males and females, and the median age of diagnosis is about 50 years [2].

The onset of PACNS is usually insidious, and the course is slowly progressive. Clinical manifestations at the time of diagnosis are non-specific with various presenting symptoms, the most common being headaches, cognitive dysfunction and stroke symptoms. A combination of symptoms is usually present in most patients [2,3].

Case report

We will be discussing a case involving a 45 year old male patient who was admitted to our hospital due to exhibiting symptoms consistent with a frontal lobe syndrome. Clinically, he manifested alterations in behavior, apathy, and loss of sphincter control.

The initial symptoms he had one month prior to the admission to our hospital were left hemiparesis, confusion and disorientation. He had no past medical history and was not taking any prescription medications. The neurological examination showed impaired consciousness, disorientation and confusion.

He had left central facial palsy with left hemiparesis. An MRI scan of the brain revealed bilateral demyelinating changes in the frontal and right temporal regions, accompanied by gyral edema observed in

T1 and high - intensity signals evident in T2 and FLAIR sequences. This finding raised a suspicion of encephalitis or low - grade astrocytoma. Because of the finding on the MRI scan of the brain, a neurosurgeon and an infectologist were consulted. A lumbar puncture was preformed, yielded normal results and therefore infective etiology has been excluded.

The neurosurgeon suggested an MRI spectroscopy to further investigate, with no indications pointing towards the need for neurosurgical intervention. In the meantime, the mother of the patient has noted that in addition to a minimal improvement on the mental status, the patient has a complete loss of sphincter control.

At the time of the admission to our hospital the neurological examination revealed a provoked rhythmic nystagmus on the left and when preforming passive movements, pyramid- extrapyramidal hypertonia is registered on the left, pathological reflexes are absent. Speech seems fluent, but an objective assessment is not possible due to insufficient cooperation from the patient, verbal contact is extremely difficult to establish, the patient cries during conversation. There is sphincter dysfunction, urinary and fecal incontinence. Onset at a young age and absence of risk factors indicated the presence of an uncommon cause of stroke.

A series of blood tests were performed, all yielding normal results, except for an elevated erythrocyte sedimentation rate (ESR= 38mm/h), the C-reactive protein levels (CRP = <3.34mg/dl). Tests of blood coagulation and fibrinolysis yielded normal results, just slightly elevated D-dimers count (1.20mg/dl).

Tumor markers (CEA=0.53ng/ml; CA 19-9=6.06U/ml; PSA=0.718ng/ml;Prolactin=10.9ng/ml), and viral markers (AntiHBsDi3=0.00 Miu/ML; AntiHCVII=0.07 nonreactive; HAVAbIgM2=0.22nonreactive; HBsAgQ2=0.21nonreactive), were all with normal results. Immunological analyzes of blood (IgG=9.66g/l; IgA=2.22g/l; IgM=0.755g/l; C3=1.52g/l; C4=0.386g/l; IgE1=361IU/ml; CRP1=<3.3mg/l; RFn= <9.38IU/ml; Antinuclear antibodies (ANA, Anti-ds DNA, Nucleosomes, PCNA, SS-A, SS-B, Scl70, AMA M2, Jo-1, Mi-2, KU), all negative. Pneumoslide testing for Legionella pneumophilla, Mycoplasma pneumonia, Coxiellaburneti, Chlamydia pneumonia, Adenovirus, RSV, Influenza A and B also came back with negative results. Urine testing yielded normal results. Factors for thrombophilia (Protein S, antithrombin III, factor II, V, VII, XIII, fibrinogen) – yielded normal results. MTHFR (methylenetetrahydrofolate reductase) mutation.

An MRI scan of the brain revealed bi-frontal involvement with a greater extent in the right frontal cortex. Bilateral extensive hyper-signal areas were observed in the parasagittal frontal cortical, peri and supraventricular regions, attributed to cytotoxic edema in an initial subacute stage, with subtle gyral post-contrast inhomogeneous accentuation. Additionally, chronic microangiopathic changes were noted in the brain's white matter, particularly marked by prominent Vichow-Robin's spaces in the frontoparietal regions.

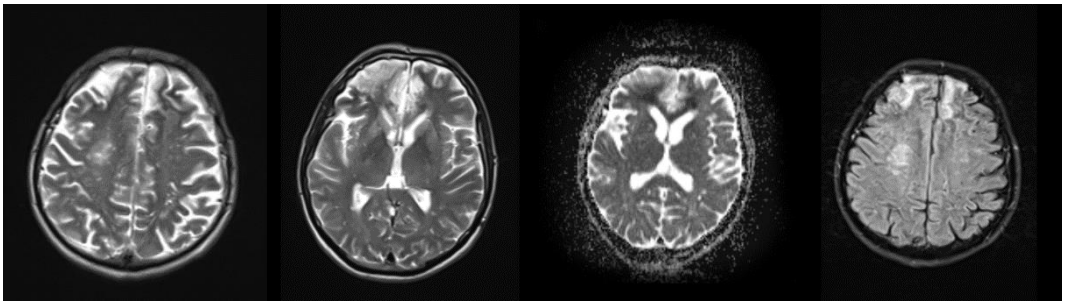

Figure 1. MRI scan of the brain showing bilateral extensivehypersignal areas in the parasagittal frontal cortical, peri and supraventricular regions, with restriction of diffusion.

CT angiography of the carotid and cerebral arteries revealed, on the right side of the terminal branching of the ACA, a gradually diminishing contrast appearance within the lumen of the ACI. This diminishment was observed from the C1 segment down to the C6 segment level. At the C6 segment, a faint

contrast image of the right ACI's C7 segment was visible, and subsequently, the contrast image of the M1 segment was restored, potentially through a right posterior communicating artery branch.

The electroencephalogram indicated a symmetrical, low-voltage brain activity in the anterior beta range and posterior alpha range. A consistent visual block was observed, transitioning into intermittent theta activity within the bilateral frontal leads. Additionally, slow sharp waves emerged on the left side, particularly in the C-T regions.

Carotid Doppler ultrasound was performed, recording a right ACC diameter of 4.7mm with normal flow and flow velocities.

The end-diastole intima-media thickness (ITM) showed a slight reduction at 0.80mm. The ACI starting from its exit, exhibited longitudinal wall thickenings. This wall was thinned with a diameter of 2.7mm, accompanied by diminished and irregular flow, reduced velocities, and initial vascular resistance along its entire length until intracranial entry. The vertebral artery demonstrated physiological anterograde flow with normal flow velocities, possessing a diameter of up to 3.8mm. The left ACC displayed a diameter of 5.4mm with consistent flow and regular flow velocities, along with an IMT of 0.90mm.

At the back wall's bifurcation, a minor isoechoic thickening was observed, extending to the ACI with a thickness up to 2m, displaying consistent functional characteristics. For the transcranial color-coded duplex (TCCD) bubble test using the transtemporal approach, nomicroembolic signals were detected in the ACM following contrast injection.

The test yielded negative results for a right to left shunt.

The follow-up MRI of the brain, using non-contrast TOF, revealed substantial cortico-subcortical and parasagittal hypersignal changes, predominantly concentrated in the bi-frontal region with more pronounced effects on the right side. These changes were accompanied by a slight diffusion restriction.

Comparable alterations were identified in the superior frontal supraventricular region at the centrum semiovale level. These distinctive features of these changes align with those typically observed in the post-ischemic conditions during the subacute stage.

There is a pronounced reduction in the lumen size of the right carotid artery within its proximal intracranial segment.

The carotid artery's appearance and flow are clear at the level of the intrapetrous segment; however, there is notably poor visual depiction of the right carotid artery within this segment.

There is no discernible view of the right carotid artery within the intracavernous segment, nor distally, which also obscures the M1 and M2 segments from view. On the left side, the view of the M1 segment is considerably delicate, with no visual access to the M3 segments and only a reduces perspective of the distal branches. The visualization of the ACA is also notably slender.

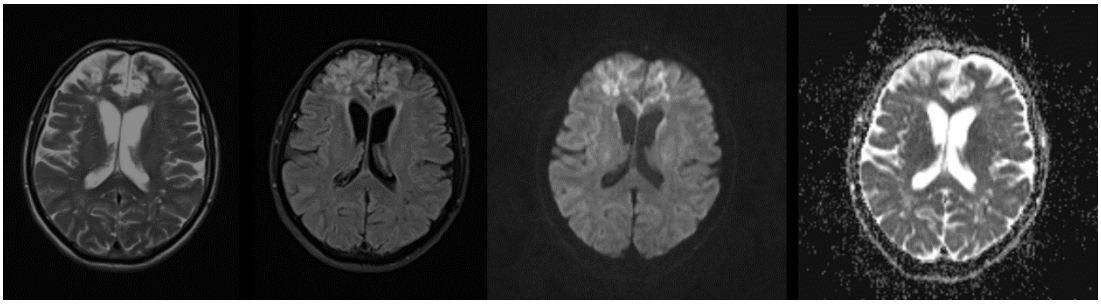

Figure 2. MRI scan of the brain showing cortico-subcortical and parasagittal hypersignal changes, predominantly in the bi-frontal region with more pronounced effects on the right side, accompanied by a slight diffusion restriction.

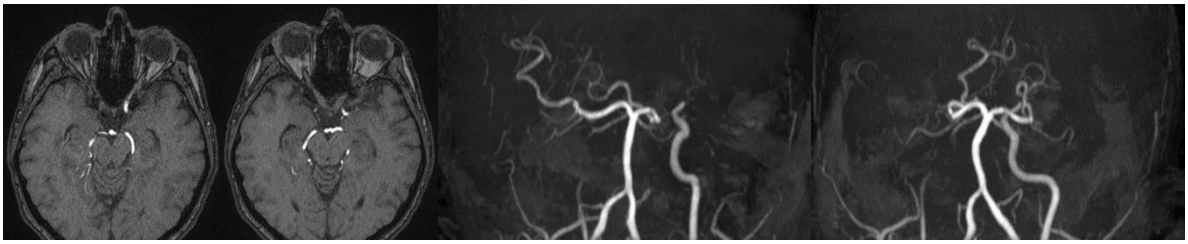

Figure 3. MRI scan of the brain showing no discernible view of the right carotid artery within the intracavernous segment; Markedly inadequate visualization of the anterior circulation.

A psychiatrist was involved in several instances, conducting psychological assessments and proposing therapy geared towards addressing the chronic psycho-organic syndrome. The intended follow-uplumbar puncture was not carried out due to the patient experiencing a panic attack.

The presence of morphologically altered blood vessels, characterized by wall thickening in the right carotid artery along with diminished and irregular flow, reduces flow velocities, and initial vascular resistance spanning the entire length up to the intracranial entry point, does not definitively exclude the possibility that it could be one of the forms of vasculitis.

He received treatment encompassing antiedematous, anticoagulant and corticosteroid therapies, along with antihypertensive, gastroprotective and neuroprotective medications. Despite the comprehensive treatment approach, the symptoms of the psycho-organic syndrome persisted when he was discharged.

Discussion

Vasculitis accounts for a limited number of cases of either hemorrhagic or ischemic stroke [1].

As vascular inflammation impacting the brain is relatively infrequent, and considering that neurological symptoms often lack specificity, arriving at a precise diagnosis can prove to be challenging.

The commencement of primary angiitis of the central nervous system (PACNS) typically manifests insidiously, with a gradual and progressive course. While acute presentations have been documented, they are less prevalent. PACNS symptoms lack specificity and usually multiple symptoms are present in the initial presentation. Foremost among these is headache, frequently accompanied by cognitive impairment and stroke.

Additional initial symptoms might include transient ischemic attack, aphasia, visual disturbances, seizures, ataxia, intracranial bleeding, and amnestic syndrome. PACNS should always be considered a possibility in the cases of rapidly progressive cognitive deterioration and unexplained shift in personality. The challenge in diagnosing PACNS arises from the absence of a definitive biomarker, coupled with the presence of a no-specific symptomatology [2].

Although the identification of multi-locular lesions on MRI, inflammatory laboratory findings in stroke patients, or the detection of intracranial stenosis through CTA, MRA, or angiography may trigger suspicion of this condition, none of these findings is dependable enough to allow a conclusive diagnosis of cerebral vasculitis. While mild systemic symptoms or an elevated CRP are possible in PACNS, organ manifestations other than the CNS are an exclusion criterion for the diagnosis [3,4].

The therapeutic approaches for primary CNS vasculitis mainly draw upon case reports and cohort studies. In most patients, the administration of glucocorticoids has demonstrated symptoms improvement. Although randomized control trials do not encompass cases of PACNS for prognostic insights, data obtained from retrospective observational studies show favorable outcomes when patients undergo corticosteroid therapy [4,5].

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