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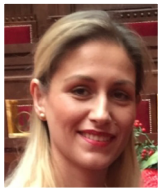
PRES SYNDROME

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Sworn declaration
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SUMMARY

The aim of this review, as well as the case report, is to become familiar with the syndrome, although it is not very common, but may still be encountered by an ophthalmologist during clinical practice. It is also interesting to point out how the clinical unit can be independent and unchangeable in medicine and, on the other hand, in the context of the reversible posterior leukoencephalopathy syndrome (PRES syndrome), the name can be changed. As such, cortex blindness arises after complete destruction of the visual cortex of both occipital lobes, often as a result of vascular circulatory disorders. PRES syndrome is characterized by magnetic resonance imaging or computed tomography, where bilateral irregular hypodensive arteries are present in the occipital lobes that cause transient cortex blindness within the syndrome, which in its name carries the word reversible.
Case report: A patient who was hospitalized at the Pneumology Department in which PRES syndrome and transient cortex blindness were diagnosed.
Key words: PRES syndrome, cortex blindness, acute condition

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INTRODUCTION

Posterior reverse leukoencephalopathy syndrome (PRES syndrome) has a rapid onset of symptoms, including headache, seizures, altered consciousness and dysfunction of vision. It is frequently though not always linked with acute arterial hypertension. If the clinical syndrome is immediately diagnosed and treated, its symptoms usually subside within one week and the changes recorded on magnetic resonance imaging (MRI) disappear during the course of a few days to weeks [1].

PRES syndrome is linked with conditions that exist in patients with diseases of the kidneys such as arterial hypertension, as well as vascular and autoimmune diseases, immunosuppressive therapy and organ transplants. This syndrome is becoming an increasingly more widely recognised disorder, with a broad clinical spectrum of symptoms and triggers, despite which the mechanism of origin has not yet been clarified [2].

CLINICAL SYMPTOMS

PRES appears over the course of a few hours, in which the most common symptoms are epileptic fits, disorders of visual functions, headaches and an altered psychological state. An increase of blood pressure is recorded in more than 70 % of patients with PRES syndrome, although a significant proportion also have normal or only slightly raised blood pressure [3].

The severity of the clinical symptoms may differ. For example, visual functions may be manifested as blurred vision, homonymous hemianopsia or even cortex blindness. Patients may be slightly confused or agitated, but may also fall into a coma. Other rarer symptoms include nausea, vomiting and brain stem deficit. Epileptic fits are frequent [4].

The most common trigger is acute hypertension, although patients often have other comorbidities, which may predispose them to the development of PRES syndrome. Values of systolic blood pressure are usually between 170 mmHg and 190 mmHg, but in 10 – 30 % of patients there is only a slight increase in blood pressure. In PRES syndrome the causes of acute hypertension are presence of acute damage to the kidneys or eclampsia, although hypertensive crisis is stated also in cases of autonomous disruption, such as in Guillain-Barré syndrome or upon the consumption of intoxicating substances. In the large number of cases recording comorbidities of patients with PRES syndrome, more than half of the patients

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had chronic hypertension and 38 % had chronic kidney disease. Autoimmune diseases – including thrombotic thrombocytopenic purpura and systemic lupus erythematosus – were present in 45 % of patients, and exposure to immunosuppressive drugs such as cyclosporine or chemotherapy was present in a similar number, especially in connection with previous transplant [5].

PATHOPHYSIOLOGY

The cause of PRES syndrome remains a subject of controversy, but the most preferred theory is that hypertension causes a disruption of the auto-regulation of the brain. Cerebral blood flow is usually regulated by dilation of blood vessels in order to maintain adequate perfusion of the brain tissue, and thereby at the same time to prevent excessive intracerebral hypertension. Auto-regulation breaks down in the case of above average arterial blood pressure, i.e. of 150 – 160 mmHg. Upon chronic hypertension it occurs at higher values. Uncontrolled hypertension leads to hyperperfusion and damage to the blood vessels of the brain, which leads to interstitial extravasation of proteins and fluids, causing vasogenic edema [6].

Irreversible damage is observed at arterial pressure above 200 mmHg. It is known that conditions that regularly occur in PRES syndrome, such as chronic hypertension and atherosclerosis, reduce the effectiveness of auto-regulation. However, the auto-regulation theory does not explain why blood pressure in PRES syndrome usually does not reach the upper limit of auto-regulation, why PRES syndrome occurs even when the patient does not have hypertension, or why the extent of edema is not directly linked with the severity of high blood pressure. In addition to this, even though this theory indicates brain hyperperfusion, evidence from a number of positron emission tomographies shows early brain hypoperfusion [7].

An alternative theory is that PRES syndrome is the result of a systemic inflammatory state, causing endothelial dysfunction. This is supported by the observation that PRES syndrome is usually linked with a systemic inflammatory process such as sepsis, eclampsia, transplantation or autoimmune disease. Angiography demonstrates reversible focal and diffuse abnormalities, which are assumed to reflect endothelial dysfunction. If blood pressure is high, the vasoconstriction that occurs during auto-regulation could worsen the already existing endothelial dysfunction, which could cause hypoxia and subsequent vasogenic edema. This mechanism would explain why control of hypertension enables recovery [8].

IMAGING METHODS

In an acute state, computer tomography (CT) enables quick assessment. It can also exclude large brain haemorrhage and pathological lesions. Although CT is not 100 % sensitive, it can demonstrate venous sinus thrombosis or arterial ischaemia or thrombosis. In several situations, including PRES syndrome, CT imaging may be normal. Typical MRI findings in PRES syndromes have bilateral abnormalities of white matter in the vascular watershed areas in the posterior regions of both brain hemispheres, which mainly affect the occipital and parietal lobes. Atypical features are common, including haemorrhage, asymmetrical changes, isolated engagement of the frontal lobes and cortical lesions [6].

Upon the use of MRI, it may not be easy to differentiate PRES syndrome clinically from other acute vascular diseases. The diagnosis requires the careful selection of suitable imaging techniques, as well as consideration of the nephrotoxic effects of certain contrast substances. Venous sinus thrombosis may be quickly diagnosed with the aid of CT. Electroencephalography can identify subclinical seizures, and can indicate further causes of encephalopathy. With the aid of lumbar puncture we can diagnose infection or subarachnoid haemorrhage, but this may be within the norm at the beginning of the disease or after treatment with antibiotics [6].

MANAGEMENT

The treatment of PRES syndrome has not yet been evaluated in any clinical trial, but it appears that quick intervention, e.g. the application of antihypertensive therapy, the discontinuation of problematic medications, or treatment according to acute appearing clinical manifestations, speeds recovery and averts complications. Antiepileptic agents should be used for the treatment of epileptic fits. Corticosteroids should theoretically improve vasogenic edema, but there is no evidence concerning their use in PRES syndrome [9].

CASE REPORT

A forty-four year old female patient was admitted to the Department of Pneumology with non-specific interstitial pneumonia, Sjogren's syndrome and rheumatoid arthritis, with chronic hypoxemic respiratory insufficiency on long-term home oxygen therapy. The patient was admitted for the purpose of administering a second cycle of pulse corticotherapy with Solumedrol, which was commenced in June 2019. Upon admittance a stable clinical condition predominated, in the initial laboratory results there was slight elevation of CRP with leukocytosis, medium-severe degree of anaemia, thrombocytosis, which is long-term, X-ray of the chest without any new lesion changes. The patient was administered two doses of physiological solution with Solumedrol 500 mg on 18 and 20 September 2019.

On a ward round at around 10:00 hours on 21 September 2019, the patient reported abdominal pains. Laboratory samples were taken, which showed no pathological finding. At around 16:00 hours the patient stated a deterioration of visual functions, headache, raised blood pressure was determined, with nausea and vomiting. The ECG finding was without pathological changes, Tensiomin 12.5 mg was applied sublingually, and a surgeon and ophthalmologist were called. The surgeon excluded an acute abdomen, and presumed an ulcerative colitis caused by a reaction to Solumedrol. The ophthalmologist examined the patient using direct ophthalmoscopy, and excluded edema of the optic nerve papilla bilaterally, as

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well as other pathology. The patient recorded a significant deterioration of central visual acuity from 1.0 to bilateral questionable light perception. Due to the acute onset of blindness, urgent CT of the head was ordered. On CT bilateral occipital irregular hypodense areas were determined, on the right with a maximum diameter of 25 mm, on the left with a maximum diameter of 35 mm, post-contrast without clear lesion saturation – suspected ischemic lesions. A neurological consultation was called after the result of the CT examination. From the CT examination the patient was transported to the department, transferred to pulmonary ICU for the purpose of monitoring, but the condition was complicated by the sudden onset of blindness. The patient was agitated, blood pressure dropped to 80/45 mmHg, P:107, adrenalin was administered intravenously, blood pressure and P were progressively stabilised, but loss of consciousness and agitation persisted, as a result of which an anaesthesiology consultation was called. The neurologist subsequently stated the need for angioCT for the purpose of investigating the cause of the condition and excluding embolism into the basilar artery. At 19:40 hours the patient was taken for a repeat CT examination, where she was sedated. The result of the repeat CT examination did not demonstrate obliteration in the watershed area of the intracranial arteries. The patient remained at the Department of Anaesthesiology and Intensive Medicine (DAIM) for short-term hospitalisation. After admittance to the DAIM the patient regained consciousness, complied with requests, she was again examined by a neurologist with a diagnostic conclusion of a condition following loss of consciousness, in differential diagnostics suspected secondarily conditioned epiparoxysm, without clear lesion symptomatology, recommendation to add MRI examination focused on the brain and orbits. Due to the increase in temperature, a haemoculture test was conducted at the DAIM with negative result, blood substitution was implemented due to anaemia, administration of chronic medication continued, but a third dose of pulse corticotherapy was not applied. On 23 September 2019 the patient was transferred back to the Department of Pneumology in a stabilised condition. With the exception of slight complaints with vision, the neurological finding was within the norm. MRI examination confirmed suspected PRES syndrome – posterior reversible leukoencephalopathy. During hospitalisation the patient was repeatedly examined by the same ophthalmologist, upon which her central visual acuity improved from original questionable light perception to 20/40 with the patient's own glasses correction. From a pneumological perspective the treatment continued with a maintenance dose of Prednisone due to increase of natriuretic peptide (NT-proBNP), and due to repeated increase of blood pressure with cephalea an internal medicine specialist was consulted, who added a small dose of diuretics and antihypertensive agents, with regular monitoring of blood pressure. The patient was in a clinically markedly improved condition, blood pressure stabilised, upon the recommendation of the internal medicine specialised antihypertensive therapy was progressively reduced until it was completely discontinued. With regard to slight elevation of C-reactive protein (CRP) in comparison with the previous values, a clinical pharmacologist recommended oral therapy with general antibiotics. The patient had cardiopulmonary compensation, with stabilised circulation and no fever symptoms, and was discharged into outpatient care on the 14th day.

DISCUSSION

Interdisciplinary co-operation is very important if various different clinical symptoms appear in a patient. In the case of our patient, who was hospitalised at the Department of Pneumology, a surgeon was called, followed by an ophthalmologist, neurologist, internal medicine specialist and anaesthetist. Timely distinguishing of symptoms is the foundation of early diagnosis. PRES syndrome, with atypical symptoms, is difficult to determine correctly, which may lead to late diagnosis or incorrect choice in the management of the patient, and subsequently to irreversible clinical development. As a result, it is essential for doctors to have a better understanding of the clinical and MRI characteristics of this pathology. After the diagnosis of PRES syndrome, the treatment, which especially includes adjuvant therapy and removal of the cause, should commence immediately in order to prevent adverse progression. After prompt and correct treatment, the clinical condition has improved and the pathological findings on CT and MRI examination have been stabilised in the majority of patients within the course of 2-3 weeks. The clinical symptoms include headaches, psychological dysfunction, seizures, blurred vision and other neurological symptoms. In our patient the first symptoms were nausea, abdominal pain and progressive deterioration of visual functions, which were also added to by raised blood pressure. The area of the lesion is limited mainly to the parietal and occipital lobes, while the frontal lobe, basal ganglia, temporal lobe, corpus callosum and cerebellum may also be affected. The prognosis is good in the case of timely and commensurate treatment [10]. Similarly as described in the literature, in our patient also the occipital lobes were affected in PRES syndrome.

A study conducted by Kini describes a 26 year old female patient in whom HELLP syndrome was manifested during pregnancy – haemolysis, elevated liver enzymes and low platelet levels – together with PRES syndrome. The coincidence of these two pathologies in one patient may be manifested in anterior or posterior disorder of visual functions, and can be evaluated as temporary cortex blindness, which can be demonstrated with the aid of CT or MRI examination or as a manifestation on the eye itself as serous elevation of the retina with attendant deterioration of visual functions [11].

CONCLUSION

In ophthalmology also it is necessary to be informed about suddenly appearing conditions, including not only injuries, endophthalmitis or retrobul-

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bar haematomas, but also sudden loss of central visual acuity. It frequently occurs that as ophthalmologists we are called upon to attend a consultancy examination on patients who do not only have ophthalmological complaints. Our patient had various different symptoms, which included a deterioration of visual functions. Precisely for this reason it is necessary to have knowledge, however basic, of acute conditions also in other fields. Only through such interdisciplinary co-operation will we then be able to respond promptly and correctly in diagnosis and adequate treatment.

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