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RESEARCH ARTICLE

OPHTHALMOLOGIC MANIFESTATIONS REVEALINGPOSTERIORREVERSIBLEENCEPHALOPATHY SYNDROME: A CASE REPORT

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is a rare clinico-radiological syndrome, secondary to many diseases especially high blood pressure. First described by Hinchey in 1996, pathophysiology is not clear. Clinical manifestations are headache, seizures, altered mental state such as confusion or coma, and also visual disturbances, such as hemianopia, blurred vision, diplopia, or cortical blindness. We report the case of a Moroccan 47-years old patient who arrived at the ophthalmology emergency presenting a sudden vision loss and horizontal diplopia, associated with headache. General examination found a high blood pressure at 240/110mmhg. The funduscopy of both eyes found hypertensive retinopathy. A brain MRI was realized showing bilateral occipital cortex T2/fluid-attenuated inversion recovery (FLAIR) hyperintensities. The diagnosis of Posterior reversible encephalopathy syndrome was retained.

Introduction:-

Posterior reversible encephalopathy syndrome (PRES) is a rare clinical and radiological syndrome. It is secondary to many diseases, especially high blood pressure [1]. It was described by Hinchey in 1996[2]. Pathophysiology is not yet elucidated. About the clinical presentation, it includes not only neurological symptoms such as headache, seizures, altered mental state, but also ophthalmological signs which can reveal the diagnosis. We report the case of a 47-year old woman who consults for ophthalmologic signsrevealing PRES.

Patient and Observation:-

We report the case of a Moroccan 47-year old patient with a history of psychiatric illness, and chronic kidney failure with dialysis, who arrived at the ophthalmology emergency presenting a sudden vision loss and horizontal diplopia, associated with headache.

General examination found a high blood pressure at 240/110mmhg. On the ophthalmologic examination, visual acuity was 3/10 in both eyes. The pupillary light reflex was present. Without the restriction of her ocular motility, she showed 10 prism diopter of exotropia in all gaze directions. Intraocular pressure was normal. Slit-lamp biomicroscopic showed normal anterior segment. After dilatation, the funduscopy of both eyes found tortuous arterioles, retinal hemorrhage, and cotton wool spots concluding hypertensive retinopathy (Figure 1).Lancaster test

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objectives horizontal binocular diplopia. With neurological symptoms, a brain MRI was realized emergently showing bilateral occipital cortex T2/fluid-attenuated inversion recovery (FLAIR) hyperintensities(Figure 2). The diagnosis of Posterior reversible encephalopathy syndrome was retained.

The patient was hospitalized in the neuro-ophthalmology department for monitoring of blood pressure, and management of her hypertension.

After two weeks of hospitalization, visual acuity improved with the disappearance of diplopia. Follow-up after 1 month was uneventful.

Discussion:-

Posterior reversible encephalopathy syndrome (PRES) is a rare clinico-radiological syndrome, secondary to many diseases especially high blood pressure [1]. First described by Hinchey in 1996 in the « New England Journal of Medicine » [2], it is characterized by acute cerebral endotheliopathy with consecutive disruption of blood-brain barrier and vasogenic edema predominantly in the parieto-occipital region of the brain [3].

Pathophysiology is not clear, but acute cerebral endotheliopathy might be triggered by severe hypertension that exceeds the threshold of cerebral autoregulation, by endogenous or exogenous toxins such as chemotherapy or immunosuppressant drugs, or by severe cerebral vasoconstriction and spasms induced via neuropeptides [3].

Clinical manifestations are headache, seizures, altered mental state such as confusion or coma, and also visual disturbances, such as hemianopia, blurred vision, diplopia, or cortical blindness. Only 27% of patients with a formal diagnosis of PRES had any sort of visual complaint, making the vision a poor diagnostic criterion for the syndrome. Table 1 resumes the visual symptoms associated with PRES and determines visual outcomes following treatment in a retrospective study about 41 patients made by Lifson et al [4].

Since its initial description, PRES has been linked to an increasing number of medical conditions or medications. A list of possible causes and contributing factors are shown in Table 2 [5].

Radiologically, the classic, so-called ‘typical’ MRI findings are symmetric (reversible) T2 or fluid-attenuated inversion recovery imaging (FLAIR) hyperintensities located in the occipital and parietal region caused by vasogenic edema [3]. But other locations were common including the frontal lobes, inferior temporal lobes, and cerebellar hemispheres [6]. Involvement of the basal ganglia, brain stem, and deep white matter including the splenium was not rare [6].

In most cases, PRES has a good short-term and long-term prognosis if treated fast and adequately and MRI findings and clinical symptoms are reversible after several hours or days [7]. PRES has to be treated early, adequately, and sufficiently long. The most important steps in acute clinical management are confirmation of diagnosis, removal of the underlying cause, for example, discontinuation of immunosuppressant drug intake, and lowering of elevated blood pressure if moderate to severe hypertension is present.

Conclusion:-

PRES is a very rare clinical and radiological syndrome. Its pathogenesis remains not clear though its association with malignant hypertension. Management of PRES includes removal of any offending agents, blood pressure, and seizure management.It should be considered in all patients presenting visual disturbances. A complete ophthalmologic examination should be realized with a complement of biological assesment and radiological exams.

Competing interests:
The authors declare no competing interests.

Informed Consent
We obtained informed consent from the patient to use the images in this case report.

Patient perspective:
She no longer has diplopia and visual acuity improved. She was satisfied with the treatment received.

List of tables:

Table 1:-Visual symptoms associated with PRES and determine visual outcomes following treatment [4].

Pt.	Visual symptoms	Visual field	Visual acuity	Outcome
1	Diplopia	NA	20/40 OU	Full resolution
2	OU vision loss	OS superior loss	20/50 OD, 20/200 OS	Full resolution
3	OS photophopsias	NA	20/20 OU	Full resolution
4	OU vision loss	Full OA	20/20 OU	Full resolution
5	Diplopia, colour desaturation	NA	NA	Full resolution
6	OU vision loss	NA	NA	Full resolution
7	OS vision loss, OS pain with EOMs	Full OA	20/20 OD, 20/40 OS	Full resolution
8	OU vision loss	Full OA	CF OU	Full resolution
9	OU vision loss	Left lower quadrantanopia	NA	Full resolution
10	OU vision loss	NA	NA	Full resolution
11	OU vision loss, diplopia	OU diffuse depression	LP	Intermittent binocular diplopia

PT, patient; NA, not assessed; OU, right and left eyes; OS, left eye; OD, right eye; EOMs, extraocular muscle movements; CF, counting fingers; LP, light perception.

Table 2:-Causes and contributing factors of PRES [5].

Hypertensive encephalopathy	Acute or chronic renal diseases Vasculitis: systemic lupus erythematosus, polyarteritis, nodosa Wegener's Endocrine disorders: pheochromocytoma, primary aldosteronism Porphyria Thermal injury Scorpion envenomation Cocaine or amphetamines abusers Over-the-counter stimulants: phenylpropanolamine hydrochloride ephedrine pseudoephedrine caffeine
Eclampsia	Carotid dissection Hyperperfusion syndrome Thrombotic: thrombocytopenic, purpura Hemolytic and uremic syndrome Guillain–Barre´ syndrome Triple-H therapy
Immunosuppressive drugs	Ciclosporin A Tacrolimus Sirolimus Vincristine Cisplatin Cytarabine L-asparaginase Gemcitabine Bortezomib Bevacizumab Intrathecal chemotherapy Combination chemotherapy

Other drugs	Interferon-a TNF-antagonist Immunotherapy with interleukin Antiretroviral therapy in HIV-infected patients Erythropoietin Granulocyte stimulating factor Intravenous immunoglobulin
Other conditions or associations	Reversible cerebral vasoconstriction syndrome Infection/sepsis/shock Blood transfusion Tumor lysis syndrome Cholesterol embolism syndrome Hypomagnesemia Hypercalcemia Hypocholesterolemia

List of figures:

Figure 1:-Funduscopy of both eyes showing hypertensive retinopathy: tortuous arterioles, retinal hemorrhage, and cotton wool spots.

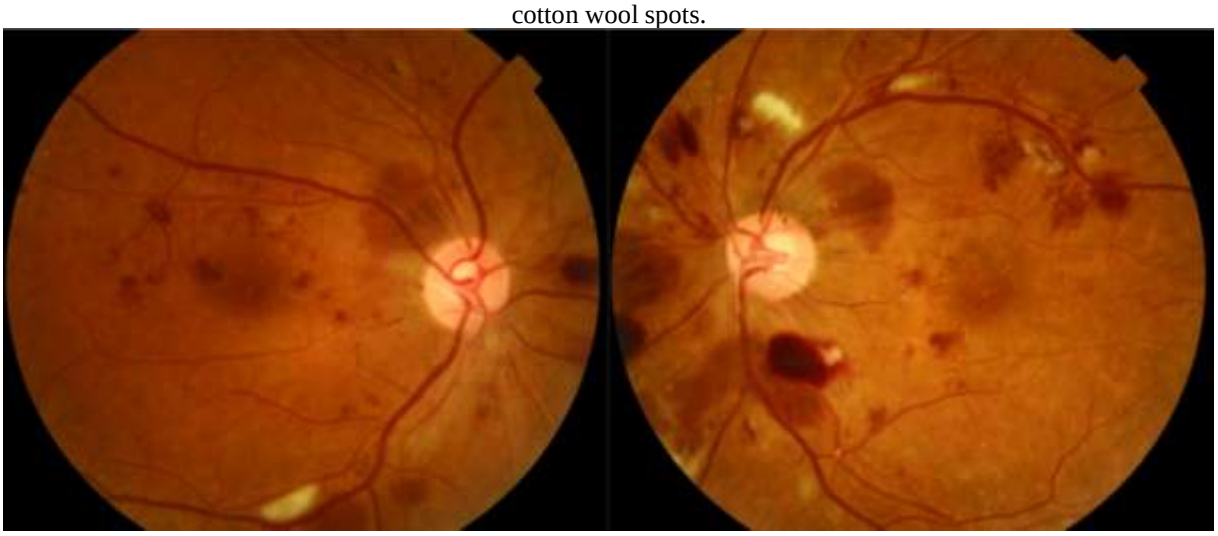
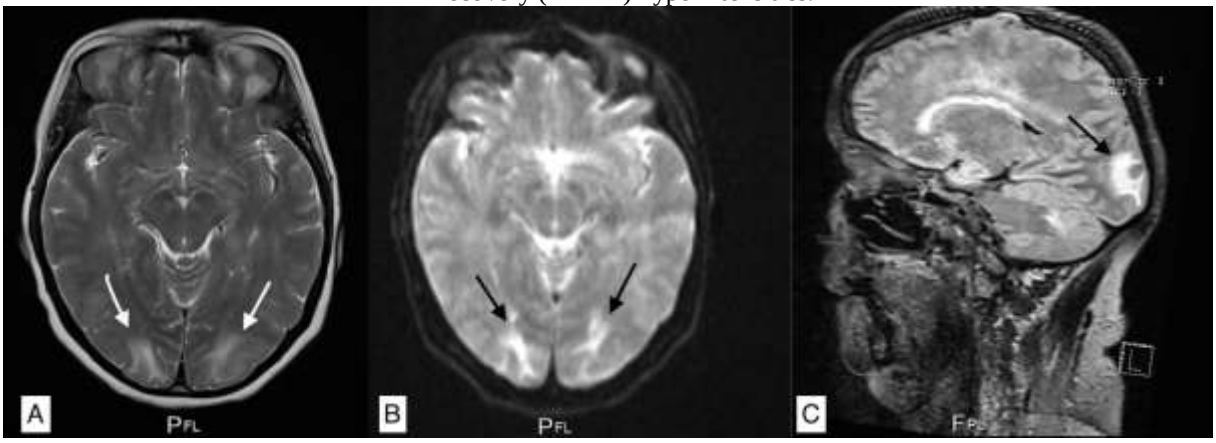


Figure 2:- Brain MRI. Axial view (A+B) Sagittal view (C). bilateral occipital cortex T2/fluid-attenuated inversion recovery (FLAIR) hyperintensities.



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