Neurosciences Quiz

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A woman with seizures and papilledema

Case Presentation

A 38-year-old woman, known case of rheumatoid arthritis and hypertension, presented with one day history of headache, blurred vision and seizures. Clinically, she was afebrile with high blood pressure (190/100 mm Hg). She was confused and disoriented. She had papilledema with normal motor examination. Reflexes were exaggerated with bilateral Babinski signs. An MRI of the brain is shown (**Figure 1**).

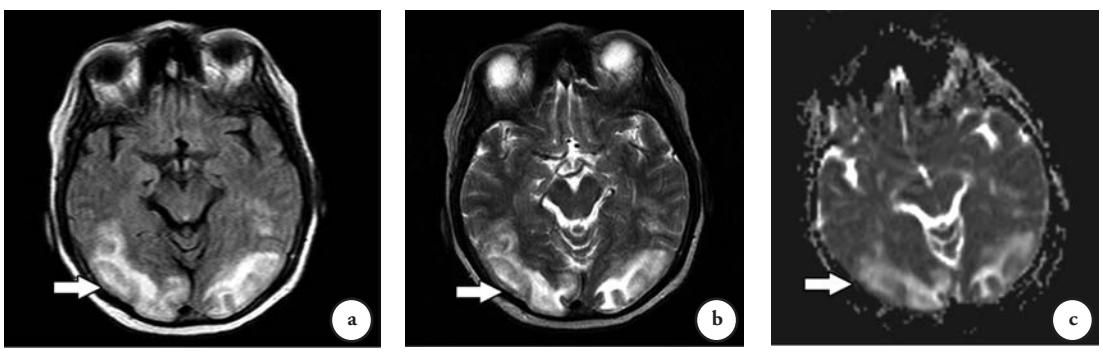

Figure 1 - Patient MRI images.

Questions

1. What are the principal abnormalities? What is the possible diagnosis?

2. What are the other features of this disorder? What are the causes?

3. What is the pathophysiology?

4. What is the treatment? What is the prognosis?

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Answers & Discussion

1. There are bilateral symmetrical changes in the parieto-occipital regions, which are of high signal intensity on both fluid attenuation inversion recovery (FLAIR) images (**Figure 1a**) and T2WI (**Figure 1b**), and of low signal intensities on T1 weighted-images (not shown). These changes are obvious on ADC map images (**Figure 1c**). The most likely diagnosis is posterior reversible encephalopathy syndrome (PRES).

2. The clinical syndrome of PRES is characterized by headaches, altered consciousness, visual disturbances, and seizures.[1] The headache is typically constant, moderate to severe, non localized, and unresponsive to analgesia. Altered consciousness ranges from mild somnolence to confusion and agitation, progressing to stupor or coma in extreme cases. Seizures are often the presenting manifestation with a tendency to recur. They are usually generalized tonic clonic but may begin focally. Status epilepticus has been reported. Preceding visual loss or visual hallucinations suggests occipital lobe origin in some patients. Visual disturbances include hemianopia, visual neglect, auras, visual hallucinations, and cortical blindness. The funduscopic examination may show papilledema with accompanying flame-shaped retinal hemorrhages and exudates but could be normal, particularly in eclamptic, and chronically hypertensive patients. The deep tendon reflexes are frequently brisk with Babinski signs often present. A few patients may have weakness and incoordination of the limbs, but other focal neurologic deficits are rare. Hypertension is frequent but not invariable. The causes of this condition are diverse, but common precipitants include the acute elevation of blood pressure and hypertensive encephalopathy, eclampsia, immunosuppressive drugs, and renal failure.

3. The pathogenesis of PRES remains unclear, but it appears to be related to disordered cerebral autoregulation and/or endothelial dysfunction. Because of the heterogeneous nature of this disorder, it is possible that different mechanisms are etiologically important in different clinical situations. To maintain the constant cerebral blood flow, cerebral vasoconstriction occurs in response to hypertension, normally via sympathetic innervation. However, when blood pressure is elevated beyond the limits of cerebral autoregulatory capacity, passive vasodilatation, and hyperperfusion occur resulting in breakdown of the blood-brain barrier and leakage of protein and fluid into the brain parenchyma (vasogenic edema). This is particularly seen in the posterior region of the brain, probably due to relative lack of sympathetic innervation.[2] The rate of blood pressure elevation is likely to be important. In chronic hypertension, adaptive vascular changes "reset" the range of autoregulation to higher systemic blood pressures. As a result, patients with PRES in the setting of longstanding hypertension may have markedly elevated blood pressures, while less severe elevations, even normal blood pressures, are associated with PRES in other settings. Children appear particularly vulnerable to PRES at lower blood pressures than adults. Endothelial dysfunction has also been implicated in the pathophysiology of PRES, especially in normotensive individuals or cases associated with pre-eclampsia or cytotoxic therapies (with nontoxic levels of these drugs).[3] Direct toxicity on vascular endothelium leads to capillary leakage, blood-brain barrier disruption, and axonal swelling, which may then trigger vasogenic edema. Based on pathologic data and radionuclide imaging, cerebral ischemia is not believed to play a major pathophysiologic role in most patients with PRES. Most patients have complete reversibility of clinical and neuroradiographic findings as seen in our patient.

4. Since PRES is usually reversible on prompt treatment, physicians should have a high clinical suspicion in the appropriate settings (pregnancy, cytotoxic therapy, hypertensive patients, and so forth). Hypertension should be promptly treated. The initial aim of treatment in malignant hypertension is to rapidly lower the diastolic (blood pressure) with the maximum initial fall not exceeding 25% of the presenting value. The use of an easily titratable parenteral agent such as labetalol is effective and safe in reducing the blood pressure to a desirable range. For patients with lower levels of hypertension, lowering blood pressure is also recommended, but treatment recommendations are somewhat limited. Seizures are treated with phenytoin, however, other antiepileptic drugs are effective and may be preferred, depending on the patient's other comorbid medical conditions and prescribed drugs. In eclampsia, magnesium is used to treat seizures and is superior to phenytoin and diazepam. Reduction in drug dosage or prompt removal of the cytotoxic drug is usually recommended in cases of PRES associated with cytotoxic agents. Comorbid conditions including electrolyte disturbances,

fluid overload, uremia, and sepsis should be treated aggressively. Prognosis is usually good with most patients recovering within 2 weeks. A small number have residual neurologic deficits resulting from secondary cerebral infarction or hemorrhage, and some patients die as a result of increased intracranial pressure or as a complication of the underlying condition.[4]

References

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