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Sir,

Reply to Tan *et al*

We welcome and thank Tan *et al*[1] for their interest and observations regarding our previously published case report.

We are in agreement regarding Snellen acuity not being a diagnostic criteria for Charles–Bonnet Syndrome (CBS). Numerous reports have highlighted the presence of CBS-type hallucinations in individuals with good Snellen acuity with restricted visual fields. It appears, therefore, that the quality of visual function and its deviation from the previous norm for the individual are key elements in production of CBS hallucinations. It is for this reason that we agree with the observation by Holroyd *et al*[2] that sudden and abrupt reduction in visual function may trigger CBS symptoms.

Tan *et al*[1] comment on reduction of light perception in the fellow eye terminating CBS symptoms in our case. We agree that the proposed theory could account for resolution of CBS on eye closure. This theory, however, is similar to the other commonly perceived theories of deafferentation described by Bartlett[3] and West.[4] The visual sensory cortex, when deprived of normal afferent input may exhibit spontaneous independent activity with resultant conscious images. This hypothesis is supported by the observation that such hallucinations may be abolished by normal or excessive visual stimulation.[3] The perceptual theory of West describes afferent input reduction below a threshold level (such as in disease of the eyes or visual pathway), resulting in the brain allowing previously registered subconscious perceptions to emerge into consciousness, resulting in a hallucinatory experience.[4]

Similarly, as the hallucinations of CBS have only been documented in acquired visual deprivation, it is possible that in-built higher centre mechanisms are in place so that eye closure may trigger intracerebral activity suggesting to the sensory visual cortex that visual stimulation has ended, rather than sensory input itself being detected by sensory mechanisms to be decreased. This may explain why darkness with the eyes open may still result in CBS but darkness as a result of eye closure may obliterate CBS hallucinations.

References

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Sir,

Idiopathic intracranial hypertension associated with depot medroxyprogesterone

Many associations of idiopathic intracranial hypertension (IIH) with various medications have not been well substantiated. Adequate proof of association requires that withdrawal of the medication decrease the CSF pressure in patients with IIH. This report describes a 23-year-old woman, who developed IIH after using depot medroxyprogesterone acetate and experienced total resolution of IIH after discontinuation of this medication.

Case report

A 23-year-old woman with a body mass index of 28.2 kg/m^2 presented with a 3-week history of retro-orbital pain and headaches associated with horizontal diplopia, photopsias, tinnitus, and nausea. Her past medical and family histories were unremarkable. Her only medication was contraception with depot medroxyprogesterone acetate (Depo Provera), which she had taken only one dose 2 months ago.

Her visual acuity was 20/20 OU with a right absolute central scotoma. Pupillary responses and color vision were normal. She had a 10 prism diopter right esotropia. Tonometry revealed 17 mmHg OU. Anterior segment examination was normal. She had bilateral papilloedema with nerve fibre layer haemorrhages in the superior peripapillary regions OD and punctate haemorrhages in the medial inferior peripapillary regions OD. No macular star was present.

MRI of the brain and orbits with and without contrast and MR venogram were normal. Her CSF opening pressure was elevated at 520 mm H_2O. CSF protein, glucose, cell count, VDRL, Gram stain, acid fast bacilli stain; routine bacterial, viral, and fungal cultures; and cytology were all normal. Laboratories including complete blood count with platelets, chemistry panel, liver function tests, PT/PTT/INR, sedimentation rate, antinuclear antibodies, RPR, VDRL, rheumatoid factor, and angiotensin-converting enzyme were normal. Other autoimmune disorders were ruled out with negative anti-double-stranded DNA, anti-SSA, anti-SSB, anti-Ro, and anti-Jo1 antibodies. Laboratory investigation, including protein C and S, antiphospholipid, anticardiolipin, and antiphosphatidylserine antibodies, did not reveal a hypercoagulable state. T3, T4, thyroid stimulating hormone, and antithyroglobulin antibodies were also normal. Urine beta human chorionic gonadotropin test was negative.

After developing IIH, she elected to discontinue this medication and not take any further ones, including carbonic anhydrase inhibitors or corticosteroids for the treatment of her IIH. After the fourth monthly visit, she had total resolution of her papilloedema and related symptoms. Repeat lumbar puncture revealed a decreased CSF opening pressure of 131 mm H_2O. Her body mass index remained the same.

Comment

Several reports have described IIH after standard oral contraceptives, containing 20–30 μg of ethinyl estradiol (EE) and after emergency contraception with 200 μg for EE over 2 days.[1] Unlike levonorgestrel implants,[2,3] medroxyprogesterone acetate has not been clearly associated with IIH. Only one report in the literature describes a 29-year-old woman who developed IIH 8 weeks after an injection of medroxyprogesterone acetate and received oral dexamethasone without follow-up data regarding resolution of her symptoms after withdrawal of medroxyprogesterone.[4]

This report demonstrates more convincing evidence linking depot medroxyprogesterone acetate with the development of IIH. Withdrawal of this medication in the patient resulted in complete resolution of IIH, as evidenced by a normal neuro-ophthalmic examination and a CSF opening pressure of 131 mm H_2O after 4 months. This wash-out period is consistent with the half-life of the medication. The serum level from a recommended dose of medroxyprogesterone 150 mg intramuscularly usually increases over 3 weeks to reach peak plasma concentrations between 1 and 7 ng/ml. The levels then exponentially decrease until they become undetectable ($<$100 pg/ml) between 120 to 200 days following injection.[5]

The close temporal relationship between the use of medroxyprogesterone acetate and development of IIH, and the resolution of IIH after medication withdrawal are only suggestive of a causal relationship. The exact pathogenesis of IIH from progestins remains unclear. Increased awareness of this adverse effect is important, as more women are using this medication for contraception and for menstrual irregularities because of its more convenient dosing of once every 12–13 weeks and its lower cost compared to oral contraceptives.