Charles Bonnet syndrome: An under reported entity in endocrinology

Sir,

Charles Bonnet’s syndrome (CBS) or “phantom eye syndrome”, described in 1760, owes its name to a Swiss philosopher who reported visual hallucinations (VH) due to visual impairment in people who are mentally healthy. It is a benign phenomenon affecting 1.4 to 40% of patients with ophthalmopathies. The well structured, permanent or intermittent, simple or complex VH, can take several aspects: geometric figures, personages, landscapes, animals and objects.

CBS affects old people with macular degeneration, glaucoma, diabetic retinopathy, and cataracts. Young persons with damaged visual cortex/optic pathways by a pituitary tumor (PT) are also concerned as in this case:

A male aged 24, consulted for epilepsy due to invasive mixed PT secreting PRL and GH. Medical history of headaches began when he was 10. Clinical exam argued for Acromegaly-gigantism with frontal, pyramidal, and vestibular syndrome. Ophthalmologic exam showed optic atrophy. After surgery, he was almost blind. Under bromocriptine (35 mg/day), and somatostatin analogues, the vision improved discreetly, neurological deficit disappeared, and the tumor size was reduced [Figure 1]. But, when questioned about bromocriptine side effects, he complained of clear visions of trees, cars, and televisions which persisted on closure of the eyes. The patient was not depressed. Conscious of the unreality of VH, he affirmed they were present before starting bromocriptine therapy.

Hallucinations are generally defined as “perception without any object to perceive”. Toxic causes, sensory deprivation, and altered states of consciousness may cause VH. In our patient a psychiatric origin was improbable as psychiatric

Figure 1: (a) Before treatment: Giant (90×70×68 mm) and multi directional somatolactotrop tumor (PRL=8170ng/ml, n<15, and GH=1430ng/ml, n=5). (b) spectacular reduction after Bromocriptine and somatostatin analogues.
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expertise ruled out this etiology. A neurological cause was also discussed because of cerebral invasion [Figure 1], but the hallucinations remained after tumor shrinkage. Toxic origin due to high dose bromocriptine seemed more probable. But, the phenomenon appeared before and persisted after stopping the drug. So the diagnosis of exclusion was CBS, as defined by the following triad: 1) Complex visual hallucinations consisting in clear, organized and well defined images on which the subject cannot exert any control, 2) eye disease causing vision deterioration and sensory deprivation, 3) preserved cognitive status: the subject being aware of the unreality of his visions. For most authors disappearance or persistence of VH on closing eyes is not a major sign.

The mechanism of this syndrome is poorly understood. But, if we consider similarities with “phantom limb syndrome” in which an individual can perceive sensation and even pain signals of a limb that has been amputated, one can understand that a rapid de-enervation (secondary to cones and rods deterioration, or brain dysfunction) induces an interruption in visual input which in return induces an over activity of sensory cells that continue to generate perceptual visions stored in the brain.[3-5]

For treatment, antipsychotics and selective serotonin reuptake inhibitors can be used with variable results. But, in general patient reassurance about the benign phenomenon which could disappear after 18 months, and functional re-education (avoiding lighting and being alone, closing and opening eyes) remain the mainstays of treatment.[1-5]

Endocrinologists should be aware of CBS, because VH are not mentioned spontaneously and are a differential diagnosis of bromocriptine side effects.

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