Arq Neuro-Psiquiat (São Paulo) 47(2):230-234, 1989

BONNET SYNDROME AND POSTERIOR PARASAGITTAL TUMOR

CLUES TO NEURAL MECHANISMS

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SUMMARY — A case of Bonnet syndrome associated with blindness due to bilateral eye disease and a posterior parasagittal meningioma is reported. It is assumed that visual afferent deprivation alone is not enough to produce the syndrome and that, in most instances, a 'cerebral factor' must be operative if hallucinoses are to occur. The distinction between hallucinosis and hallucinations is favored and a common neural circuit for the mediation of hallucinotic imageries in general is suggested. One should not immediately put the blame on obvious eye or visual pathways affections when facing cases of Bonnet syndrome, as they are not likely to explain the complex array of images perceived by any given patient. On the contrary, the possibility of a clinically covert intracranial disease should be always raised and intensively looked for.

Síndrome de Bonnet e tumor parassagital posterior: indícios dos mecanismos neurais.

RESUMO — Relatamos o caso de um paciente com síndrome de Bonnet com cegueira ocasionada por doença ocular bilateral e meningeoma parassagital posterior. Admite-se que, isoladamente, a deprivação visual aferente é insuficiente para produzir a síndrome e que, na maior parte dos casos, um 'fator cerebral' deve operar para que a alucinose aconteça. Favorecemos a distinção entre alucinose e alucinação e sugerimos que um circuito neural comum medeie as imagens alucinóticas de modo geral. Não parece prudente responsabilizar de imediato afecções oculares ou ópticas óbvias quando diante de casos de síndrome de Bonnet, pois, provavelmente, não justificarão as imagens complexas percebidas pelo paciente. Ao contrário, deve-se investigar exaustivamente a possibilidade de doenças intracranianas clinicamente inaparentes.

The occurrence of hallucinosis in blind areas of the visual fields was first described in 1760 by Charles Bonnet (1720-1793), Swiss naturalist and philosopher, on his'Essai Analitique sur les Facultés de l'Âme'. Since then, many identical cases have been documented, and in 1935 Morsier proposed to collect such cases under the eponym 'Charles Bonnet syndrome' [19] or, simply, 'Bonnet syndrome' (BS). Clinically, a gross defect of at least a significant portion of one or both visual fields seems to be a prerequisite for the emergence of the hallucinosis, which tends to be restricted to the amblyopic areas [14]. The classic observations were made on old patients who had become blind due to bilateral cataracts, but it appears now certain that visual failure resulting from injuries anywhere along the optic pathways, from the eye properly up to the calcarine cortex, may be associated with this same symptom-complex [25]. Like 'hallucination', 'hallucinosis' implies perception in the absence of external physical percepts [3]. However, sharply contrasting with the former, the latter term implies, further, that the individual detains clear consciousness and insight regarding the 'falseness' or 'unreality' of what is being perceived, besides being capable of telling what is truly real or not among the variety of objects which com-

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pound his visual world [21]. The content of the hallucinosis, on its turn, is made up of complex, vivid and colored naturally appearing images of landscapes, human faces, unknown people, crowds, pets, buildings, beasts, trivial objects of daily life, and the like. These may be animated by peculiar movements or expressions which lend them corporality, liveliness and seeming outward and autonomous existence. Yet, they can be neither touched nor heard to make sounds of any kind. Rarely are they frightening and the majority of patients communicate a feeling of indifference or even amusement towards them. The apparitions may be continuously present or supervene episodically many times a day or just on a few occasions throughout the whole life of the individual, even though the basic condition that has led to blindness persists. Neuroleptics do not exert any effect on them, but in some instances antiepileptic drugs do [23]. Writers do not always respect this distinction, referring to any kind of perception in the absence of external stimuli as an hallucination [3]. We favor the view that some difference does exist, though no sharp boundaries between them in practice can always be drawn. What we have characteristically seen in hallucinotic patients is that many times they react appropriately to the hallucinoses as if they were real (thus, like really hallucinating), but after they are gone, their reality is almost always denied. At other times still, patients seem to infer that what are seeing just cannot be real. In other words, no distinction seems to exist between a visual hallucination and a visual hallucinosis in what concerns their pictorial content and the behavioral reactions they may induce on the patient. However, we have been impressed by the emotional attitude of patients in one case and the other, as if the critical matter which distinguishes them is their irreducible belief' in the reality or falseness of the perceived non-existent images.

No case of BS in Brazilian medical journals could be found. Besides, the association of the syndrome with intracranial tumor has been seldom noted. It thus seems opportune to report one new case and recall some of the basic features of such a peculiar condition.

CASE REPORT

JF, an 86-year-old white, widowered engineer born in Minas Gerais, came to consultation in December 1987 complaining of 'visions'. At the age of 20, already coursing the fourth grade of the Politechnic School of Rio de Janeiro, he casually occluded his right eye, then realizing he could not see with the left eye alone. In the absence of antecedents of ophtalmic disease, a diagnosis of congenital chorioretinitis was made. About this epoch, he was recognized to suffer from daltonism too, owing to the fact that he was unable to distinguish certain types of minerals and stones solely on the basis of their colors. In 1950, at the age of 56, spectacles for astigmatism were prescribed by an ophtalmologist. Along the following years, visual accuity went on declining, despite regular use of eye drops and a new examination four years before consultation disclosed involvement of the peripheral vision of the right eye due to glaucoma. On this occasion, he was submitted to prostatectomy. In the immediate postoperative period, he felt his vision abruptly further decline, a right 'retinal hemorrhage' being diagnosed. Since then, he became totally blind, only capable of seeing 'a circle of clarity' if lights were turned on in a previously darkened room.

Soon after surgery and although blind, he unexpectedly read two telegrams printed on a newspaper leaf, one concerning President Tancredo Neves and the other, President Reagan, respectively. He perfectly recalled them to be lengthy. In time, the 'visions' used to coming on daily, lasting from a few minutes up to a couple of hours, with no predictable time schedule to recur nor any identifiable associated triggering factor. Their content varied, yet it used to obbeying to certain patterns which came and went at irregular intervals. Often, he saw a hut at the top of a hill and animals, as bears, tigers and chickens with chicks. Moreover, naked women, faces of strangers and 'beautiful' landscapes, such as lawns, a crystalline river flowing next to his legs and the Guanabara Bay, were also commonly perceived. The apparitions, from....., which no sound, smell or tactual impressions could be derived, were animated by lively and naturally appearing colors, with sharp contours and details, undistinguishable from real perceptions. In no moment, however, did he believe in the reality of such 'stuffs', seeming bewildered when asked to offer them a plausible explanation. Most of the time, they were 'nice' and might be even wanted, excepting a few animals at sexual intercourse, which were felt as 'disagreeable'. Only one of such visions was not colored and, significantly, portrayed a black-and-white movie. Lately, a 'kind of factory' full of machineries, wheels, pulleys and ropes that made him feel uneasy, was being seen. Another frequent late vision was one of cattle ('many oxes in the midst of dust'). Sometimes these experiences were reported in laughs and giggles.

Antecedents included two grandsons with daltonims, two with keratoconus and another one with congenital cataracts. His clinical and neurologic examinations showed him to be lucid and oriented with integrally preserved memory and thinking operations. An ophtalmologic examination revealed a blind man who could discern brightness from darkness only with the left eye. Intraocular pressures were 38 mmHg in the right eye and 12 mmHg in the left (normal = 12 to 18 mmHg), respectively. The right optic nerve head was atrophic, the left could not be seen because of cataract. Pupillary reflexes were abolished. There was resting tremor of both hands magnified by the finger-to-nose test bilaterally. Eye movements were full. No meningeal signs were noted. The remainder of the examination was normal. Laboratory tests were all normal, including ECGs, plain skull and chest X-rays, carotid duplex-scans and urinalysis. An EEG exhibited sparse sharp and theta waves, most evident in the left temporal areas. A CT scan of the head showed an enhancing parasagittal ball-shaped mass measuring around 3.2 cm in diameter, close to the falx, in contact with the medial surface of the left parietal lobe above the corpus callosum, unaccompanied by mass effects on the lateral and third ventricles, which conserved their anatomic shape and volume. The falx and the pineal body were calcified, the sylvian fissures widened. The posterior fossa and its structures showed no signs of abnormality. Radiologic diagnosis was falx meningioma (Fig. 1). The patient was not operated on and died on July 1988 with no changes in his hallucinotic status.

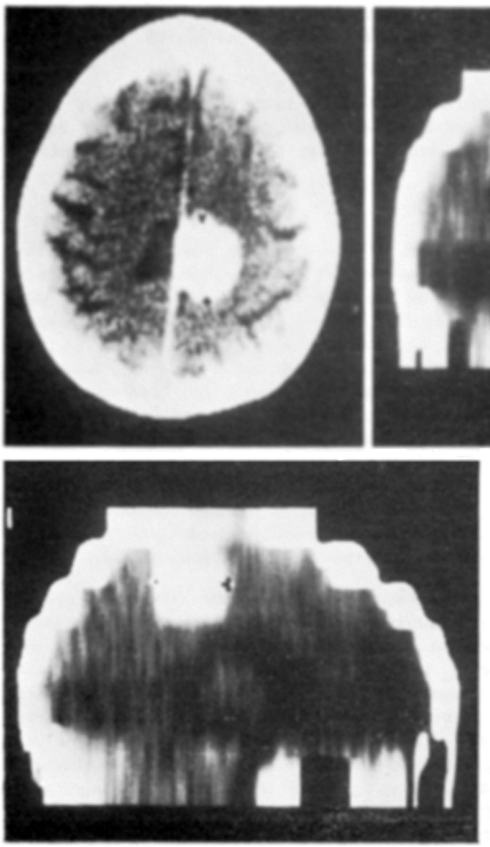

Fig. 1 — Case JF. Axial computed tomographic scan with coronal and sagittal reconstructions, showing round enhancing mass in close contact with the falx in the left parieto-occipital region of the cerebral hemisphere. The radiologic aspect is compatible with falx meningioma.

COMMENTS

There can be little doubt concerning the syndromic diagnosis of this patient [1,21]. Cases as these should not be just labeled as 'demented' or 'hysteric'. In fact, BS stands as the most representative paradygm of the possibility of the occurrence of perception without percepts in mentally sane people. The classic cases of BS are given by the emergence of hallucinosis in old people with falling vision due to a variety

of eye diseases [26]. Most writers believe that the visual defect per se suffices to explain the hallucinotic events arguing for a mechanism akin to the one known to be at play in conditions of sensory deprivation. This interpretation is certainly not exact, as it does not explain why BS does not occur in the large majority of blind people. On the other hand, there are positive data pointing to the fact that pathologic processes confined to the optic pathways are not likely, by themselves, to be accompanied by BS [10]. Thus, it seems that a second factor, besides the reduced quantity of afferent impulses travelling along the visual pathways itself must be active in BS. We suspect that this factor may be an affection of the brain. In the majority of the cases in whom the intracranial structures were investigated be it by radiological or per-operative means evidences of either gross or more subtle cerebral disorders were found, such as tumors, angiomas, atrophy [6,8,9,12-14,17,18,24-26]. There is then a suggestion that the finding of an expanding intracranial mass in BS patients, as was the case with ours, may represent the extreme point of a spectrum of pathologic alternatives which, in many other cases, may be not so evident or discarded as just meaningless. On the other hand, the close phenomenologic similarity linking BS to Lhermitte's peduncular hallucinosis should be noted [16],the main distinction between them being, indeed, the presence or absence of the campimetric defects. The pathologic correlates of peduncular hallucinosis are still obscure, but the responsible lesions are usually found in the high brainstem, thalamus or along the basal forebrain [2,5,7,9,20,22]. In addition and again indistinguishable phenomenologically from BS and Lhermitte's hallucinosis, are the ones occuring in instances of drug intoxication, withdrawal from alcohol and other neurodepressants, focal epilepsy and direct brain stimulation of selected regions, in particular Brodman's areas 18 and 19 [3,4,10,11]. That the production of hallucinosis depends on a number of mechanisms is already indicated by these brief citations. Nevertheless, this should not preclude the continued search for a distributed though specifiable neural circuit which, if destroyedor stimulated by either electrical or chemical means, correlates with the appearence of hallucinosis.

Until more systematic data are collected, we speculate that a subgroup of BS cases is caused by cortical injuries lodged in the parieto-occipital areas. Lance [14] presented a beautiful series of such cases although he curiously did not seem to have recognized their patients as fulfilling criteria for BS. In all of them the same lesion produced the campimetric defect and acted as an irritative stimulus for the elicitation of seizures which might have clinical and electroencephalographic concomitants. The patients of Brust and Behrens [6] and Sowa et al. [23] shared identical characteristics. Antiepileptic drugs probably exert a beneficial effect on such individuals who obbey to classic Jacksonian patterns of fits and, more recently, were grouped by Cogan as belonging to an 'irritative' type [8]. On the other hand, there is a second subgroup of BS patients that comprises the majority of cases and shows a campimetric defect, in general caused by eye disease, and a kind of hallucinosis identical to the one observed to occur in Lhermitte's peduncular hallucinosis. This appears to be what Bonnet himself described. We hipothesize that in many such cases the same general circuit which takes part in the production of any type of hallucinosis be it 'chemical' 'electrical' or anatomic, is called into play by an associated cerebral affection. If this really proves to be the case, the campimetric defect might have only a permissive value for the emergence of the apparitions. However, this statement should be taken as a provisional hypothesis and we are presently selecting patients to probe it.

A case identical to ours in many essential aspects was reported in 1965 by Mooney et al. [18]: a 45-year-old commecial artist developed BS due to a parasagittal meningioma. Perimetry disclosed a complete right lower quadrantanopsia with macular sparing and incomplete involvement of the right upper homonymous quadrants with wide macular sparing, suggesting 'compression of some branches of the left calcarine artery, with resulting ischemia of the striate cortex'. The symptoms remitted after excision of the tumor and interestingly, the patient painted some beautiful pictures of what he had seen while partially blind. During surgical intervention, it could be seen that 'there was marked elevation and separation of the branches of the posterior cerebral artery, one of which had a kinked and stretched circumscribed course'. This distortion was caused by a meningioma which 'indented the medial aspect of the parieto- occipital hemisphere in the region of the precuneus lobule' and was 'imbedded in the parietal lobe'.

Although no anatomic confirmation was available in our case, we guess that the patient also had a posterior parasagittal meningioma [15] and that this was the immediate cause of his hallucinosis, perhaps by an ischemic mechanism operating on the paras-

triate cortex. Meningiomas are slowly growing tumors[1] and it is possible that when the patient suffered his final decline of vision four years before consultation, the tumor was already there. Again, abrupt visual decline may have acted as a conditioning factor for the visions to become manifest. So in this case, as may commonly occur, different agents might have been active in the production of blindness and hallucinosis, respectively. Had we attributed our patient's complaints to 'visual afferent deprivation' only, the diagnosis of intracranial tumor would have been certainly missed. Until more restrictive criteria are elaborated, it seems advisable to submit every patient presenting with BS to a protocol investigation for intracranial disease, even if there is an obvious clinical explanation for his campimetric defects.

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