Unusual presentation of more common disease/injury

Frontal cortex dysfunction due to extensive hyperostosis frontalis interna

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An 87-year-old patient was found to have an unusually protrusive hyperostosis frontalis interna, discovered on MRI examination during an assessment of cognitive decline. Neuropsychological evaluation suggested direct repercussions of the frontal lobe compression on executive functions, as well as psychiatric disorders and possibly memory loss.

BACKGROUND

Hyperostosis frontalis interna (HFI) is an unexplained irregular thickening of the inner table of the frontal bone.¹ It is a common finding on brain imageries of postmenopausal women.² The potential repercussions of HFI on cognitive functions have been debated, but only a few cases have been reported to date.^{3 4} We describe here a case of remarkably extensive HFI in a patient exhibiting executive function disorders and memory loss.

CASE PRESENTATION

An 87-year-old woman was referred to the geriatrics department because of inappropriate behaviour, sleep disorders with circadian inversion and memory loss. The symptoms had been observed over a period of 2 years, but the patient had no complaint on cognitive impairment. Although she was able to fulfil basic activities of daily living, she needed assistance for dealing with more complex tasks such as accountancy.

The patient had a history of psychiatric disorders, with a tendency towards paranoia, emotional indifference and aggressiveness. Her pathological personality had led to multiple conflicts within the family. However, a progressive worsening in behaviour had been noticed, associated with lost of interest and self-neglect. She had no history of seizures or endocrinopathy. She had been treated with oxazepam, escitalopram, valsartan and hydrochlorothiazide.

At the time of initial consultation in March 2009, she presented with emotional lability from euphoria to irritability, uninhibited talking and distractibility. An impairment of executive functions was noted, with apraxia and non-semantic phasic disorders. Both semantic and phonemic fluencies were severely altered. The total MMSE (mini mental state evaluation) score was of 17/30, albeit preserved capacities in mental recall. Global physical examination was normal apart from a discrete extrapyramidal syndrome.

Neuropsychological evaluation in August 2009 showed rapid and global deterioration of cognitive functions, with

Figure 1 Axial T2* (A) and sagittal T1 (B) MR images showing severe, bilateral thickening of the inner table of the frontal bone with cortex compression.

Table 1 Patient's performance at neuropsychological testing performed in August 2009, 5 months after the initial consultation. A global alteration of cognitive functions is revealed, affecting mental recall and executive functions. Apart from recognition, similarities, prehension behaviour and Luria motor series, all test are pathological. Phonological fluency is severely altered.

Assesment				Score
MMSE				15/30
Mental recall	RL/RI-16	Immediate recall		13/16
		1st free recall		4/16
		2nd free recall		4/16
		3rd free recall		5/16
		1st recall total		8/16
		2nd recall total		10/16
		3rd recall total		12/16
		Delayed free recall		5/16
		Total free recall		8/16
		Recognition		16/16
		DMS 48		
		Set 1		69%
		Set 2		67%
Executive functions	Digit span (WAIS-IV)	Standard score		6/19
		Forward		4
		Backward		3
	Trail making test	Part A		285 s (<10th percentile)
		Part B		Impossible to test
	FAB	Overall score		9/18
		Similarities		2/3
	Lexical fluency (Mental flexibility)	Lexical fluency (Mental flexibility)		0/3
		Prehension behaviour		3/3
		Motor series «Luria» test		3/3
		Conflicting instructions		0/3
	Go- no go	Go- no go		1/3
		Phonological		1
	Categorical	Categorical		6
		Mental calculation		3/8

FAB, Frontal assesment battery; MMSE, Mini mental test evaluation. RL/RI-16 is a test using the Gröber and Buschke procedure (Gröber E *et al Neurology* 1988;**38**:900–3), assessing the patient's capacities in free and cued recall of a subset of 16 words. DMS 48, visual Delayed Matching-to-Sample task (Barbeau E, *et al Neurology* 2004;**62**:1317–22). WAIS-IV, Weschler adult intelligence scale.

respect to orientation, storage and retrieval process in verbal episodic memory, visual recognition, work memory, executive functions (mental flexibility, inhibition, fluency), language and praxis. Table 1 presents the subset of tests regarding memory and executive functions, which were performed. The MMSE score was then 15/30 and the frontal assessment battery was 9/18 (table 1).

INVESTIGATIONS

The MRI examination performed in July 2009 revealed an extensive bilateral and symmetrical HFI, type D,¹ compression and pushing back of the majority of the frontal lobe (figure 1). A global but asymmetric cortical loss with evolved atrophy of the right hippocampus and moderate dilatation of the anterior temporal region of the right ventricle were also observed (figure 2).

DIFFERENTIAL DIAGNOSIS

An association with Alzheimer's disease appears likely given the results of MRI examination and neuropsychological evaluation.

Figure 2 Coronal T1-weighted MRI picturing atrophy of the right hippocampus.

OUTCOME AND FOLLOW-UP

The patient initially presented selective impairment of executive functions. However, a global and severe cognitive decline appeared rapidly, leading to a high level of dependency and entry in a specialised nursing home, within 6 months after the initial consultation.

DISCUSSION

Although HFI is a common, often incidental finding among older woman, such severe cases have rarely been observed.^{2–5} HFI is usually bilateral, although unilateral cases have been described,⁶ and spares the midline. It was originally described as part of Morgagni-Stewart-Morel syndrome,⁷ (associated with virilism and obesity). Many associations have been reported since,^{2–8–9} but authors now agree that HFI exists independently.

The cause of HFI remains uncertain. Hormonal influences on bone growth in the skull have been postulated. Sexual hormones for example, oestrogen stimulation,¹—could play a part in the emergence of HFI and explain its predominance among female. Other metabolic disorders or endocrinopathies have been highlighted, such as acromegaly (especially when associated with hyperprolactinemia),^{10–11} diabetes or leptin hyper secretion.^{8–12} Genetic predisposing factors are also involved.^{6–13}

The large majority of patients appear to be asymptomatic. In severe cases, HFI can lead to compression of soft tissue, dural irritation and atrophy due to increased pressure in the brain.⁶ It is virtually established that HFI can cause diverse psychiatric disturbances, such as aggressiveness, paranoia or depression.¹⁴ Our patient seems to match the description.

Potential consequences of HFI on cognitive functions are less well documented. To our knowledge, two cases have been published previously, both, as might be expected, showing alterations of the frontal lobe (executive function disorders, alterations of work memory and mental flexibility).^{3 4}

Our patient initially presented selective alterations of executive functions. However, the cognitive impairment rapidly evolved to become global, suggesting a possible entanglement with Alzheimer's disease (also indicated by atrophy of the right hippocampus on MRI examination, as shown on figure 2). The executive dysfunction could possibly be explained partly by Alzheimer's disease. However, the severity of the impairment strongly suggests that the frontal lobe compression due to HFI has played a part. For example, the deficit in phonological fluencies is striking, and seems much more impaired than could be expected in a context of Alzheimer type dementia alone.¹⁵ The symptoms of 'frontal type' behaviour have preceded the memory loss, and were clearly at the foreground at the time of initial consultation. Furthermore, we can propose the hypothesis that the frontal cortex compression due to HFI could have altered the patient's ability to compensate the memory loss with organisational strategies, explaining rapidity of the evolution of dementia.

Learning points

- ▶ Our case tends to confirm that frontal lobe compression due to extensive HFI can lead to psychiatric disorders and cognitive impairment, especially with regard to executive functions.
- ▶ In the event of coincidental degenerative dementia, the presence of HFI could be a bad prognosis with respect to rapidity of evolution of cognitive decline.

Competing interests None.

Patient consent Obtained.

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