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Progressive Dementia and Behavioral Changes: Pick's Disease: A rare disease or an underdiagnosed disorder?

[Presentación](#)

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Pick's disease is a nosological entity with frontal dementia, early cortical dementia with severe frontal lobe disturbances, absence of apraxia, and absence of gait disturbance at onset. This disorder is underdiagnosed in clinical practice. When specific criteria for the clinic, imaging and neuropathologic diagnosis including the presence of Pick bodies, are used, the diagnosis of Pick's disease is achieved. However, taking into account that the definitive diagnosis of PD is achieved only with pathological study, in vivo diagnosis requires of the combination of neuroimaging techniques.

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More than 100 years ago, Arnold Pick described several patients who presented with progressive behavioral changes (apragmatism, outbursts of rage, and later stages, mutism) and who, at autopsy, had characteristic frontal or temporal lobar atrophy [1]. Alois Alzheimer histologically characterized the disorder when he described "argentophilic globes" in the cytoplasm of neurons and the presence of ballooned neurons and spongy cortical wasting in the absence of neurofibrillary tangles or plaques [2]. Pick's disease is considered a relatively rare neurodegenerative disorder, affecting subjects in their 60s with the progressive development of frontal lobe type features (e.g., difficulty planning, reasoning, abnormal social behavior), language disturbances (decreased fluency followed by echolalia, mutism), later followed by memory and gait abnormalities and occasional parkinsonism [3]. In autopsy studies of progressive dementia, only about 5% are due to PD. The underlying cause is not known, but there does appear to be a hereditary component, with clear autosomal dominant transmission in some families. The disease usually progresses inexorably over 2 to 5 years to death. At the present time there is no specific treatment available [4]. We report our findings in a patient with the purpose of to attract attention about this entity with the aim to avoid unnecessary, expensive and dangerous treatment when this kind of patients received a wrong diagnosis.

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Case presentation

An 81-year-old, right-handed woman, retired dressmaker, was referred to the Neurology Department in December 1999 because of progressive dementia and behavioral changes. Poor memory and speech difficulty had been increasing for about 5 years, above all in the last two years. She was unable to sew on a button or a zip, despite she knew very well her job. She had a compulsive repetition of a word or syllable, echolalia, outbursts of rage and hyperactivity. Eight months ago she had been visited in another hospital where a Neuropsychological assessment showed an intelligence score according to her age, but with features of a frontal syndrome. In this sense, disinhibition, a decrease in the verbal fluency and presence of fabrications highlighted. Computed tomography (CT) of the brain revealed a parasagittal left frontal meningioma. There were no reported ischemic lesions. A SPECT cerebral perfusion (Figure 1) disclosed markedly reduced perfusion in both frontal lobes, prevailing in right side, by what her symptoms were attributed to the meningioma. A thyroid ultrasonography revealed a pseudocyst in the right side and her test of thyroid function disclosed a hypothyroidism. In this hospital, on examination, there were no cardiovascular abnormalities. Blood

pressure was 140/80. On testing higher cortical function, her speech was somewhat rambling and hesitant with word finding difficulty; she was compulsive repetition of word or syllable. She scored 4 in the Global Deterioration Scale (GDS) [5] and 23/30 in the Mini-mental State Examination (MMSE). Cranial nerve examination was normal. There were primitive reflexes (hand grasp and glabellar reflexes), and the rest of the neurological examination was normal. She was referred to the Endocrinology Department to treat her thyroid problem. Neuropsychological assessment showed visuospatial and visuoconstructive difficulties, and she had commensurate difficulties with calculation, that no changed after two months of substitutive treatment with thyroid hormones. At follow up the patient was noted more retracted and with periods of mutism. Initial investigations were undertaken. Blood studies disclosed a thyrotropin concentration increased to 10,35 mUI/l (0,25 – 5,00). Brain magnetic resonance imaging (MRI) disclosed a severe frontal, temporal and parietal lobar atrophy; a parasagittal left frontal meningioma of about 1,5 to 2,5 cm, with no mass effect (Figure 2). There were no reported ischemic lesions. Proton nuclear magnetic resonance spectroscopy (MRS) disclosed decreased levels of N-acetyl-L-aspartate, mio-inositol and creatine. We thought that the patient had a Pick's disease.

COMMENT

Clinical features suggestive of Pick's disease include personality change, social and interpersonal disinhibition, apathy, and language disorders. Disproportionate frontotemporal atrophy on structural imaging and prominent frontal hypometabolism on functional imaging usually supports the diagnosis [6-8].

In contrast to Alzheimer's disease, where the atrophy is relatively mild and diffuse, the pathologic change in Pick's disease is a severe wasting of the frontal and temporal lobes. The parietal lobes are involved less frequently [6-8].

This patient, an 81-year-old, right-handed woman, became 5 year ago with dementia and behavioral changes, increased above all in the last two years. She scored 4 in GDS and 23/30 in MMSE (the latter scale lower than the normal, even in person with a low level of schooling), hyperactivity, compulsive repetition of the word, high verbal fluence, and verbal perseverance. This picture could be related to her age associated to her hypothyroidism or to the meningioma effect, or both. However, seeing as the substitutive treatment with thyroid hormones did not modified her neuropsychological performance, hypothyroidism was ruled out as the cause of her cognitive and behavioral alterations. On the other hand, the meningioma, which was localized on the representative area of the leg's motility, did not caused her clinical picture, since if it would do a compressive effect, would give a right's leg paresia, but this do not happen, even less to be the cause of a as diffuse problem as this.

Neuroimaging studies disclosed a big fronto-temporo-parietal atrophy, more evident than seen in Alzheimer's disease patients (MRI), markedly reduced perfusion (hypometabolism) in both frontal lobes (SPECT), and decreased levels of N-acetyl-L-aspartate (lower than seen in patients with Alzheimer's disease and vascular dementia) and decreased of mio-inositol and creatine (MRS), suggests of a severe neuronal loss (9). In our serie, patients with Alzheimer's disease had a decreased of N-acetyl-L-aspartate and creatine, but an increase of mio-inositol, and the patients with vascular dementia had a decreased of of N-acetyl-L-aspartate and an increase of creatine. By all this information, we thought that the patient had a Pick's disease (10, 11), since she carried out 5 of 6 criteria of Pick's disease (Table 1).

Definitive diagnosis of Pick's disease is achieved with pathological study: biopsy or necropsy. Biopsy did not was performed due to hemorrhages related to biopsy in elderly patients with dementia are common (12,13). On the other hand, surgical treatment for asymptomatic meningioma in elderly patients is not recommended, therefore we did not see the advisability of surgery and she was not operated (14). Patient was undergone to paroxetina, and a follow up every 6-month and after every year in order to make a careful clinical and neuroimaging observation.

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TABLE 1.

Criteria of Pick's Disease

1. Clinical: some o several of following:
 - § Personality changes: hyperactivity, outbursts of rage, abnormal social behavior.
 - § Language disorders: dysphasia with palilalia, compulsive repetition of a word, echolalia.
 - § Memory disorders: later, poor concentration, progressive oversights.
2. Fronto-temporal atrophy disclosed by structural neuroimaging: MRI, TC.
3. Exclusion of frontal infarcts or multi-infarction disclosed by MRI, TC.
4. Massive neuronal loss disclosed by decreased levels of N-acetyl-L-aspartato, disclosed by SMR.
5. Frontal bilateral hypometabolism disclosed by functional neuroimaging: PET, SPECT.
6. Pick's body: argentophylic neuronal inclusions, disclosed by biopsy or necropsy.

LEYENDAS DE FIGURAS

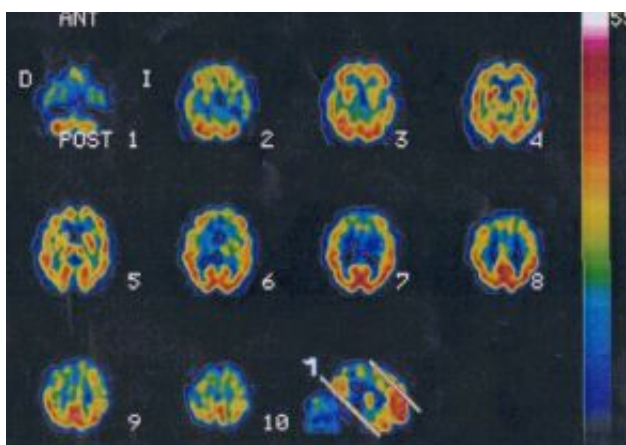


Fig 1. SPECT. Representative spectra depicting the hypometabolism of both frontal lobe, prevailing in right side

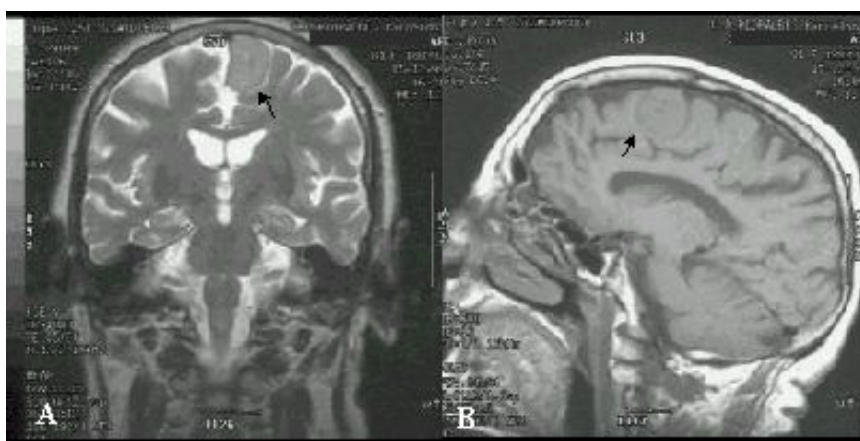


Fig 1. MRI. T2 y PD, depicts meningioma location midfrontal (A) and parasagittal (Binswanger) in the left frontal lobe (arrows).

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