PSICOSOMÁTICA
Y PSIQUIATRÍA

17 / AÑO 2021
ABRIL • MAYO • JUNIO

ARTÍCULO ORIGINAL

ORIGINAL ARTICLE

Recibido: 22/02/2021. Aceptado: 27/05/2021

DIOGENES SYNDROME: A LATE-ONSET CASE IN FRONTOTEMPORAL DEMENTIA

SÍNDROME DE DIÓGENES: UN CASO DE INICIO TARDÍO EN LA DEMENCIA FRONTOTEMPORAL

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Potential conflicts of interest: The authors declare no competing interests and no conflicts of interest regarding the publication of this case report.

Funding/support: None.

Disclaimer: Consent was received from the patient to publish the case report, and information has been de-identified to protect anonymity.

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ABSTRACT

Diogenes Syndrome (DS) is a behavioural disorder characterized by severe self-neglect. In addition, DS is characterized by domestic squalor, social withdrawal, syllogomania and refusal of help that may be precipitated by stressful events. Primary DS reports are rarely described. Secondary DS is related to mental disorders. The time span in which the syndrome develops is undefined, though it is most accurately characterized as a reaction to stress that occurs late in life. This specific behavioural disorder referred to as DS is a significant functional problem contributing to increased morbidity and mortality and may be due to one or several underlying mechanisms, including personality disorder and frontotemporal dementia. We describe a case of DS in a 90-year-old man which presented with a 5- year history of personality changes, progressive behavioural changes and decline in memory. Neuropsychological testing found executive deficits, decreased verbal fluency and disturbed memory. CT-Scan imaging revealed cerebral atrophy possibly more prominent in the frontotemporal regions. His hoarding, compulsive and collecting behaviours decreased with sertraline, donepezil and risperidone. DS is a severe psychiatric condition with a multifactorial aetiology. It is strongly related to functional and cognitive decline, specially in elderly patients with comorbid frontotemporal dementia. DS represents a clinical, social and ethical challenge that requires a multidisciplinary team approach. Further investigation into the hoarding behaviour´s aetiology may lead to the development of treatment options for patients with frontotemporal dementia.

Keywords: Diogenes Syndrome, Hoarding, Frontotemporal Dementia, Self-neglect.

RESUMEN

El síndrome de Diógenes (SD) es un trastorno conductual caracterizado por una auto negligencia extrema. Además, el SD se caracteriza por insalubridad del domicilio, falta de higiene, aislamiento social, silogomanía y el rechazo a la ayuda, que pueden estar precipitados por eventos estresantes. Los casos de SD primario raramente se describen. El SD secundario está relacionado con trastornos mentales. El período de tiempo en el que se desarrolla el síndrome no está bien definido, aunque de forma más precisa se puede caracterizar como una reacción a estresores que se produce en una etapa avanzada de la vida. Este trastorno conductual específico, denominado SD constituye un problema funcional importante que contribuye a una mayor morbilidad y mortalidad, pudiendo deberse a uno o varios mecanismos subyacentes, incluidos el trastorno de personalidad y la demencia frontotemporal. Describimos un caso de SD en un varón de 90 años con un cuadro de 5 años de evolución de cambios de personalidad, progresivas alteraciones de comportamiento y deterioro de la memoria. Las pruebas neuropsicológicas hallaron déficits ejecutivos, disminución de la fluidez verbal y alteración de la memoria. La prueba de neuroimagen (Tomografía axial computarizada) reveló atrofia cerebral posiblemente más prominente en las regiones frontotemporales. Su tendencia a la acumulación, compulsión y coleccionismo disminuyó con sertralina, donepezilo y risperidona. El SD es una enfermedad psiquiátrica grave de etiología multifactorial. Está fuertemente relacionado con el deterioro funcional y cognitivo, especialmente en pacientes ancianos con demencia frontotemporal comórbida. El SD representa un desafío clínico, social y ético que requiere un enfoque multidisciplinar. Investigación adicional sobre la etiología del comportamiento de acumulación podría conducir al desarrollo de opciones de tratamiento para pacientes con demencia frontotemporal.

Palabras clave: Síndrome de Diógenes, acumulación, demencia frontotemporal, auto-negligencia.

INTRODUCTION

Diogenes Syndrome (DS) is a behavioural disorder characterized by severe self-neglect, domestic squalor, social isolation and excessively hoarding (syllogomania) (Biswas et al., 2013; Cipriani, Lucetti, Vedovello, & Nuti, 2012; Finney & Mendez, 2017). Primary DS reports are rarely described (Finney & Mendez, 2017). Secondary DS is related to other mental disorders including dementia, probably due to one or several underlying mechanisms (Greve, Curtis, Bianchini, & Collins, 2004) and it is a significant functional problem contributing to increased morbidity and mortality. New-onset DS in older age may be secondary to dementia (Cipriani et al., 2012). However, subtle frontal lobe dysfunction is reported in some cases of DS not fulfilling diagnostic criteria of dementia (Chan, Leung, & Chiu, 2007). DS may have a special relationship with behavioural variant Frontotemporal Dementia (bvFTD) (Cipriani et al., 2012), which is the most frequent frontotemporal Lobar Degeneration phenotype (Galimberti & Scarpini, 2013). It commonly presents with behavioural changes and deterioration of personality. (Galimberti & Scarpini, 2013; Mendez, Perryman, Miller, Swartz, & Cummings, 1997).

CASE DESCRIPTION

Initial presentation: A 90-year-old man presented with progressive personality, behavioural changes, memory decline and impairment of social insight. Over the past 5 years, he had become disinhibited, acting inappropriately, demonstrating excessive familiarity with strangers. He exhibited a decline in self-care accompanied by hoarding behaviours, collecting items such as dozens of futile objects and excessive hoarding of garbage. His relatives reported memory loss. On examination, the patient was alert and cooperative. He showed no insight into his disease and denied any changes in his behaviour or personality.

Cognitive evaluation, laboratorial and neuroimaging finding: Neurologic examination was remarkable for a frontal grasp reflex and restlessness. The Mini-Mental State Examination was 19/30. Complete blood count, electrolytes, glucose, liver function, and lipid profile were all within normal limits. Brain computed tomography scan showed cerebral cortical atrophy more prominent in the frontotemporal regions (Figure 1). Neuropsychological evaluation found deficits in executive tasks, decreased verbal fluency and relative sparing of episodic memory, concluding a moderate cognitive dysfunction impairment, amnestic type.

Diagnosis and treatment: Hoarding and self-neglect which can occur in DS, can also be found in dementia and secondary DS in a patient with bvFTD was a suspected diagnosis. His hoarding, compulsive and collecting behaviour decreased gradually with sertraline (100 mg once a day) and

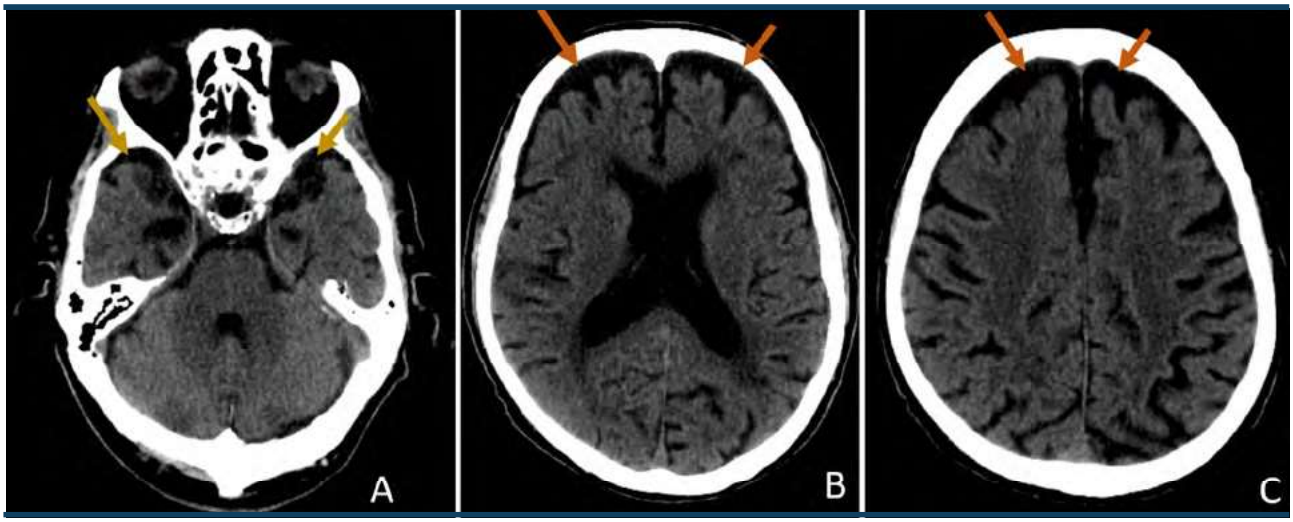

Figure 1. *CT Scan revealed symmetrical enlargement of the cerebrospinal fluid spaces in temporo-polar topography (yellow arrows) and anterior frontal (orange arrows), in a context of diffuse cortico-subcortical atrophy, of bilateral frontal-temporal cortical predominance.*

risperidone (0,5 mg twice a day).

Clinical evolution: He was admitted in nursing-home with some improvement in physical state and environmental hygiene. However, he shows a lack of concern or awareness of their condition. Further history indicated a significant improvement in selfcare and hygiene involving dressing and grooming.

DISCUSSION

This case illustrates many features of DS reported in the literature and the importance of suspecting DS when elderly patients present with non-specific symptoms that may otherwise be disregarded. DS is a severe psychiatric condition with a multifactorial etiology and it is strongly related to functional and cognitive decline (Galimberti & Scarpini, 2013), especially in elderly patients with with moderate or severe dementia. (Hwang, Tsai, Yang, Liu, & Lirng, 1998)

DS can be distinguished from psychiatric disorders (Fontanelle, 2008) or can also be a part of dementia, schizophrenia, obsessive-compulsive disorder and affective disorders. (Amanullah, Oomman & Datta, 2009) The frontal lobe dementia tends to occur approximately 10 years prior to the typical age that DS patients are affected. (O'abrien, 2017) Clearer delineations between disorders need to exist however. An alterative suggestion was that DS "may be a final common pathway of different psychiatric disorders. (Fontanelle, 2008) In the presented case, the patient started showing behavioural changes secondary to bvFTD before hoarding behaviours. Early detection would help patients through introduction of psychosocial interventions, before the clinical symptoms are maintained and difficult to treat.

Previous studies have implicated abnormality in the frontal lobe in association with decrease self-awareness and personal hygiene (Finney & Mendez, 2017; Guimarães, Fonseca, & Garrett, 2006), compulsions and the release of a need to accumulate objects (Finney & Mendez, 2017), like in this case report. The spectrum of DS in a bvFTD includes a decline in self-awareness and self-care and collecting behaviour to a tendency for clutter and a disordered environment (Mendez et al., 1997). The literature suggests a combination of frontal lobe disturbances, namely lack of insight or self-awareness (Mendez et al., 1997), which results in a clutter and disorganization (Finney & Mendez, 2017). Compulsive behaviour is characteristic of bvFTD (Finney & Mendez, 2017) and there is an orientation to environmental stimuli which may facilitate collecting behaviour (Cooney & Hamid, 1995). There are several common aspects between DS and FTD (e.g., carelessness, indifference, physical neglect, aggressiveness, stereotyped and persistent behaviours). This suggests that altered function of frontal lobe may be one of the determining factor in DS, but there are also remarkable differences between these two pathologies like less cognitive impairment in DS patients, precocious and progressive language disturbance in FTD (Finney & Mendez, 2017). DS in bvFTD represents a clinical, social and ethical challenge that requires a multidisciplinary team approach.

There is no specific therapy approved for FTD (Guimarães et al., 2006), nevertheless selective serotonin reuptake inhibitors (SSRIs) seemed to have some improvement in reducing hoarding (Finney & Mendez, 2017; Guimarães et al., 2006). Our patient improved with SSRI treatment (sertraline). Other pharmaceutical that may be of benefit include zolpidem for insomnia, paroxetine for hoarding, and sodium valproate or quetiapine for secondary bipolar disorders (Amanullah, Oomman & Datta, 2009).

Timely diagnosis may reduce both acute and chronic physical illness, increase personal and home hygiene and safety, and improve public health outcomes. Management of DS can be difficult, as patients often deny that there is a problem, may refuse any help, and can present late to medical attention (Irvine & Nwachukwu, 2014). The prognosis depends on capability of re-integrating into society, and often relies on making small changes away from unhealthy living condition. (Irvine & Nwachukwu, 2014) Some prognostic factors include early age at onset and poor physical health, which may already be advanced due to self-neglect. (Irvine & Nwachukwu, 2014) Further investigation into the hoarding behaviour´s aetiology may lead to the development of treatment options for patients with dementia.

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1. **Potential conflicts of interest:** The authors declare no competing interests and no conflicts of interest regarding the publication of this case report.
2. **Funding/support:** None.
3. **Disclaimer:** Consent was received from the patient to publish the case report, and information has been de-identified to protect anonymity.