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CASE REPORT/**CASO CLÍNICO**

Ekbom Syndrome: A Case Report
Síndrome de Ekbom: Relato de Caso

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Abstract
Delusional parasitosis, also known as delusional infestation or Ekbom syndrome, is a somatic type of delusional disorder, usually mono-symptomatic, in which patients are convinced they are being infested with animal parasites while no objective evidence to support their belief exists. Complaints are usually about skin infestation, but involvement of the gastrointestinal tract has also been described. We describe a case of a 59-year-old woman with delusional parasitosis claiming to be infected with *Strongyloides stercoralis.*

Resumo
A parasitose delirante, também conhecida como delírio de infestação ou síndrome de Ekbom, é um tipo de delírio somático, normalmente monossintomático, no qual os doentes estão convictos de estarem infestados por parasitas, não existindo, no entanto, qualquer evidência que suporte esta crença. Normalmente, a "infestação" refere-se à pele, mas "infestações" do trato gastrointestinal encontram-se também descritas na literatura. Relatamos um caso de uma senhora de 59 anos de idade com um delírio de infestação por *Strongyloides stercoralis*.

Keywords: Delusional Parasitosis/diagnóstico; Delusional Parasitosis/tratamento

Palavras-chave: Delírio de Parasitose/diagnóstico; Delírio de Parasitose/tratamento

INTRODUCTION

Delusional infestation was first described in 1937 as "pre-senile delusion of infestation" by Dr. Karl-Axel Ekbom, a Swedish neurologist.[1] Delusional parasitosis (DP) is a somatic type of delusional disorder, usually mono-symptomatic, in which the patient is convinced of being infested with animal parasites (usually insects or worms), and rarely viruses and bacteria, while no objective evidence to support their belief exists.[2,3] The disorder is most frequently seen in middle-aged, often socially isolated, women (average age is 57 ± 14 years).[3-6]. In 1988, it was reported that DP was diagnosed in about seven out of 10 000 psychiatric admissions. However, the incidence of this disorder is probably much higher because most cases of DP are treated by dermatologists and the prevalence of the disease reported by psychiatrists only reflects a minority of this population.[2] Patients may describe tactile, visual or auditory hallucinations related to the infestation. The "*matchbox sign*" is the classic presentation, i.e. a box or container where various materials that have been collected by the patient as evidence of infestation, including anything from dust/dirt, plant or animal fibers, scabs and skin debris, to photographs of old/previous lesions or parasites are gathered.[7,8] The most frequent imaginary sources of infestation are other people (50%), particularly those returning from a trip to developing countries but also pets and real infestations with parasites which were controlled successfully in the past.[2] One possible reason for the delusional symptoms is the false interpretation of an itching sensation, which is a common symptom. Depending on the

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location of the infection, excoriation erosions and cuts are frequent forms of self-harm caused by attempts to eliminate the parasites. Skin damage can be caused by obsessive cleansing and the application of aggressive chemical or caustic substances to remove the fictitious parasite.[4,6]

Folie à deux, also known as shared psychotic disorder[9] or induced delusional disorder,[10] a condition in which psychiatric symptoms (delusions or hallucinations) are transferred from one individual to another, is not uncommon in this disease.[8]

Past or comorbid psychiatric conditions are reported in roughly 80% of patients with DP. The most common of which is depression (74%), followed by substance abuse (24%) and anxiety (20%).[8]

Delusional parasitosis can be categorized into primary, secondary and organic forms. In primary delusional parasitosis, the patient has the delusion of being infested with parasites, but no other psychiatric or organic disorder is present. The secondary and organic forms occur secondarily to other disorders, namely psychiatric and organic disease, respectively. There are several mental disorders that can be associated with Ekbom syndrome, such as schizophrenia, schizophrenia spectrum disorders, depression, dementia, affective psychosis, substance–induced psychosis and psychosis caused by a general medical condition (eg., iron deficiency, anaemia). Anxiety disorders and somatoform disorders should be carefully distinguished from delusional states and may be comorbid with delusional disorder. DP must also be differentiated from obsessive--compulsive disorder or, according to DSM-5, "skin picking disorder", which can result in ritualized skin cleaning and consecutive skin lesions but without the unshakeable belief of it being infested by living creatures.[2]

The organic type of DP is linked with thyroid disease, anaemia, vitamin B12 deficiency, hepatitis, diabetes, infections (e.g., HIV, syphilis), cocaine abuse[1,4,6] and neurological diseases such as multiple sclerosis.[11]

A thorough evaluation is required to establish the diagnosis, including a complete history and physical examination. It is important to rule out genuine parasitosis. Formication and pruritus caused by medications such as amphetamines, cocaine, opioids, dopamine agonists, topiramate or alpha--adrenergic agents must also be considered. A review of current medications is essential.[1] Pruritus related to systemic diseases including hyperthyroidism, renal or liver disease may present as a generalized itch or a crawling sensation in the skin. Exposure to fiberglass may produce itching of unknown etiology and fiberglass-contaminated clothing placed in the laundry can result in itching in an entire family.[8]

There is a case report describing a delusional parasitosis as presenting symptom of occipital lobe cerebrovascular accident.[12]

A complete blood cell count, metabolic panel, thyroid--stimulating hormone and obtaining a biopsy specimen of lesional and perilesional skin for direct immunofluorescence may help to rule out organic disease and help establish an effective physician-patient relationship.[8] If substance induced psychosis is suspected, a urine drug screen is highly valuable as well as a search of controlled substances that are being prescribed via a database network search.

Brain computerized tomography (CT) is also important – the emergency physician should consider a psychiatric cause of delusions as a diagnosis of exclusion, made only after medical causes have been ruled out.[12]

DP patients often refuse psychiatric referral, as they do not believe that they have a psychiatric problem. In a study conducted in Argentina, psychiatric referral was possible for only 1 in 12 patients (8%). When offered, patients became upset and angry, accusing the physicians of incompetence. Although aggressive reactions are rare, an attempt to murder a family doctor has been reported.[2]

Antipsychotic drugs are the most effective agents to treat DP. In the past, pimozide was used as the first-line agent, but it has a less favourable side effect profile compared with second generation (atypical) antipsychotic agents, and electrocardiograms are recommended to measure Q-T interval before treatment.[8] While pimozide is still used successfully to treat DP, newer atypical antipsychotic agents such as risperidone, olanzapine, quetiapine and paliperidone are good options because of their favourable side effect profiles.[3,8] Risperidone is the treatment of choice for delusional infestation, with a recommended dose range of 0.5-2 mg daily. Low dose olanzapine (2.5-12 mg daily) has also been shown to be effective in DP. Third-generation antipsychotics, such as aripiprazole, are likely effective but have not yet been studied in delusional infestation.[1]

Due to the paucity of well controlled medication treatment studies, response rates for antipsychotic treatment has not been well-established and reported response rates vary.[2]

Longer duration of untreated psychosis in patients with delusional infestation, like in other psychiatric diseases, is associated with significantly less favourable clinical outcomes.[13]

CASE REPORT

a. History of Present Illness

A Woman of 59-year-old, divorced, with the 6th grade of education. She worked as a nursing assistant in a nursing home.

Presented to the Emergency Department of a central general hospital in August of 2018, where she was first observed by Internal Medicine. Her complaints were swelling and pain in the upper limbs. Upon physical examination, no evidence of oedema or skin alterations was found. The patient claimed the symptoms were caused by a parasite and that she had been infected for a long period of time. A complete blood panel and urine drug test was unremarkable. She had a CT head scan the previous year, after complaining to her general practitioner of persistent head and earaches, showing no abnormalities. She had also a normal electrocardiogram performed two months prior.

The patient was then referred to the psychiatric team for assessment. She accepted the referral, albeit with some reticence. She reported to the psychiatrist that she had been

infected by the parasite *Strongyloides Stercolaris*, describing its life cycle in great detail. It had supposedly happened 15 years before, when she had been living in France with several other people, amongst them a woman of African descent. She claimed this woman had "*thrown the larva*" at her, which then it lodged under her skin. After this, she said she had never been able to get rid of it. She had insisted multiple times with her general practitioner (GP) to be referred to a tropical medicine specialist, because she considered that general practitioners and internal medicine specialists were not able to handle such infections.

She claimed to have been able to catch the larva once, which made a "crutch"-like sound. The patient said she could feel the larvae under her skin, and that "*they are more active at the end of the day*".

The patient was convinced the larvae were inside her and that she sometimes saw them on her skin but was unable to catch them. She explained that, in the past, larvae had been expelled through the ocular mucous membrane. In an attempt to rid herself of the infestation, she had bought albendazole, without a medical prescription, and taken it for 4 straight months, once daily, claiming that her gastric upset had improved, but that "*the larva was still in the intestine*".

She denied taking any other substances or using any other methods, such as sharp objects, to get rid of the parasite.

The patient denied restricting social contact and claimed to remain functional at work. She did not believe she could pass the parasite onto others.

b. Previous Psychiatric History

Reviewing the patient's medical records, it was found that she had previous psychiatry follow-up, despite denying so. Her psychiatrist described an infestation delusion and possible hallucinatory activity in the form of formication, as well as delusional interpretations. As far as it was possible to establish, the symptoms had been present since 2004.

In March of 2009, her mental condition worsened, showing severe anxious and depressive symptoms congruent with the delusional system. She was then voluntarily admitted to a psychiatric hospital. Nevertheless, she denied having any kind of psychiatric disorder, claiming that the admission was only meant to draw blood samples.

The patient underwent viral tests and an urine test for illicit drugs as well as a formal cognitive assessment, which revealed no changes.

As an inpatient, she was started on risperidone up to 6 mg/daily, with reasonable tolerability but with poor response. She was then started on olanzapine up to 10 mg/daily, with better response and good tolerability. Upon discharge, follow-up outpatient appointments were scheduled. She stopped complying with the follow-up and psychopharmacological treatment after the first appointments. She has not accepted referral to a Psychiatric consult by her GP since.

c. Mental Status Examination

Mental status examination showed adequate self-care, the patient was awake and oriented in all domains, with no apparent amnesic deficits. Her posture was hostile, and she exhibited an irritated mood. Speech was organized, her tone of voice in crescendo, quickened, and centred in the complaints related to the "infection", which she described in great detail. She constantly called the Psychiatrist's competence into question. Psychotic symptoms were present, namely delusional ideas of endo- and ectoparasitic infestation and delusional interpretations of ordinary bodily sensations and excretions, as well as haptic hallucinations (in the form of formication). Visual and auditory illusions were most likely present in the past, but not upon the current evaluation. No intention of self-harm or harm to others was perceived. Sleep and appetite were maintained. She displayed no insight towards her condition or to the necessity for treatment.

d. Follow-up

Upon our observation, we spoke with the patient in an attempt to validate the complaints of discomfort but alerting to the harmful use of daily albendazole. The patient's willingness to initiate psychopharmacological treatment was also questioned, without imposing on her wishes. The patient was discharged with indication to suspend treatment with albendazole, and she refused referral for psychiatric consult.

From what we were able to gather, the patient did not comply with the proposed treatment. She continued having quarrelling interactions with her GP and with doctors in other primary care centres, demanding further diagnostic exams and antibiotic treatment. She has not since been referred to Psychiatric evaluation.

DISCUSSION

The present case illustrates a patient with Ekbom syndrome, who closely resembles the typical characteristics described in the literature, namely female gender, between the fifth and sixth decades of age, single, divorced or widowed and a low level of education.

The patient presents with DP, arising spontaneously as a mono-delusional disorder, meeting the criteria of the International Classification of Diseases, 10th revision (ICD--10)[10] for persistent delusional disorder and the criteria of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5)[14] for delusional disorder, somatic type.

Reassurance regarding the lack of evidence of organic disease rarely provides relief, and patients often see multiple physicians in search of someone who will believe them.[8]

Symptoms of DP are very real and distressing to the patient, who often seek medical help and advice from GPs, dermatologists and parasitologists.[2]

In the diagnosis and follow-up of these patients, it is important to have effective communication between different specialists, such as general practitioners, dermatologists and psychiatrists.

The most challenging and intricate problem for practitioners is to sufficiently build patients' trust and alliance, to be able to engage them in psychopharmacological treatment.[2]

The emergence of such cases in a psychiatric emergency setting, and regular clinical discussion meetings between psychiatry specialists and primary health care doctors, can

alert both GPs and psychiatrists to this disease, which has a large impact on patient's quality of life. Many are never observed by a psychiatrist, which makes awareness among colleagues of other specialties even more relevant.

Treatment is only possible within a solid doctor-patient relationship, based on trust. This should be worked on prior to offering psychopharmacological treatment. The primary challenge for psychiatrists and GPs is to not "lose" the patient to follow-up. Poor adherence is the main factor leading to ineffective treatment.[7]

Considering the long duration of untreated psychosis, the history of reiterated non-compliance with treatment and follow-up and the complete absence of insight, the reported case seems to have a poor prognosis.

In conclusion, more information would be ideal, possibly with a close relative, to clearly define the intricacies of this patient's psychiatric history, social stressors, and possible iatrogenic contributions to the psychosis she experienced. This was not manageable in the Emergency Department setting.

Most of the literature on this subject is limited to case reports and open studies. Further research including significant controlled and randomized studies should focus on thoroughly evaluating the current recommendations for the treatment of delusional parasitosis, due to its major impact on the patient's quality of life.

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