Royal Eschizophilia. Genealogical delusion or Mignon delusion as a pathoplastic realization

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Dear Editor,

Genealogical delusion has been considered a variety of megalomania in which the patient feels related or descendant of Kings, Princes or any special lineage. Hitler is considered as a historical case of Aryan genealogical delusion. It has been described in patients that have felt to be members or successors of the Royal family of France and Savoy[1] and it can appear both in the manic psychosis and schizophrenia.[2] In the glossary of the Present State Examination (PSE) within delusions of great identity we can find the delusion of being God and also the Mignon delusion, in which the patient believes that he comes real kinship or a most distinguished family.[3] M. Sanati describes in Iran a case of Mignon syndrome.[4]

Case report

C.S. is an unmarried 50-year-old patient, she has two older siblings, a sister and a brother. There were no particularly relevant events during her childhood and adolescence. They belong to a middle class, with a normal schooling and a good socialization. At the end of the Bachelor's degree in Economics and doing the doctorate courses, they promise her a doctoral scholarship in which she puts many hopes.

The first period of the disease began when she knew that she wasn't obtaining scholarship, she was 24 years old. This was experienced as a rupture of her life projects: "everything went wrong ", "they cheated on me". The acoustic hallucinations began, identifying the voices with a university teacher saying to be in love with her and that she had to work for him. Progressively other voices appeared that she also identified with University people, this voices saying that she was a prostitute or a Priestess. During this first period she entered in an increasingly restless state due to the hallucinations. For two years, without insight of the disease, she distanced from all labour activities, keeping increasingly disturbed and isolated until she was medicated with antipsychotics.

In the second period of the disease, 6 years, the antipsychotic medication did not cease the symptoms but only helped her being more calm. She also gained much weight, which also influenced her not wanting to leave home. With risperidone the hallucinations improved but it had to be suppressed because she started suffering from amenorrhea.

Acoustic hallucinations are imperative in second person and command her quit smoking in certain places, not ending the meal and not eating certain foods. She was also forced to do somethings without her will. She identifies one of the voices as the one that commands her the most and a second voice that rewards her if she manages to resist the first one: "you´ve won a shower, a coffee or a tea". It also appears delusions of allusive meaning (the TV is connected with the voices), delusions of control and of influence.

The delirious realization was the following: she got to understand that she was a Princess, daughter of the Kings of Spain. The voices say to her that in the future she will have a new life. The Kings know everything through the Masons and people of the University which she identified "by the accent of the voices". The voices terrify her because they are telling her that she will become sick and will suffer a depression disease.

In the third period of the disease that covers from 32 to 35 years old she had the antipsychotic medication reduced. When we saw her the first time (35 years old) she was only taking fluphenazine decanoate 25 mg/month. She lost weight and even though delusions and hallucinations carried on she was able to work in shops and stores during those three years.

Fourth period. When she was 35 years old we prescribed her risperidone (5 mg/day) and carbamazepine 200 mg 3 times a day. She improved significantly, returned to her job and although the acoustic hallucinations remained, she felt less overwhelmed by then. The control delusions happened only at the bar and the paranoid self-references "related to teachers because they knew the issue". But the amenorrhea and bulimia reappeared and she began to be unable to control the amount of food intake. Thus we decreased the dose of risperidone to 2 mg/day and 60 mg at breakfast and dinner of Ziprasidone were introduced.

Three months later the voices are heard "only occasionally" (at lunchtime and at bedtime), she remained being active and with very little delusions of control. She decreased the bulimia events and had twice the menstruation.

Since she is 37 years old she feels active, having monthly the menstruation, she works in a trade and has a good relationship with her colleagues. The hallucinations are only of imperative type and only when she is at home: "don't eat that", "don´t wear that!", but they have no much emotional

impact on her and she deals to "ignore them". Delusions are encapsulated without affecting her life although its remains as a reality.

With 39 years old she only has hallucinations at night before sleeping and being sometimes imperative and sometimes dialoguing (hearing conversations "between them they speak").

She stopped taking medications with 40 years old and starts to suffer again from psychotic symptoms and hallucinations which overwhelm her a lot because of their intensity. She has them when she goes down the street, she notices self-paranoid references in her workplace and delusions of control of people crossing the road.

We reintroduce the medication with carbamazepine 600 mg/day, Ziprasidone 80 mg 3 times a day and Risperidone 3 mg/day. Yet it takes two years to gain the prior state before quitting the medication. With 42 years old the hallucinations are only "a loose comment" and the paranoiac delusion is mild and she says "I control it". She continues working and resume her social relations, but the amenorrhea return (prolactin 68.33 ref. 3.4-24.1 ng/ml). The delusion was again encapsulated.

When she is 47 years old without any specific trigger the intense imperative voices reappear when she is at home, on the street, at work and when she´s less active. The hallucinations refer to her directly (in second person) or are dialoguing voices. We replaced Risperidone by Paliperidone (3 mg/day) and add Aripiprazole (15 mg/day) to the treatment. After two months she recovered, having low intensity voices they didn't overwhelm her too much and which appeared only when she was more stressed or nervous at work. With this improvement she remains until today.

Complementary tests

Electroencephalography study (EEG): Normal.

Analytical study: Prolactin, first analysis 92.19 ng/ml (Ref. 3.4-24.1 ng/ml) taking 5 mg a day of Risperidone. Second analysis of prolactin 81.43 ng/ml taking 2 mg a day of Risperidone and taking 120 mg of Ziprasidone. When she is taking Paliperidone 3 mg/day, Prolactin: 11.14 ng/ml.

Discussion

the pathoplasty or pathoplasticity determines the form of a disease. Pathoplasty is the shaping of the disease. Pathoplastic factors refers to what causes the variation in the disorder between individuals or cultures.

It has been described cases of monarchs who have had a mental illness,[5] but in a country like Spain with no monarchy for many years (until 1978) no delusional paranoid of monarchical identification has been described. This only appeared after the restoration of the monarchy. It's a rare case in psychiatry and clearly a realization of the patoplastia of our era.[6]

In the case described by Barrowclough and Tarrier[7] the patient was convinced a member of the Royal family was inhabiting his body and it was therefore his duty, as a gentleman, to vacate it.

In other cases it has been described "delusional memories" in which suddenly the patient remembers something that makes him understand that he belongs to royalty. As in the case described by Mckenna PJ[8], a patient realised he was of royal descent when he remembered that the fork he used as a child had a crown on it. Also Rojo-Sierra describes a patient who said: "I remembered that when I was doing the military service and the King was reviewing, he looked at me. Then I fell into the account that the King wanted to tell me with that look that I am his son". In this case, a real memory has suddenly acquired a new meaning.[9]

Also H.E. Nelson (1997) describes a patient that believed to be son of a Royal Prince and this caused problems at home because talking about this royal connection caused rows between him and his father.[10]

In our case the paranoid disease is primary, it's not an experience of bodily influence or delusional memory.

In the case described by M. Sanati (1992), successive drug trials with Trifluoperazine 30 mg/day, Haloperidol 30 mg/day and Thiothixene 30 mg/day, failed to remove his delusions but ameliorated his depression and agitation.[4]

Our patient had a very good response to Risperidone combined with Carbamazepine. The side effects of Risperidone (amenorrhea and bulimia) made us introduce Ziprasidone which not only maintained the clinical improvement that had been achieved, but also improved still further the patient. However, hallucinations and delusion never entirely disappeared and the delusion got encapsulated.

Over 15 years of evolution it has not been possible to reduce medication and when she stopped taking it we need more doses of antipsychotics to get the improvement that she had previously. Occasionally, during these 15 years, she took other antipsychotics such as Chlorpromazine or Quetiapine but with no improvement.

The syndrome of Mignon and the Capgras/Sosias syndrome (having parents or relatives being replaced by other people) oftenly go together but not in our case.

Conclusions

Our case of psychotic royal family identification (Mignon delusion / genealogical delusion) it is not very common and it is really a rare psychiatric case in Spanish literature. It is significant the positive evolution of this psychotic patient, who currently performs with normality her labor and social relations, with a remaining encapsulated psychotic delusion without significant influence on her life. But certainly, after 15 years, the antipsychotics have been unable to completely eliminate the hallucinations neither the encapsulated delusion.

CONFLICT OF INTERESTS

The authors declare not to have conflicts of interest.

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Steroid-responsive auto-immune encephalitis as a paradigm of neuropsychiatric differential diagnosis: case report and review

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Dear editor,

Autoimmune encephalitis (AE) has gained clinical attention over the past years, especially since the identification of autoantibodies directed at brain structures, which led to the emergence of several new clinicopathological entities[1]. These advances have made it possible to identify previously unrecognized clinical conditions in patients with a wide range of unexplained neuropsychiatric symptoms and signs, often leading to psychiatric hospitalization[2].

There is a wide differential diagnosis for encephalopathy (including metabolic conditions, paraneoplastic syndromes, idiopathic autoimmune limbic encephalitis and others) but the clinical features and findings in several diagnostic tests (such as blood and cerebrospinal fluid analysis, electroencephalography and neuroimaging) often lead to an accurate diagnosis[3]. However, unawareness on these conditions and frequent early psychiatric symptoms often cause under and misdiagnosis[4].

We report a clinical case of a patient initially evaluated in a psychiatric setting who turned out to be diagnosed with steroid-responsive autoimmune encephalitis associated with autoimmune thyroiditis (SREAT), a subtype of AE. We further review the current perspectives on these conditions and their impact on psychiatric practice.

Case Report

A 31-year-old woman, with an unremarkable medical history besides a previous episode of undefined depressive-anxious disorder, and without any current medication, was admitted in a metropolitan psychiatric emergency de-

partment (ED) due to a 3-day behavioral disturbance. Recently, the patient and her family had a minor flu-like syndrome, without need of specialized medical care. Two days previously to the admission, her mother noticed she was "quite different": that day, she left work without warning, something she never did before. Next day, contrary to her usual routines, she did not prepare dinner for her family and went to bed early. The next morning, she abandoned home, leaving her 2-year-old daughter alone, and drove her car until a minor accident occured, to which she presented post-event amnesia. She then wandered in the vicinities, until she was intercepted by some neighbors, who noticed her speech was incoherent. They took her to the nearest general ED, where basic tests were performed, including blood alcohol concentration, urinary drug screening and brain CT – all normal, except for a slight increase of aspartate transaminase. There was no history of voluntary or accidental exposure to drugs or other toxic substances.

The mental state examination revealed that the patient was alert, slightly apathetic, disoriented to time and space, with a defensive posture. Affect was labile, at times incontinent. The speech was disorganized and incoherent; she claimed not to remember the car crash, while declaring persistently that a neighbor and a work colleague had installed several cameras inside her house, revealing likely persecutory delusions; she also described insomnia in the previous days. It was not possible to confirm or exclude hallucinations. She expressed hetero-aggressive ideation associated with the persecutory delusions, stating that she wanted to kill her neighbor and colleague; no suicidal ideation was found. She lacked insight for her condition.

The acute onset and the disorganization of affect, speech and behavior, raised the hypothesis of a dissociative disorder. However, according to the patient's husband, there were no recent relevant stress factors; also, the absence of previous significant psychiatric personal history and the rapidly aggravating evolution, made this diagnostic hypothesis unlikely. For that reason, a neurologic evaluation was demanded.

The neurologic examination revealed slight symmetric hyperreflexia of the four limbs, without other relevant clinical findings in addition to the previously described. The patient was apyretic. For further assessment and treatment, the patient was hospitalized in the Department of Neurology.

During her stay blood tests were performed, disclosing high levels of anti-thyroglobulin (235.4 UI/mL) and anti-peroxidase (15.7 UI/mL) autoantibodies; other immunological tests were negative (including anti-nuclear, cell-surface, and onconeuronal autoantibodies). Syphilis and HIV infection were excluded. Cerebrospinal fluid (CSF) analysis was unremarkable. Other tests including brain MRI, electroencephalogram, and abdominal and pelvic ultrasound were normal. Thyroid ultrasound was suggestive of thyroiditis, despite normal thyroid function tests.

In this patient, the acute onset of altered mental status, short-term memory loss and psychiatric symptoms with delusions suggested the diagnosis of autoimmune encephalitis. Given the finding of thyroiditis and the presence of above-normal serum thyroid antibodies, plus the exclusion of alternative causes which could mimic autoimmune encephalitis, a probable diagnosis of autoimmune encephalitis associated with autoimmune thyroiditis was made and the patient was prescribed IV methylprednisolone 1g/day for 5 days, followed by oral prednisolone 1mg/kg body weight. There were no relevant complications during her stay and an extraordinary complete remission of neuropsychiatric symptoms was observed by the second day of IV treatment, thus confirming the diagnosis of SREAT.

The patient was re-evaluated a few months later in the neurology outpatient clinic. She remained asymptomatic.

Discussion

SREAT is a rare specific type of AE associated with overt or subclinical thyroid disease, most commonly hypothyroidism. It was first described in 1966 as Hashimoto's encephalopathy[5], and it is still a controversial entity, deemed immune mediated and with good response to immunosuppressive drugs, being frequently considered on the differential diagnosis of autoimmune encephalitis. Its incidence is not well established, but probably remains underdiagnosed. Women are predominantly affected, and the age of presentation varies widely[6].

Serum levels of anti-thyroid autoantibodies (anti-thyroid peroxidase and/or anti-thyroglobulin) do not correlate with disease severity, and thyroid function can be normal. According to recent literature reviews[3,6], CSF analysis may be normal or show non-specific findings, most likely slight hyperproteinorraquia; electroencephalogram findings were abnormal in about 87% of cases, showing mainly diffuse slowing without epileptic activity, consistent with encephalopathy.

The clinical picture is pleomorphic, often with neuropsychiatric symptoms and signs, and its presentation may vary from mild confusion to status epilepticus or coma, thus the diagnosis is often challenging[6]. Psychotic symptoms are reported to occur in about 25% of cases, while seizures, confusion, and memory impairment are the most common clinical findings (but esteemed to occur in less than 50% of

cases); only about 10% of cases present isolated psychiatric symptoms[3].

Core diagnostic features include cognitive impairment with high levels of antithyroid autoantibodies and steroid responsiveness (up to 90% of cases)[6].

Its pathophysiology is not well-understood; a direct causal relationship between thyroid antibodies and encephalopathy is unlikely[7], and it is still unclear whether the reported cases represent only encephalopathy of other etiology with co-occurrence of high levels of anti-thyroid auto-antibodies, which is a common marker of autoimmunity in many autoimmune neurologic disorders[8] and may be present in up to 13% of healthy individuals. Several other etiologies (immunological and vascular) have been proposed and it is likely that more than one mechanism could lead to an autoimmune or inflammatory encephalopathy responsive to corticosteroids[3].

We present a clinical report of a case of SREAT, initially suggesting a possible dissociative or psychotic disorder. Atypical clinical features for a purely psychiatric disorder led to a neurologic evaluation, further confirming the etiology. This report typifies a paradigmatic neuropsychiatric case, demanding thorough psychiatric and neurologic investigation to achieve a reliable diagnosis and effective treatment that, otherwise, could have been missed, with important clinical consequences.

CONFLICT OF INTERESTS

The authors declare that there is no conflict of interests regarding the publication of this paper.

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