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Asperger Syndrome
Can the disorder be diagnosed in the adult age?

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Introduction. Asperger Syndrome (AS) is characterized by a qualitative disorder of social interaction, a pattern of restrictive, repetitive and stereotyped behavior, interests and activities, with normal intellectual capacity and normal language skills in the areas of grammar and vocabulary. Since its inclusion in international taxonomies, there has been much controversy regarding its nosological validity.

Clinical case. A patient with a diagnosis of AS in adulthood is described. Results from the psychopathological, personality and cognitive functioning assessment are included.

Conclusions. Asperger Syndrome can also be diagnosed in adulthood and should be suspected whenever retrospective information and clinical assessment point to this diagnosis.

Key words:
Asperger, autism, differential diagnosis, comorbidity, neuropsychology.

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Síndrome de Asperger
¿Un trastorno del desarrollo diagnosticable en la edad adulta?

Introducción. El Síndrome de Asperger (SA) se caracteriza por la alteración cualitativa de la interacción social, un patrón de comportamiento, intereses y actividades restrictivos, repetitivos y estereotipados, con capacidad intelectual normal y unas habilidades lingüísticas normales en las áreas de gramática y vocabulario. Desde su inclusión en las taxonomías internacionales ha existido una gran controversia en torno a su validez nosológica.

Caso clínico. Se presenta el caso de un paciente en el que se realiza el diagnóstico de SA en la edad adulta. Se presentan los resultados procedentes de la exploración psicopatológica, la exploración psicométrica de la personalidad y la evaluación del funcionamiento cognitivo.

Conclusiones. Es posible diagnosticar el síndrome de Asperger en la edad adulta, especialmente cuando los datos retrospectivos y la exploración actual lo permiten.

Palabras clave:
Asperger, autismo, diagnóstico diferencial, comorbilidad, neuropsicología.

INTRODUCTION

Asperger Syndrome (AS) is characterized by the qualitative disorder of social interaction, a pattern of restrictive, repetitive and stereotyped behavior, interests and activities, with normal intellectual capacity and normal language skills in the areas of grammar and vocabulary. The disorder was first accepted in the Diagnostic and Statistical Manual of Mental Disorders in its fourth edition and is still present in its fourth revised edition.[1] Since its inclusion in the international taxonomies, there has been much controversy regarding its nosological validity.[2] Some authors continue to include it in the autistic continuum, while others consider it to be a disorder with its own entity.

Much of the current controversy regarding the nosological validity of the syndromes revolves around the significant overlapping existing between this and other generalized developmental disorders, especially with high functioning autism (HFA). Although differences do not seem to exist between both groups in the different clinical aspects, higher levels of anxiety have been observed in the AS group.[3] According to the DSM-IV-TR, the differential diagnosis of AS should be made with schizophrenia, autistic disorder, Rett's disorder, childhood disintegrative disorder, obsessive-compulsive disorder and schizoid personality disorder. Some authors[4] also indicate the need to consider the schizotypal personality disorder, attention deficit hyperactivity disorder (ADHD), non-verbal learning disorder, social-emotional learning disorder and even semantic-pragmatic disorder of

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communication. Table 1 shows the principal keys for the differential diagnosis.

The comorbidity of AS with mood disorders, both with bipolar and the depressive pictures, stands out. This could suggest that the social withdrawal entailed in the syndrome could be a depressogenic factor.[5] AS is associated in very few occasions with schizophrenia, even though many patients with AS may be erroneously diagnosed of psychotic pictures, possibly due to the presence of overevaluated ideas and bizarre behaviors.[4] Comorbidity also exists with the anxiety disorders[6] and specifically with obsessive-compulsive disorder (OCD). In fact, it has even been suggested that the dual diagnosis OCD-AS has such complexity in regards to the therapeutic management that it should be considered as a special subtype of OCD.[6] Cormorbidity with the personality disorders, such as schizotypal and schizoid disorder is another controversial subject since the latter could be considered as an adult form of AS.[7]

Neuropsychological characteristics

In general, it is considered that the neuropsychological deficits pattern in AS is similar to the Nonverbal Learning Disabilities Syndrome (NVLDS), frequently observing a delay in linguistic development and superior performance in non-verbal cognitive tasks.[8, 9] Furthermore, a verbal intellectual coefficient superior to the manipulative one is generally observed in subjects with AS, while this pattern in the opposite in high functioning autism (HFA).[9] On the other hand, only quantitative differences rather than qualitative ones have been suggested between patients with AS and HFA.[9] Thus, both groups would share a deficit in their meta-representation skills, abstract reasoning and nonverbal communication while patients with AS occasionally would obtain better performance in the tasks related with theories of the mind and abstract reasoning, this fact being attributable to their superior intellectual level and compensatory verbal strategies.

The following case describes a male patient with Asperger syndrome diagnosed in adult age. The interest of this case is because AS is a disorder initiating in childhood and is generally diagnosed during school age and linked to specific educational centers and occupations. Thus, it is extremely rare to identify it in an adult psychiatric facility. On the other hand, the case proposes diagnostic doubts, which will be discussed.

CLINICAL CASE

D.G. is a 23-year old young man who was admitted to the Psychiatric Acute Unit due to deterioration in his social withdrawal level and who manifested heteroaggressive intentionality with a firearm.

Clinical history

Only son, born of dystocic delivery (possible perinatal hypoxy). He had delayed psychomotor development, and did not start walking until 1-2 years and did not use understandable language until five years. During the preschool years, he initiated psychological treatment due to withdrawal and emotional non-expressiveness. He preferred to be alone, did not express affect, did not relate or communicate with his peers and also did not show interest for games or toys. He was indifferent towards his setting, above all for the social setting. During the school years, he had important learning problems, above all in calculation tasks. During this stage, he underwent a psychometric examination for his intellectual level, this showing a quotient superior to the mean.

In adolescence, the patient had problems integrating into his peer group. He was very withdrawn, practically mutistic and always focalized on very restrictive interests. He was only interested in reading and drawing. Furthermore, in that period, he had stereotypic rituals in his behavior and personal habits. The patient had an obsessive interest in keeping empty recipients in the refrigerator and reacted with hostility if anyone hindered his performing that ritual.

At 19 years of age, he was enlisted into Military Service, a few days after which he presented with a picture of psychotic symptoms consistent with experiences of influence, delusional ideas of self-reference, auditory pseudohallucinations and derealization. In the military hospital, he was diagnosed of Brief Psychotic Disorder and Schizoid Personality Disorder, and was referred to a Day Hospital (DH) for follow-up.

At 23 years, the patient was admitted voluntarily to our Psychiatry service due to worsening of his withdrawal and social isolation. Furthermore, he verbalized in the presence of his mother his intentions of taking a shotgun and ""doing away with everyone." On admission, he had a very stiff body posture, very limited facial expression and was akinesic and had some tic on the psychomotor level. His body expression was deficient and language quite strange, with strange prosody. He used very pedantic and formal vocabulary and he seemed to speak with a "foreign accent." He excessively prolonged the gap between response and seemed to be hyper-controlling the information offered. He stated that his only problem was "that it was hard for him to relate with the others" and that lack of sleep had deteriorated his appearance. As he explained, "extreme bags under his eyes" and unusual tension in his jaws" could be seen on his face,

Table 1 Keys for the differential diagnosis of AS with the most problematic diagnostic categories

AUTISTIC DISORDER	**ASPERGER DISORDER**
- It is generally diagnosed before 3 years of age - Intelligence quotient is generally low (in approx. 70% of the cases) - They generally lack language (approx. 25% of the cases.) - They lack high level obsessional interests	- It is generally diagnosed at older ages. - Intelligence quotient is generally normal or superior - They have language. Grammatically and semantically correct, although with some peculiarities - They have high level obsessional interests (mathematics, astronomy, drawing, etc.)
Autismo de alto funcionamiento	
- Their nonverbal communication skills are generally superior to their verbal skills	- The opposite: great deficit in nonverbal communication, with conserved verbal skills (similarities with the nonverbal learning disability syndrome)
RETT DISORDER	
- Gender-linked: it only occurs in girls - Specific pattern of progressive deterioration with deceleration of head growth and loss of previously acquired skills - Severe psychomotor delay	- Predominance in males - There is no specific pattern present in Rett Disorder and there is no loss of already acquired skills - There is no severe psychomotor delay
CHILDHOOD DISINTEGRATIVE DISORDER	
- Normal development until 2 years of age, with lost, before 10 years, of previously acquired skills (loss of language, bladder control, adaptive behavior, etc.)	- There is no loss of previously acquired skills (no problems with bladder control, no loss of language, etc.)
ADHD	
- There is no qualitative alteration of social interaction or in the pattern of interests, activities and behaviors	- In addition to attention deficit, with possible hyperactivity-impulsiveness, there is a qualitative alteration in social interaction and in the pattern of interests, activities and behaviors
SCHIZOPHRENIA	
- There are delusions and/or hallucinations - Altered language that responds to formal thought disorders such as illogicity and loose association - It is generally diagnosed in early youth - Affectivity and personal contact (empathy) are altered, but it is not the most important abnormality	- There are no delusions and/or hallucinations (and if there are, consider it as a dual diagnosis) - Speech may be altered, but less illogicity and loose association are observed - It is generally diagnosed in childhood - Affectivity and interpersonal contact are the most affected area (it is hypothesized that there is a central deficit in empathy or development of the theories of the mind.)
SCHIZOID PERSONALITY DISORDER	
- Pattern of restriction of emotional expression and keeping a distance in social relationships. WITHOUT there being alteration in the pattern of interests, activities and behaviors	- The social isolation is more pronounced and also the emotional lack of expressiveness. Furthermore, alteration in the pattern of interests, activities and behaviors is added (and the other criteria proposed that have still not been included in the DSM-IV)
SCHIZOTYPAL PERSONALITY DISORDER	
- Cognitive and/or perceptive distortions as well as the eccentricities of the behavior	- Eccentric behavior, due to the unusual imaginative capacity and of the peculiar obsessional interests can be observed, but there are no cognitive and/or perceptive distortions (and if there are, consider the dual diagnosis)
OBSESSIVE-COMPULSIVE DISORDER	
- There may be restricted interested in the subjects related with the obsessions - There is no social or communication deficit - There are no sensorial alterations or coordination deficits - There is stress associated to the obsessional subjects or to the compulsive interests	- The restricted interests are a comprehensive part of the syndrome - The subjects of the obsessional interests are physical type (e.g. work instruments), never human or psychological - There is no stress associated to the idiosyncratic interests

something that "stood out" and that others commented about and made fun of him. A clear auto-referential ideation was observed in this theme.

The patient stood out by his interest and skill in writing and above in drawing. He surprised the professionals and patients with fantastic and creative comics. He was prescribed antipsychotic medication with atypical neuroleptics, but refused to take it, complaining about their side effects and hypersensitivity to the sedative effects was observed. D.G. self-defined himself as "very egoistic" and "very selective" in regards to his limited friendship relationships. He mentioned being very fond of reading and being very interested in ideological and political subjects. He expressed, although very concisely, anti-Marxist ideology that was close to the thinkings of Hitler.

Psychometric and neuropsychological evaluation

- **Minnesota Multiphasic Personality Inventory (MMPI-2).** Clinically significant elevation on the *Psychopathic Deviate (Pd), Schizophrenia (Sc), Depression (D) scales.* On the Morey personality disorders scales, he obtained an elevated score on *schizoid personality disorder*. This combination of scales is frequently observed in very isolated schizoid personalities with social intelligence problems who lack empathy, and who are generally unpredictable and unsatisfied.
- **Wechsler Adult Intelligence Test (WAIS-III).** Total intellectual quotient of 107 (normal-middle intellectual level), score on verbal scale of 103 and score of 111 on manipulative scale.
- **Continuous Performance Test (CPT-II)**. Profile of scores consistent with sustained attention deterioration. Lack of attention, without impulsivity and without vigilance alteration were observed.
- **Wisconsin Card Sorting Test (WCST).** The scores (failures in 77% if the trials and 45% perseverative errors) indicated alteration of the executive functions.
- **Rey auditory verbal learning test (RAVLT)** No difficulties in immediate verbal memory was observed and an ascending learning curve, indicative of normality, was obtained.
- **Verbal fluency test with phonetic code (FAS)**. Verbal fluency decreased.
- **Trail Making Test, Form B (TMT-B).** The patient performed the test in a time superior to that of his age group, indicative of poor visual-manual coordination.

On discharge, a clear improvement in the relationship and expressive setting and of the management of criticism of megalomanic delusional ideas was observed. He was diagnosed (for the first time in his life) of Asperger Syndrome. The diagnostic hypotheses of this patient are AS (versus autistic spectrum disorder), delusional disorder and/or schizotypal personality disorder.

DISCUSSION

Based on these data, many diagnostic doubts arise. AS is a Pervasive Developmental Disorder or PDD that is diagnosed in childhood or adolescence and our patient did not receive this diagnosis, although he was treated for being "extremely timid and with social withdrawal." On the other hand, the syndrome is similar to a schizoid personality disorder (or schizotypal) that should be diagnosed in the early youth, the time of evolution in which our patient was found when he was admitted to our service. Therefore, what diagnosis should be made? Should we consider the retrospective information we have and thus diagnose in an adult subject a disorder having its initiation in childhood? Should we only consider the DSM-IV-TR criteria according to which perhaps we should make some addition diagnosis to explain all the symptoms observed or should we consider the criteria proposed by several authors and make a single diagnosis?

We have retrospective information according to which an onset of the symptoms in childhood can be confirmed. Although not recorded in the personal data, the current symptoms of the patient have an onset in the pre-school period. In the second place, the clinical and neuropsychological examination data highly coincide with the data from most of the studies on patients with AS. Furthermore, the current symptoms of the patient, although they overlap with that of some disorders on axis II, do not totally fit in with the criteria defined for the latter and do adapt to the clinical characteristics of the AS. In the third place, we have the information provided by different professionals and by the associations of patients and family with the disorder that indicate the high frequency with which these persons are poorly diagnosed due to the confusion created by the comorbidity of the syndrome and the laxity of the criteria existing in the DSM-IV-TR and the ICD-10.

Although a development disorder should never be diagnosed based on a certain cognitive profile, the results obtained are, largely, those frequently observed in AS. Some authors[10] have recently found data that confirm an elevated episodic memory and good verbal memory. In the patient, this skill was observed more during the interview (he could recall more or less relevant dates with surprising accuracy) than in the Rey test (normal score). In any event, much similarity between AS and non-verbal learning disorder has been described, it standing out that the learning deficits, if they occur in subjects with AS, cannot generally be explained by their incapacity to acquire verbal information.[8, 9, 11, 12] The alteration in visual-manual coordination[8, 9, 13] and alteration

in attentional capacity have been extensively documented.[8, 9, 14] In this aspect, subjects with AS are similar to those with autistic disorder and ADHD. In regards to the alteration in the executive functions, this is one of the cognitive deficits that has been more corroborated in recent works in which patients with AS are compared with autistic patients, with ADHD and even with schizophrenia.[15]

In the Wechsler Intelligence Scale (WAIS-III), some results that also coincide with those found in studies published in recent years are obtained. These are, for example, a score on the Cube subtest that is superior to the mean and an elevated score in vocabulary. In the responses to the latter subtest, very formal definitions "from the dictionary" appear, since he uses pedantic and ostentatious language. The subtests that evaluate skills to understand social situations are among the lowest. This information reflects what for many authors is the core pathology of the patient, the deficits in the social and affective sphere.

CONCLUSION

Considering all these data and in line with that of the authors who take the position of the need for a more detailed description of the disorder in the currently diagnostic classifications, we orient this patient as having a case of Asperger Syndrome [299.80], without making any additional diagnosis, since we consider that all the symptoms observed are explained with that syndrome. However, more research in this area is needed in order to extend the criteria and to be able to avoid diagnostic errors in these patients. The great symptomatic variety of the syndrome, together with the limited specificity of the criteria contemplated at present in the diagnostic classifications make the diagnosis of AS highly unlikely, if not impossible. Unfortunately, many individuals with this are erroneously diagnosed and treated, with the consequences that this entails.

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