CASE REPORT

Paroxysmal non-kinesigenic dyskinesia, post-streptococcal syndromes and psychogenic movement disorders: a diagnostic challenge

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SUMMARY

We report a case of a 15-year-old boy presenting with sudden attacks of hyperkinetic movements of the limbs, trunk and neck. Clinical features were suggestive of paroxysmal non-kinesigenic dyskinesia, but the elevated antistreptolysin O antibody titre and history of recurrent upper airways infection led us to consider a post-streptococcal syndrome as a possible diagnosis. The patient started therapy with benzathine penicillin, sodium valproate and clonazepam without any significant improvement. A successive psychiatric assessment revealed the presence of a psychogenic movement disorder. Psychodynamic psychotherapy and individual counselling were started with progressive improvement of psychological symptoms and gradual resolution of hyperkinetic episodes, without any recurrence recorded during the follow-up (10 months).

BACKGROUND

It is important to not underestimate the possibility of a psychogenic movement disorder (PMD) during the early diagnostic stages in a case such as this, because of the decreased probability of a positive outcome when the psychotherapeutic approach is delayed and because the condition requires different management strategies as compared with organic movement disorders. A correct differential diagnosis is therefore crucial to avoid useless and potentially harmful drug therapies in paediatric patients.

The clinical features of movement disorders can be various and heterogeneous, and correct clinical diagnosis is often a challenge, even for expert neurologists. Paroxysmal dyskinesias are defined as disorders of the nervous system that cause transitory periods of involuntary movement, and can be classified as kinesigenic dyskinesia (induced by sudden movement), non-kinesigenic dyskinesia and exercise-induced dyskinesia. These three types of dyskinesia can have different clinical presentations and ages of onset, but the distinctive feature in all three is the recurrence of abrupt attacks of involuntary movement such as chorea, athetosis, dystonia or ballismus, without loss of consciousness.^{1 2} Paroxysmal non-kinesigenic dyskinesia (PNKD) can be sporadic, familial,³ or secondary to some organic cause or pathology,⁴⁻⁶ such as group A β -haemolytic *Streptococcus* infection in children.

In particular, Sydenham's Chorea (SC) and paediatric autoimmune neuropsychiatric disorders (PANDAS) are common entities associated with

streptococcal infection,⁷ which sometimes can go unrecognised.⁸

SC is a well-defined disease, characterised by involuntary choreiform movements sometimes associated with behavioural or psychiatric symptoms, occurring 1–6 months after streptococcal infection.⁹ Young patients display syndromes ranging from mild to severe in intensity, including wide ballistic purposeless movements and, possibly, inability to walk.⁸ SC can be associated with mild hyperintensity in T2-weighted images of the basal ganglia in MRI of the brain studies,¹⁰ and antibasal ganglia antibodies have been detected in serum of 46% of children affected by rheumatic chorea.¹¹ SC is a clinical diagnosis and is usually self-limiting and not harmful or irreversible.⁸ Neurological manifestations can be controlled with dopamine receptor blockers or antiepileptic drugs such as valproic acid or carbamazepine;¹² moreover, antibiotic prophylaxis treatment with penicillin is indicated in order to reduce the risk of developing carditis or valvular disease.^{9 13}

PANDAS has been described as a defined entity in 1997¹⁴ and is characterised by the sudden appearance of tics and/or obsessive-compulsive disorder after a recent group A β haemolytic *Streptococcus* infection. Unlike SC, PANDAS is a relapsing-remitting disease usually without cardiac involvement, hence antibiotic prophylaxis is not strictly required. Neurologic and psychiatric symptoms are generally well-controlled with haloperidol.^{8 15 16}

PMDs are heterogeneous manifestations of motor disturbance, not justified by organic conditions and usually associated with underlying psychiatric disease.¹⁷ The hyperkinetic attacks often start in early childhood and can be precipitated by triggering factors such as alcohol or caffeine, fatigue, anxiety, distress or excitement.^{18 19} Neurological examination performed between episodes is normal and commonly no alterations in MRI can be detected.

Diagnostic criteria were defined by Fahn and Williams.²⁰ They identified four levels of certainty for the diagnosis (Documented, Clinically established, Probable and Possible) on the basis of clinical characteristics of atypical movements and of the response to psychotherapy and psychological suggestion.^{20 21} In particular, movements should be inconsistent over time and incongruent with typical neurological symptoms, and present specific clinical features (acute onset, spontaneous remission,

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unresponsiveness to appropriate medication, responsiveness to placebo, movement increasing with attention and decreasing with distraction).^{20 21} These kinds of motor disturbances can be associated with specific psychiatric disorders,²¹ but it is important to consider PMD as a possible diagnosis even if there is no evidence of psychiatric disease or psychological distress and if clinical data support an organic pathogenesis, as PMD can be associated with different medical conditions, such as viral infection, trauma, encephalitis or substance intake.²²

Moreover, diagnosis can be difficult because comorbidity with organic movement disorders has been described in 10% to 25% of cases.^{17 22} The most common motor presentations are tremor, dystonia, gait disorders and myoclonus,^{17 23 24} but parkinsonism, chorea and tics have also been reported.²³ Psychogenic paroxysmal movements have been described and differential diagnosis with PNKD could constitute a diagnostic challenge.²⁵

CASE PRESENTATION

A 15-year-old boy was admitted to our ED reporting of recurrent episodes of abnormal movements for 5 days. He presented with sudden attacks of wide involuntary aimless movements of the limbs, trunk and neck, sometimes with ballistic or choreic features, and sometimes with clonic aspects (video 1). During the attacks the patient was conscious and oriented in time and space (video 2), but was unable to walk, stand or sit. The abnormal movements were not precipitated by any triggering factor such as psychological stress or sleep deprivation, but they occurred mostly in the morning, with a frequency of 3–4 times a day. The duration of each episode was a few minutes to up to 20 min, and the episodes did not occur during sleep. The patient was admitted to the ED of another hospital, 4 days before our medical examination, where he underwent a head CT scan, the result of which was normal. He started therapy with delorazepam for a supposed psychological aetiology, without any improvement.

The patient did not report major illnesses or recent diseases other than frequent upper respiratory tract infections during the previous winter, and denied history of brain injury, intoxication, medications or drugs intake. There was a family history of depression.

INVESTIGATIONS

On physical examination, the boy had normal vital signs; his musculoskeletal system, heart, lungs and abdomen were normal.

Video 1 Fragment from a hyperkinetic attack; the patient presented with involuntary movement of the head and limbs, with clonic contractions of the head and neck in all directions, and ballistic movements of the limbs and the body.

Video 2 The patient maintained consciousness during hyperkinetic attacks and answered the physician's questions about his age and birthdate.

Throat examination showed mild pharynx hyperaemia. His neurological examination, performed between the hyperkinetic attacks, was normal and did not reveal any extrapyramidal or cerebellar signs. During the episodes of dyskinesia the patient was awake, oriented and compliant, without impairment in any cognitive domain. Results of blood cell count and chemistry panels were normal, except for an elevated antistreptolysin O antibody titre. Throat culture was negative for group A streptococcus. An EEG showed sporadic slow frequencies in anterior fields, more evident in the left hemisphere. Brain magnetic resonance (MRI) and echocardiogram were normal. A lumbar puncture was performed and cerebrospinal fluid analysis showed normal protein level and cell count. No neurotropic viruses were detected. Thyroid function tests, immunological tests, copper and ceruloplasmin levels were normal. Genetic testing for Huntington disease revealed normal CAG expansion on chromosome 4.

TREATMENT

Considering a post-streptococcal syndrome as a possible diagnosis, the patient began therapy with benzathine penicillin (1.2 million units, intramuscularly, every 2 weeks, for 6 doses), sodium valproate (300 mg, orally, twice a day) and clonazepam (0.5 mg, orally, three times a day). The patient reported only mild improvement in duration and frequency of the hyperkinetic attacks. Therefore, despite his parents' disapproval, the patient underwent a psychiatric assessment that revealed mild depressive symptoms, difficulty in waking up in the morning with severe drowsiness and progressive isolation from friends since the beginning of high school. In particular, many problems seemed to be associated with the development of abnormal movement attacks, such as poor scholastic performances and feelings of unsuitableness and incompetence.

OUTCOME AND FOLLOW-UP

Psychodynamic psychotherapy and individual counselling were started, which lasted for 8 months. The patient experienced progressive improvement of psychological symptoms and gradual resolution of hyperkinetic episodes, without any recurrence, for the following 10-months' follow-up.

DISCUSSION

We described a young patient displaying sudden attacks of hyperkinetic movement suggestive of PNKD. In our patient, we investigated several different diagnostic hypotheses such as

thyrotoxicosis, encephalitis, Wilson disease, autoimmune or rheumatic disorders, epileptic syndromes and Huntington disease, without any positive result. Brain imaging and interictal scalp EEG were normal and there was no family history of movement disorder. Hence, the patient's medical history of recurring upper airways infections and his elevated antistreptolysin O titre oriented us toward a diagnosis of a post-streptococcal syndrome.

Our patient did not meet the diagnostic criteria for PANDAS because of the lack of tics and obsessive-compulsive disorder. At the same time, his neurological presentation was atypical for SC, as he had remitting-relapsing episodes and normal neurological findings between attacks; moreover, the results of a cardiological examination were found to be normal. However, his medical history and the elevated antistreptolysin O titre prompted us to start specific treatment with benzathine penicillin, valproic acid and clonazepam. Our decision was also driven by the fact that clonazepam and sodium valproate are known to be effective in the treatment of idiopathic PNKD,^{18 19} and supported by the description of a case of paroxysmal dyskinesia and tic disorder associated with group A β haemolytic *Streptococcus* infection.⁷ The young patient experienced only a mild reduction in frequency and duration of hyperkinetic episodes. Meanwhile, with some delay, we persuaded the patient and his parents to undergo a psychological and psychiatric assessment, which revealed mild depression, sleep impairment and a possibly conversive origin of the hyperkinetic episodes.

Excluding the atypical clinical presentation, our patient met the diagnostic criteria for PMD. In fact, his movements were various in performance, amplitude and distribution during different hyperkinetic attacks, with acute onset and remitting relapsing presentation. He did not display psychological physical signs, but he presented mild depression and conversive symptoms. Finally, his motor disorder recovered with psychodynamic psychotherapy and individual counselling.

While some authors report poor prognosis in patients with long disease duration, our patient experienced complete remission despite the relatively long period (3 months) between the onset of symptoms and diagnosis.

According to the literature, a multidisciplinary therapeutic approach to the patient affected by PMD gives satisfying results, in term of improvement or sometimes global remission of the motor disorder.²⁶ The importance of establishing a therapeutic alliance and gaining trust of the patient, who should correctly understand and accept the diagnosis, is underlined in the literature. The psychotherapy should be supported by physical therapy (especially for the suggestion-effect) and sometimes by pharmacotherapy (in case of association with a well-defined psychiatric disorder). Placebo, transcranial magnetic stimulation and hypnosis have been described as possible therapeutic approaches.^{27–29}

Learning points

- Consider psychogenic movement disorders as a possible origin of unusual movement disorders.
- Pay attention to possible misdiagnosis in order to avoid useless and potentially harmful drug therapies in paediatric patients.
- Reduce the time between symptom onset and diagnosis in order to improve prognosis.

In conclusion, our patient was treated as if suffering from a dyskinesia with organic aetiology, but the lack of a complete psychiatric assessment led us to a misdiagnosis. We therefore recommend that psychological and psychiatric assessment should be routinely carried out in patients, especially in children and adolescents, with unusual movement disorders.

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Competing interests None.

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