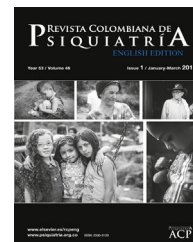




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Case Report

Dystonic storm in consultation-liaison psychiatry[☆]



Laura Canon Ángel^a, Laura Milena Saavedra Ramírez^b,
Francisco Javier Muñoz Molina^c, Gabriel Fernando Oviedo Lugo^{d,*}

^a Hospital Universitario San Ignacio, Pontificia Universidad Javeriana, Bogotá, Colombia

^b Departamento de Psiquiatría y Salud Mental, Pontificia Universidad Javeriana, Bogotá, Colombia

^c Facultad de Medicina, Departamento de Psiquiatría, Pontificia Universidad Javeriana, Hospital Universitario San Ignacio, Bogotá, Colombia

^d Facultad de Medicina, Departamento de Psiquiatría y Salud Mental, Pontificia Universidad Javeriana, Hospital Universitario San Ignacio-Centro de Memoria y Cognición Intellectus, Bogotá, Colombia

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ABSTRACT

Dystonia is a movement disorder characterised by sustained muscle contractions that produce repetitive twisting movements or abnormal postures. It can be classified according to the aetiology as primary (idiopathic and genetic forms), or secondary. The presentation associated with generalised, intense episodes and with exacerbation of severe muscle contractures and usually refractory to traditional pharmacotherapy is known as dystonic status or dystonic storm. In the present article, a case is presented of a 33-year-old patient with a history of congenital deafness, stimulant use disorder and on psychopharmacological treatment with antipsychotics, who presented with a severe dystonic reaction that evolved to a status dystonicus.

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Tormenta distónica en psiquiatría de enlace: a propósito de un caso

RESUMEN

Las distonías son trastornos del movimiento caracterizados por contracciones musculares sostenidas que producen movimientos repetitivos de torsión o posturas anormales. Pueden clasificarse según la etiología como primarias (formas idiopáticas y genéticas) o secundarias.

Palabras clave:

Distonía

Trastorno distónico

Tormenta distónica

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* Corresponding author.

E-mail address: goviedo@javeriana.edu.co (G.F.O. Lugo).

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Trastornos del movimiento
Tratamiento
Estimulantes del sistema
nervioso central

La presentación asociada con episodios generalizados, intensos y con exacerbación de contracturas musculares intensas que suelen ser refractarias a la farmacoterapia tradicional se conoce como *status* distónico o tormenta distónica. Se presenta el caso de un paciente de 33 años, con antecedente de sordera congénita, trastorno por consumo de sustancias psicoactivas y tratamiento psicofarmacológico con antipsicóticos, que presentó un cuadro distónico grave que evolucionó a un *status* distónico.

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Introduction

Dystonic storm, or dystonic status, is a condition that can appear in patients with primary or secondary dystonia. The symptoms consist of severe dystonic spasms and generalised rigidity, which are usually refractory to pharmacological treatment. Additional symptoms are rhabdomyolysis, anarthria, respiratory failure, dysphagia and, in some cases, hyperthermia. The picture can be complicated by acute renal failure and aspiration pneumonia. It is more common in males and those under 15 years of age. In 67.4% of cases, the triggering factor can be identified.^{1,2}

We describe the case of a 33-year-old man, evaluated in the emergency department and hospitalised, with a history of congenital deafness, substance use disorder and antipsychotic medication use, who presented a picture of severe dystonic storm from which several differential diagnoses emerged. The patient was evaluated and treated by the authors of the article. [Table 1](#)

Case report

The patient was a 33-year-old man who was admitted to the emergency department following a sudden onset of clinical symptoms of generalised dystonic movements. He had a significant history of congenital hearing impairment (anacusis) and psychoactive substance use disorder, mainly derived from cocaine use (high purity cocaine and base paste) from the age of 14. He was in outpatient psychiatric treatment due to psychotic symptoms related to substance use (diagnosis of substance-induced psychotic disorder at the referring institution), and had been taking clozapine 150 mg/day during the last six months, which had been suspended 15 days prior to this admission. During the institutional evaluation by our department, he was diaphoretic, tachycardic, without axial stiffness, with repetitive dystonic movements, mainly of the trunk and limbs, with alternating opisthotonus posture without altered state of consciousness ([Fig. 1](#)). A simple skull CT scan and then a gadolinium-enhanced brain MRI were requested, which were within normal limits.

Elevated total creatine kinase (CK) values of 4238 U/l were found (normal range, 55–170 U/l). A follow-up test was carried out at 12 h, which showed a progressive increase up to 9708 U/l. Drug tests were requested, which were positive for cocaine and benzodiazepines. A haemogram showed leukocytosis of $13.2 \times 10^3/\mu\text{l}$, with the other values within normal limits. A

lumbar puncture was performed, which reported 3 cells/ μl , without red blood cells, glucose at 54.5 mg/dl and proteins at 32.6 mg/dl. No germs or bacterial antigens were observed. Urinalysis showed myoglobinuria. Electrolytes, kidney function tests, VDRL test and blood cultures were all within normal limits. Differential diagnoses of dystonic storm, atypical neuroleptic malignant syndrome, serotonin syndrome and suspected strychnine toxicity were made.

Upon admission to the emergency department, a dose of 5 mg of haloperidol was administered along with 5 mg of midazolam for psychomotor agitation. After this there was an increase in the dystonic movements and the patient required transfer to the intensive care unit. Treatment with diazepam 10 mg/6 h was started, with an initial improvement in symptoms. However, due to the reappearance of paroxysmal dystonic movements and dysautonomic symptoms, it was necessary to start the patient on a midazolam drip, requiring up to 96 mg/day. Due to the risk of respiratory failure, orotracheal intubation was required. It was decided not to restart antipsychotic treatment due to the initial differential diagnosis of neuroleptic malignant syndrome.

The benzodiazepine dose was gradually decreased, with adequate tolerance and remission of the motor symptoms. Given the intermittence of symptoms such as abnormal movements and dysthermia, the multidisciplinary team considered that dystonic storm was the most likely diagnosis.

Discussion

Dystonic storm is a life-threatening, abnormal movement emergency. It is characterised as a hyperkinetic disorder that leads to severe dystonia. It was first described by Jankovic and Penn³ in 1982. Their patient was an 8-year-old boy with deforming muscular dystonia, who suffered an acute worsening of dystonia and elevated CK and serum creatinine together with myoglobinuria.

The diagnosis should be considered when severe generalised dystonia occurs in conjunction with hyperthermia, tachycardia, tachypnoea, hypertension, sweating and autonomic instability. Its presentation can be tonic or phasic. In the largest case series available, of the 89 episodes studied, 69% were tonic.¹ The risk of dystonic storm appears to be correlated with the severity of the dystonia and occurs more frequently in patients with poorly controlled baseline dystonia. Dystonic storm can occur as an idiopathic symptomatic expression or due to trauma, encephalitis, acute exposure to neuroleptics, or neurodegenerative disorders. The differ-

Table 1 – Aetiology of dystonic status.^{1,4}

Primary dystonia: DYT1 generalised dystonia, DYT1 negative primary generalised dystonia

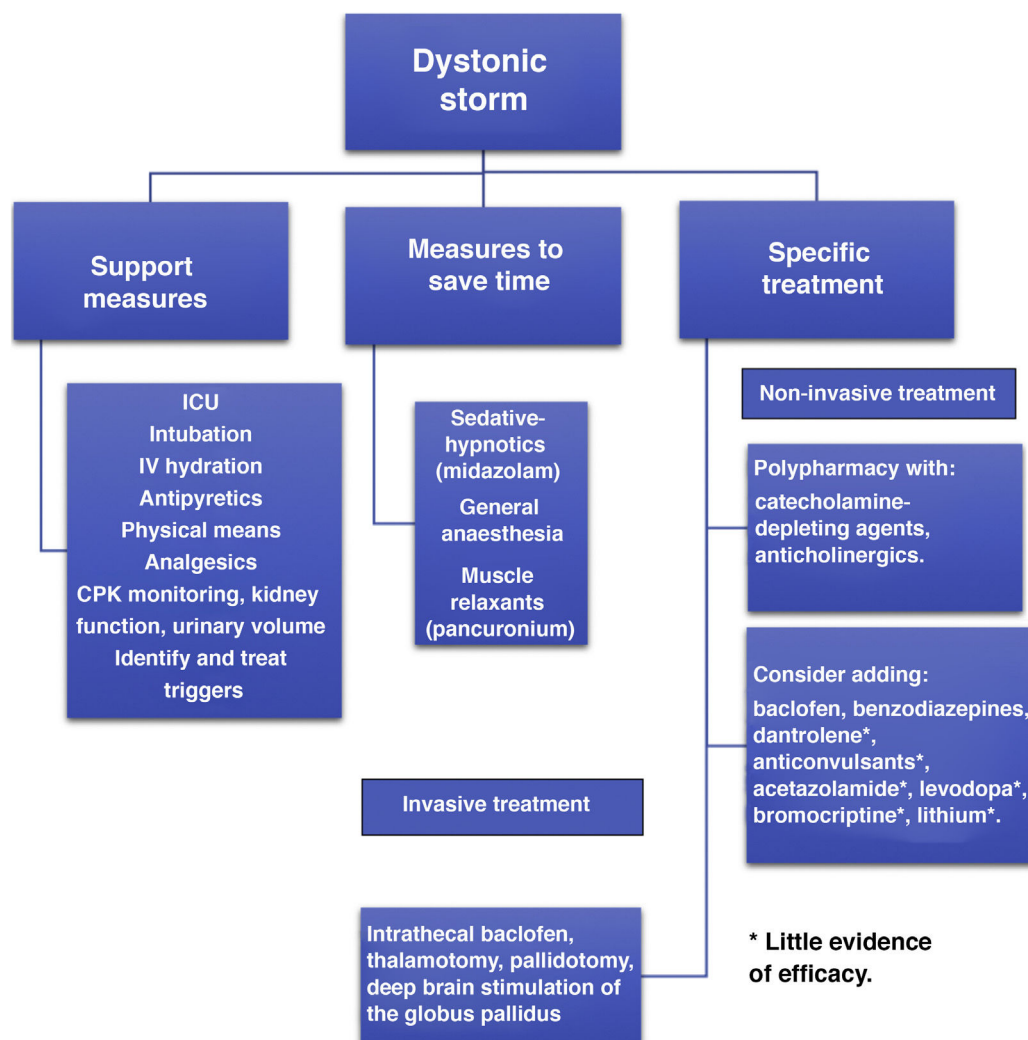
Brain damage: cerebral palsy, head trauma

Metabolic disorders affecting subcortical areas: mitochondrial diseases (Leigh syndrome), glutaric acidemia type 1, Wilson's disease, progressive hereditary degenerative dystonia

Neurodegeneration associated with pantothenate kinase, neuroacanthocytosis, ceroid neuronal lipofuscinosis

Miscellaneous: genetic syndromes (ARX syndrome, megalencephalic leukoencephalopathy with subcortical cysts (MLC), SOX2 anophthalmia syndrome, ataxia-telangiectasia, familial fatal hypoparathyroidism)

Drug-induced acute and tardive dystonia

**Fig. 1 – Patient in opisthotonus posture in dystonic storm.**

ential diagnosis should be made with bacterial meningitis, neuroleptic malignant syndrome, serotonin syndrome and withdrawal of baclofen. It often occurs after a trigger, such as fever, infection, stress, general anaesthesia or changes in medication. Medications that have been associated with it are dopamine receptor antagonists, penicillamine and zinc. Abrupt discontinuation of tetrabenazine, lithium or anticholinergic medications has also been associated as a trigger for dystonic storm.⁴

The pathophysiology of dystonic storm remains unknown. Termsarasab et al.⁵ propose a hypothetical model of the mechanisms underlying all abnormal movement storms.

Specifically for dystonic storm, they consider that the worsening of the dystonia can be caused by an increase in efferents of the globus pallidus that results in an increase in muscle tone mediated at the spinal level, fever, rigidity and an increase in CK.^{6,7}

In the conditions associated with antipsychotic medication, there is an acute saturation of dopamine D2 receptors. Preclinical studies indicate that sigma 1 and sigma 2 receptors found in motor regions of the brain that are related to postural control may be associated in all forms of dystonia.⁸ Recent use of stimulants (cocaine and derivatives, methamphetamine) has been associated with severe acute dystonia and dystonic



Fig. 2 – Treatment algorithm for dystonic storm.^{1,2,4,10} CK: creatine kinase; ICU: intensive care unit; IV: intravenous.

status.^{9,10} We consider that, in this patient, this circumstance played an important role in the development of the condition, due to the recent consumption of cocaine derivatives and high purity cocaine at the same time as the neuroleptic medication used for the treatment of chronic psychotic disorder.^{6,11} Clozapine has a low rate of movement disorders like dystonias and a therapeutic role in some cases of tardive dyskinesia, which may be related to the serotonergic blockade it induces. Serotonergic blockade boosts the availability of dopamine in the striatum and minimises extrapyramidal effects. Kovacs et al. reported a case of dystonic storm that remitted with deep brain stimulation (DBS) and sustained its improvement process with clozapine as concomitant treatment.^{12–14}

The therapeutic algorithm is summarised in Fig. 2.

Conclusions

Dystonic status is a rare condition that requires prompt diagnosis and treatment, considering that treatment is carried out in an intensive care unit to avoid metabolic, renal and respiratory complications.¹² Deep brain stimulation is a therapeutic

strategy that can be considered early. Clinical practice guidelines are required to systematically support the approach to this problem.

Conflicts of interest

The authors have no conflicts of interest to declare.

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