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Case Report

Challenges in Diagnosing and Managing Delayed Onset Paroxysmal Generalized Dystonias Associated with Bilateral Thalamic Hemorrhagic Venous Infarction Due to Extensive Cerebral Venous Thrombosis (CVT) in a 15-Year-Old Adolescent: A Case Study

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Abstract

Cerebral venous thrombosis (CVT) constitutes less than 1% of stroke occurrences. The prevalence of abnormal movements following a stroke remains unclear, with movement disorders observed after a stroke ranging from 13 to 22% of secondary disorders. Nonetheless, these disorders are present in only 1% to 4% of stroke cases, with dystonias being the most commonly linked abnormalities in ischemic strokes. The lesions that lead to dystonias primarily affect the basal ganglia, thalamus, brainstem, cerebellum, and specific cortical regions. Dystonias make up about 30% of abnormal movements observed after a thalamic stroke, with the lesions typically being unilateral. This research discusses instances of delayed paroxysmal dystonias related to bilateral hemorrhagic infarction in the thalamus of an adolescent with cerebral venous thrombosis. A 15-year-old adolescent was admitted due to a rapidly developing disturbance of consciousness. Upon arrival, he exhibited a non-massive left hemicorporal pyramidal syndrome, left-sided tonic seizures, and a state of confusion. Magnetic resonance imaging revealed a bilateral thalamic hemorrhagic focus and cerebral venous thrombosis. Anticoagulant therapy was initiated, and the clinical progression during the acute phase was satisfactory. Ten days later, the patient displayed generalized but asymmetrical paroxysmal dystonias, predominantly affecting the left hemibody. This case presented diagnostic challenges, as the abnormal dystonic movements were linked to focal tonic seizures, alongside management issues, given that most antidystonic medications are sedative in nature, complicating treatment due to the pre-existing disturbance of consciousness. Nevertheless, the patient's condition improved under cautious administration of anticholinergic agents and GABAergic agonists.

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1. Introduction

Dystonias represent a diverse array of hyperkinetic movement disorders characterized by involuntary and prolonged muscle contractions, leading to abnormal postures and repetitive movements [1]. This condition ranks as the third most prevalent movement disorder, following Parkinson's disease and essential tremor [2]. Despite the relative commonality of dystonic syndromes, they are frequently misdiagnosed [1]. Furthermore, advancements in the classification of dystonias have been made, alongside a deeper understanding of the underlying causes of dystonic movements, facilitated by research in genetics, neurophysiology, and functional imaging [1]. Various lesions are linked to secondary dystonias, with vascular lesions implicated in approximately 13 to 22 percent of cases [3, 4]. The structures commonly affected include the basal ganglia, thalamus, brainstem, cerebellum, and certain cortical areas, such as the parietal and frontal regions [3], suggesting a potential correlation between lesion location and the type of dystonia [3].

Pallidonigral lesions have traditionally been regarded as contributors to dystonias [5, 6]. However, damage to the centromedian and ventral intermediate nuclei of the thalamus has been linked to the onset of hemidystonias following a stroke, attributed to disruptions in the cortico-striato-pallido-thalamo-cortical loop [5]. This report presents a clinical case of paroxysmal generalized dystonia resulting from bilateral thalamic venous infarction in a 15-year-old adolescent. The case posed diagnostic challenges due to its association with epileptic seizures, as well as complications in therapeutic management, particularly concerning medication selection, given that the patient initially exhibited clouding of consciousness.

2. Case Presentation

The patient, a 15-year-old adolescent with normal psychomotor development and educational progress, has a significant medical history that includes septic cerebral venous thrombosis of the superior sagittal sinus, which occurred approximately seven months ago, as well as focal epilepsy as a consequence of this condition. Financial difficulties led to the cessation of both anticoagulant and antiepileptic therapies. The patient was readmitted to our department following a rapid deterioration in vigilance over a span of three days. Upon physical examination at admission, a state of obnubilation was observed (GCS score of 13/15), accompanied by focal epileptic seizures and signs of febrile meningeal syndrome. Magnetic resonance imaging (MRI) of the brain revealed extensive cerebral venous thrombosis extending from the left lateral sinus to the internal jugular vein, with intra-parenchymal effects manifesting as bilateral thalamic hemorrhagic venous infarcts. An anticoagulation protocol was established, in conjunction with targeted treatment for meningitis and the reintroduction of antiepileptic drugs.

The clinical progression demonstrated a decrease in seizure occurrences, yet the impairment of vigilance continued to be present. Approximately ten days post-admission, the patient exhibited abnormal movements, characterized by episodes of painful generalized contractions affecting the neck, trunk, and both upper and lower limbs, predominantly on the right side. These episodes occurred paroxysmally without impacting the baseline level of alertness, exhibiting a tendency to worsen throughout the day, ultimately leading to a diagnosis of late paroxysmal generalized dystonias.

The therapeutic approach entailed the administration of trihexyphenidyl at 6 mg in three doses, clobazam at 25 mg in three doses, and baclofen at 30 mg in three separate administrations. This regimen was followed for a period of 30 days, during which positive results were achieved. After this initial phase, clobazam was gradually reduced and ultimately discontinued, while the child continued to receive trihexyphenidyl and baclofen at the same dosages for an additional three months. At the end of this period, a near-total improvement in dystonias was noted, allowing for the cessation of trihexyphenidyl. However, some hypertonia persisted without any abnormal limb postures, necessitating the continuation of baclofen at a dosage of 20 mg administered in two doses.

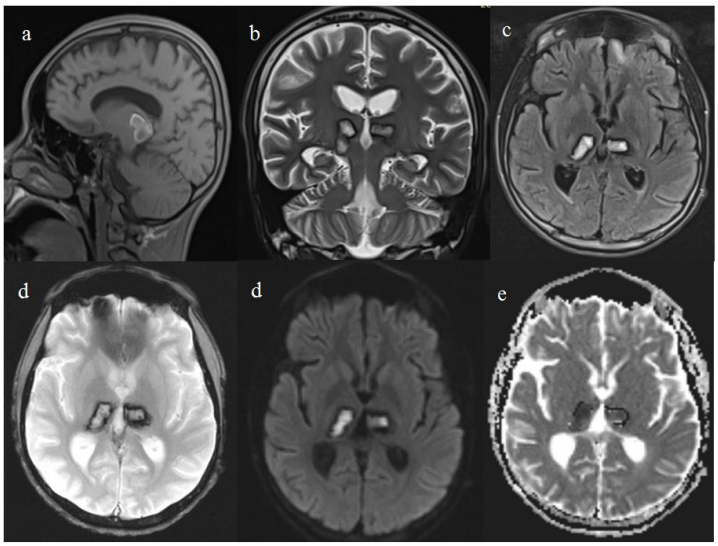

Figure 1. *Morphological sequences of brain MRI.*

Bithalamic hemorrhagic venous infarction at the late sub-acute stage presenting as heterogeneous hyper intensity on the sagittal cut in T1-weighted spin echo sequence (a), hyper intensity on the axial cuts weighted in T2 (b) and in FLAIR (c). Additionally, heterogeneous signal is observed, characterized by a hyper intense center surrounded by a hypo intense rim on the axial T2* cut (d), as well as hyper diffusion (d) and hypo ADC (e).

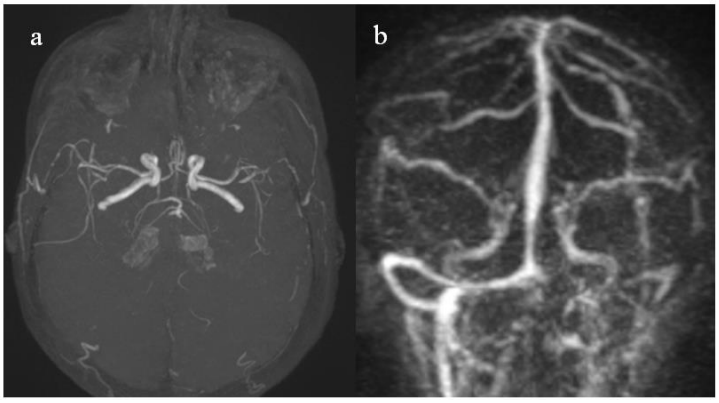

Figure 2. *Vascular sequences of the brain MRI.*

The intracranial arteries are patent on MIP (a) and there is a void in the right sinus and left sigmoid in venography (b), suggestive of extensive lateral sinus vein thrombosis on the left.

3. Discussion

In this report, we presents a case of late-onset generalized dystonia resulting from bilateral thalamic hemorrhagic venous infarction in a 15-year-old adolescent with left lateral sinus vein thrombosis. We discuss the diagnostic and therapeutic challenges associated with this condition. Cerebral venous thrombosis accounts for less than 1% of all strokes [7, 8] and predominantly affects children and young adults [9]. Thalamic hemorrhagic venous infarcts, whether unilateral or bilateral, are commonly linked to deep venous system occlusion [10].

The prevalence of movement disorders following a stroke remains uncertain [11]. A study involving 1,500 stroke patients indicated that 3.7% of them developed movement disorders [11], while the research conducted by Ghika-Schmid et al. reported an incidence of 0.08% [12]. Since Oppenheim's initial description of dystonia in 1911 [2], the clinical understanding of this heterogeneous condition has significantly advanced, largely due to the efforts of scholarly societies. The most recent definition, articulated by the International Parkinson and Movement Disorder Society in 2013 [2, 13], characterizes dystonia as a movement disorder characterized by sustained or intermittent muscle contractions that lead to abnormal, often repetitive movements or postures, or both.

Dystonic movements are typically patterned, twisting in nature, and may include tremors. This disorder is frequently triggered or exacerbated by voluntary actions and is associated with excessive muscle activation. The classification of dystonias is structured along two primary axes: the first axis focuses on clinical characteristics, while the second axis examines the etiology of dystonia [2, 13]. In our analysis, the phenomenology was indicative of paroxysmal generalized dystonias, characterized by exacerbation during the daytime at the onset of voluntary movements, with the etiology being attributed to acquired vascular dystonias.

In the study conducted by Suri et al., it was found that dystonias are the most frequently observed abnormal movements following a stroke, representing 23% of cases, while chorea and myoclonus account for 16% and 15% [4], respectively. The onset of dystonia can occur anywhere from one day to five years after a stroke, which may significantly impact the time required for partial recovery of motor functions and the formation of pathological circuits [3, 4, 14]. Dystonias are observed in 28% of ischemic stroke cases and 30% of hemorrhagic strokes [4]. Suri's analysis of abnormal movements following a stroke revealed that all cases of dystonia were associated with posterior-lateral thalamic damage [4]. This is consistent with findings from Navnika et al., who identified that the movements most commonly linked to thalamic lesions were dystonias (30%) and hemiataxia (21%) [14]. In a study involving 102 patients who exhibited dystonia following thalamic stroke, 32 cases (31%) were isolated, while 70 cases (69%) were associated with other movement disorders. The distribution of dystonia was focal in 51 cases, hemispheric in 7 cases, bilateral in 6 cases, segmental in 3 cases, and multifocal in 2 cases [14]. Dystonia has been predominantly linked to damage in the posterolateral thalamus, with lesions typically found in the ventroposterior and ventrolateral nuclei [14]. However, various studies have also documented instances of dystonia associated with lesions in the centromedian nucleus, a component of the intralaminar nuclei that projects to the striatum and forms the thalamostriatal pathway, in addition to those involving the striatopallidal complex [5, 6].

The most recent model concerning dystonia, developed by Neychev [15], emphasizes the dysfunction of the cerebellum as a central factor. This model posits that lesions in the cerebello-thalamic projections may lead to an imbalance between agonist and antagonist muscles, thereby resulting in dystonia. The centromedian nucleus receives inputs from the pedunculopontine pathways, while the pedunculopontine complex exerts influence over the ventrolateral nuclei of the thalamus [14]. In the study conducted by Navnika et al., delayed onset forms (exceeding 7 days) were the most prevalent, accounting for 64% [14]. Despite the presence of a hemorrhagic lesion in our patient, an increasing number of studies suggest that ischemic lesions are more frequently associated with the onset of dystonias [11, 14]. A hypothesis suggests that younger patients who have undergone a hemorrhagic stroke are more predisposed to the development of movement disorders, owing to their heightened resilience and neuroplasticity [4, 14]. Nonetheless, the emergence of these disorders may be postponed based on the extent of the hemorrhage [4, 14]. It is noteworthy that in both hemorrhagic and ischemic strokes, approximately 70% of abnormal movements, including dystonias, are likely to diminish spontaneously [4]. In the instance of our patient, we recorded a significant enhancement throughout the four-month medication regimen.

A variety of therapeutic approaches is available, including counseling and education, oral medications, intramuscular injections of botulinum neurotoxins (BoNT), as well as physical therapy and occupational therapy, in addition to neurosurgical interventions [16]. It is important to note that none of these methods have been evaluated in the context of a double-blind randomized clinical trial [16]. The majority of the evidence supporting these treatments is derived from small controlled trials, non-blinded studies, retrospective analyses, and case reports [16-18].

In the realm of pharmacological treatments, the therapeutic classes utilized encompass anticholinergics, benzodiazepines, dopaminergics, and muscle relaxants [16]. Anticholinergics are among the most commonly prescribed medications, functioning through the inhibition of anticholinergic receptors [16]. Small-scale studies have indicated that trihexyphenidyl demonstrates a 70% efficacy at an average daily dose of 30 mg [19], regardless of the dystonia's etiology, although it may exacerbate certain dystonic conditions in children with cerebral palsy [20]. Effective dosages typically range from 6 to 40 mg per day, administered in three to four doses [16]. Common adverse effects include memory disturbances, confusion, agitation, depression, as well as symptoms such as dry mouth, constipation, urinary retention, blurred vision, and worsening of closed-angle glaucoma [16]. For our patient, we chose to administer the minimum effective dose of 6 mg without escalation, due to concerns regarding vigilance impairment.

Benzodiazepines, including alprazolam, chlordiazepoxide, clonazepam, and diazepam, function by enhancing GABA receptor transmission and have demonstrated efficacy in the management of paroxysmal dystonias [16]. For our study, we opted to administer clobazam at a dosage of 25 mg, divided into three daily doses over a month-long period. It is important to highlight that there is a lack of large-scale, double-blind, controlled studies investigating the use of benzodiazepines for dystonia [16]. Anecdotal evidence indicates that these medications may be particularly effective in alleviating phasic symptoms of dystonia, such as eye blinking in cases of blepharospasm or dominant tremors [16]. Additionally, baclofen, a GABA receptor agonist, is commonly employed in the treatment of dystonia [16, 21]. Effective oral dosages range from 30 to 120 mg per day, typically administered in three to four doses [16]. Due to its sedative properties, we have chosen a dosage of 30 mg. Anecdotal evidence suggests that this medication has yielded promising results in children experiencing dystonia associated with hypertonia,

particularly in the lower limbs [16].

Additionally, we have noted positive outcomes, although hypertonia was more pronounced in the upper limbs of our patient. Large-scale studies are essential to develop appropriate therapeutic protocols for dystonia, taking into account clinical manifestations and underlying etiology.

4. Conclusion

Late-onset dystonia is a complication that may arise following thalamic injury, particularly in cases of hemorrhage among younger individuals. A thorough semiological assessment is essential for accurate diagnosis and treatment direction. Early-stage intervention does not necessitate invasive treatment, as spontaneous resolution may occur over time. The formulation of a treatment plan must consider the potential adverse effects of the chosen therapeutic class to avoid exacerbating other associated clinical symptoms. Consequently, large-scale research is imperative to develop structured recommendations that will assist clinicians in addressing this issue.

Abbreviations

GCS	Glasgow Coma Scale
MRI	Magnetic Resonance Imaging
FLAIR	Fluid-Attenuated Inversion Recovery
ADC	Apparent Diffusion Coefficient
MIP	Maximum Intensity Projection
BoNT	Botulinum Neurotoxins
GABA	Gamma-Aminobutyric Acid

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Author Contributions

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Ethical Approval

The study was approved by our local ethics committee.

Consent

Oral informed consent was obtained from the patients for the publication of this case report.

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Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Conflicts of Interest

The authors declare no conflicts of interest.

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Research Field

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