Self-Induced Stretch Syncope: An Unusual Non-Epileptic Paroxysmal Event. A Case Report and Literature Mini-Review

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ABSTRACT: Stretch syncope (SS) is a benign, uncommon, distinct condition described mainly in adolescent males. It is responsible for paroxysmal events started by stereotyped stretching actions with neck hyperextension, culminating in alteration of consciousness. Motor manifestations are often present and may be associated with a generalized slowing of the electroencephalographic activity, challenging the diagnosis. Despite a few cases reported in the literature, different mechanisms have been implied in the pathogenesis, involving both local and systemic hemodynamic phenomena. Here, we report on an 8-year-old girl with self-induced SS, providing new insights into the related neurophysiological profile and discussing the possible etiology. Our evidence of transient and dynamic vascular impairment supports the hypothesis of SS as a multifactorial disorder.

KEYWORDS: Syncope, non-epileptic paroxysmal event, stretching maneuver, pediatrics

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Background

Stretch syncope (SS) is a rare condition observed primarily in young male adults characterized by stereotyped episodes of neck hyperextension standing or sitting, resulting in loss of consciousness.[1] The patient's clinical history is generally unremarkable, and typical pre-syncopal symptoms may not be reported.[2] SS may be easily mistaken for epileptic seizures since facial and upper limb twitching is often associated with electroencephalogram (EEG) generalized slow-wave activity, leading to an erroneous diagnosis and treatment.[3] SS also mimics non-epileptic paroxysmal events, whose differential diagnosis in childhood may be particularly challenging.[4,5]

To our knowledge, only 16 cases (12 males, age range 7-26 years) have been described; a detailed mini-review is available in Supplemental Table 1. Several underlying mechanisms have been proposed without establishing a definitive etiology.

Case Presentation

An 8-year-old girl presented a 5-month history of paroxysmal episodes characterized by variable degrees of altered consciousness and postural instability, occasionally combined with facial grimaces, upper limb tremors, and falls to the ground. Stereotyped movements of head tilting and upper limb stretching always preceded the episodes. These events occurred exclusively during wakefulness and were generally brief (5-20 seconds) and followed by a prompt and complete recovery. The parents traced the onset of symptoms to a particularly stressful period. Their frequency increased rapidly over time.

There was no family history of neurologic disorders or recurrent syncope. Despite mild anxiety, she completed primary school excellently and developed adequate peer relationships.

Before our observation, the young patient had two hospital admissions. Blood chemistry and neurological and cardiovascular assessments were unremarkable except for a quadrigeminal cistern lipoma. Standard supra-aortic trunk and transcranial Doppler examination were normal.

She was admitted to our Institute on suspicion of cataplexy. She underwent a 4-day long-term video-polysomnography monitoring with EEG, bilateral electrooculography, chin, bilateral anterior tibialis electromyography, single-channel electrocardiogram (ECG), respiratory, and peripheral oxygen saturation. Daytime and nocturnal sleep features were unremarkable; a 5-naps Multiple Sleep Latency Test ruled out hypersomnia. Fifty to 100 typical episodes were recorded daily, occurring in a sitting or standing position; she could also provoke them under request. The episodes were preceded by a head tilting against the right shoulder, less frequently the left, and associated with back and neck hyperextension, variably along with breath-holding and stretching of the upper limb(s). Shortly after this stretching maneuver, she could manifest a grimacing smile or behavioral arrest with variable

loss of the postural tone: from a subtle swaying to head dropping, asymmetrical limb weakness, sagging of the trunk, or, rarely, fall to the ground during prolonged episodes (Supplemental Videos 1 and 2). The severity of consciousness impairment varied. During the episodes, EEG showed bursts of monomorphic, slow-wave activity (2-3 Hz), lasting from 3 to 12 seconds, predominant on the frontotemporal regions and mainly ipsilateral to the side of the head tilt (Figure 1a and 1b), less frequently diffuse (Figure 1c). Mild heart rate increases with low amplitude QRS complexes were documented during the stretching. Oxygen saturation and arterial pressure measurement were not detectable due to artifacts. The execution of the Valsalva maneuver (VM) and passive triggering movement were ineffective. The attacks were more frequent during medical assessments or clinical interviews. When questioned, the patient reported a compulsive urge to stretch to contrast an indefinite numbness sensation involving her chest, shoulders, and head, which would resolve after she had performed the stretching maneuver. She declared that she was not disturbed by the above symptoms; instead, she seemed to seek them. To clarify the clinical picture, further sonographic studies were performed. Extracranial and transcranial color-coded duplex sonography (ECCS and TCCS) was normal at rest and in a neutral head position. Then, different combinations of head movements (lateral rotation, hyperextension, and flexion) and maneuvers (Adson's maneuver, VM, and compression test) were explored while lying down and or sitting. The TCCS revealed a reversal of flow and holosystolic pattern in the V4 segment of the right vertebral artery (VA), with the head turned to the left at a 45° declination angle both in a lying and sitting position. Concomitantly, the girl started complaining of mild pre-syncopal symptoms. A specular but milder pattern was observed on the left VA while the head was turned to the right. Back to a neutral position, a normalization of flow velocity and direction in the VA occurred after 1 to 2 minutes and was followed by a gradual increase of the diastolic flow component and restoration of the anterograde flow. The middle cerebral artery (MCA) investigation showed a minor blood flow asymmetry during the dynamic tests with a decrease in its flow ipsilateral to the head movement, though only <30% compared to the contralateral artery.

At ECCS, the right dominant internal jugular vein displayed an accelerated flow and loss of spectrum with a venous phasic course. Brain magnetic resonance angiography and chest X-ray were normal.

The patient and her family were made aware of the benign and self-induced nature of the phenomena and advised to avoid triggering motor behaviors. Cognitive behavioral therapy was recommended.

At one-year follow-up, the girl was episode-free and anxiety symptoms were significantly reduced. The last attack occurred during our hospitalization; the urge to precipitate the attacks resolved after 4 months of psychological intervention.

Discussion

SS was first outlined in 6 adolescents by Pelekanos et al.,[1] who identified the mechanical compression of the VA together with a predisposition to faint and the effect of the VM as causative agents. The hypothesis of extracranial compression was further endorsed by the evidence of decreased blood flow in the posterior circulation, documented during the provocative position in 2 patients.[6] A similar finding was observed in another adolescent, whereby the combination of a particular neck anatomical background in addition to the effect of a Valsalva-like maneuver were implied.[7] Again, in the case reports of 2 young girls, the authors attributed SS to a decreased blood flow due to vertebrobasilar insufficiency/extrinsic compression of the VA, even if they did not document it.[8,9] Interestingly, a drop in the systemic blood pressure followed by a diffuse cerebral dysfunction was recorded during the stretching maneuver in 4 other patients diagnosed with SS, suggesting a form of vasodepressor faint rather than a vertebrobasilar insufficiency.[2,10] Adrenergic dysfunction during VM was also proposed as an additional factor.[11] Most recently, in the case of a 7-year-old girl, the dynamic compression of the VAs was correlated to an inadequate cardiovascular response to a "false signal" from the afferent pathways, inducing reflex syncope and leading to transient hypoxia of the brainstem.[3]

Here, we report a case of self-induced SS in an 8-year-old girl, presenting for the first time evidence of a lateralized EEG pattern during the episodes in addition to the typical generalized one, and provide a literature mini-review (Supplemental Table 1).

Our patient's TCCS examination displayed blood flow changes in the posterior circulation during neck rotation, pointing to a possible vascular conflict in the VA district. We excluded static vascular compressions or malformations, abnormalities of the posterior fossa/craniovertebral junction, supernumerary ribs, and vascular conflicts at supra-aortic trunk emergence and course. However, it is well known that the VAs may be susceptible to mechanical compression or stretching in the atlantoaxial region during head rotation toward the contralateral side (particularly during childhood), compromising the vessel's blood flow distally to the point of distortion and possibly leading to vertebrobasilar distribution symptoms.[12,13]

Nevertheless, it should be emphasized that this ultrasound finding of impairment in the posterior circulation was inconsistent with our patient's neurophysiological data. The frontotemporal alterations recurrently observed at the EEG during the episodes (Figure 1a and 1b) were ipsilateral to the stretching side and rather pointed to the MCA territory, the involvement of which was not confirmed at the ultrasound. EEG focal slow-wave activity generally indicates a focal cerebral pathology of the underlying brain region, likely due to brain structural abnormalities or comprised blood flow.[14] In this regard, stretching may also impair the ipsilateral blood

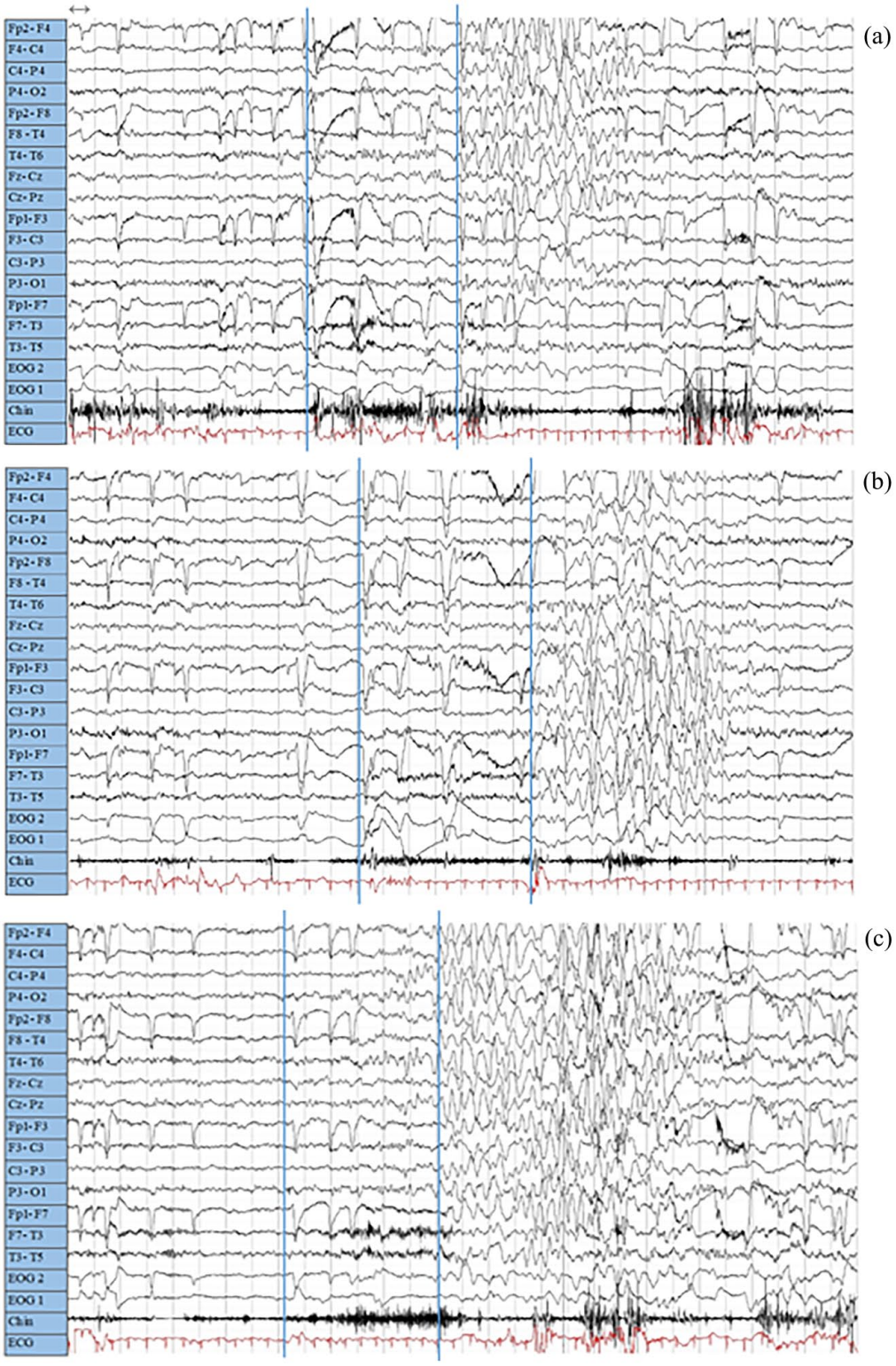

Figure 1. (a) The patient is standing and tilts her head on the right side (first mark) while hyperextending her back and neck. At the end of the stretching action (second mark), high amplitude slow-wave 2.5 Hz activity appears in the right hemisphere, lasting 6 to 7 seconds and mainly involving the frontotemporal areas. The ECG shows mild sinus tachycardia. (b) The patient is seated and tilts her head on the left side (first mark) while hyperextending her back and neck and stretching backward the ipsilateral arm. At the end of the stretching action (second mark), high amplitude 2 Hz slow-wave activity appears in the left hemisphere, lasting 7 to 8 seconds, mainly involving the frontotemporal areas and spreading to the contralateral homologous regions. The ECG shows mild sinus tachycardia. (c) The patient is standing and tilts her head on the right side (first mark) while hyperextending her back and neck backwards. Concomitantly to the end of the stretching action (second mark), the EEG trace shows a generalized high amplitude 2.5 Hz slow-wave activity lasting 9 to 10 seconds, more evident in the right frontotemporal regions.

drainage from the sinus transverse to the jugular vein, indirectly leading to transient interruption of the arterial blood flow at a microcirculatory level.[15,16] Conversely, diffuse slowing is more often associated with transient cerebral hypoperfusion linked to reduced systemic blood pressure. Similarly to vaso-vagal syncope,[17] her EEG (Figure 1c) on other occasions shows an initial generalized slowing of the cerebral activity while blood pressure drops, followed by a diffuse flattening with a variable time delay during the asystolic phase, documenting the global impairment of cerebral circulation.

The presence of both focal and diffuse EEG slowing suggests the combination of a transient comprise of the local blood flow supplied by the MCA and cerebral hypoperfusion due to a drop in the blood pressure as probable etiopathological players.

A dynamic computed tomography angiography and cerebral near-infrared spectroscopy measurement could have been helpful in further clarifying the clinical picture. However, they were not performed given the benign condition and young age of the patient.

Conclusion

SS is a non-epileptic paroxysmal disorder that presents with a spectrum of clinical symptoms involving variable impairment of consciousness and loss of muscular tone, culminating in syncopal episodes. According to the literature, this condition is rare and predominant in childhood; its genesis is still unclear and likely multifactorial.

The evidence in our case appears challenging to interpret. Sonographic findings suggestive of posterior circulatory alterations during lateralized head movements coincided with heterogeneous neurophysiological data of local anterior involvement in most of the episodes (probably mediated by an altered venous outflow), diffuse in the others (probably on a systemic basis of hypoperfusion/syncope). The contribution of multiple etiological mechanisms seems plausible in explaining the different phenomena we observed.

Well-conducted medical history, including affective and relational background, neurovascular assessment, and long-term complete video-EEG monitoring (including electromyography and cardio-respiratory parameters) are essential in the differential diagnosis of unexplained paroxysmal episodes in pediatric age. Psychopathological aspects should be investigated, as they may influence onset and recurrence.

Declarations

Ethical approval and consent to participate

The study was approved by our local ethics committee (Comitato Etico Interaziendale Bologna-Imola, CE-BI, protocol number 17009).

Consent for publication

The participant signed consent regarding publishing their data.

Author contributions

Anna Mercante: Conceptualization; Writing—original draft; Data curation; Methodology. Fabio Pizza: Conceptualization; Writing—review & editing; Supervision; Methodology. Federica Pondrelli: Writing—review & editing. Andrea Zini: Investigation; Writing—review & editing. Luigi Cirillo: Writing—review & editing; Investigation. Paolo Tinuper: Writing—review & editing. Rocco Liguori: Writing—review & editing. Ludovica Migliaccio: Data curation; Investigation. Stefano Vandi: Data curation; Investigation. Giuseppe Gobbi: Writing—review & editing. Giuseppe Plazzi: Conceptualization; Writing—review & editing; Supervision; Methodology.

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Competing interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Availability of data and materials

The authors confirm that the data supporting the findings of this study are available within the article and its supplementary materials. Comprehensi data are available from the corresponding author (G.P.) upon reasonable request.

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Supplemental material

Supplemental material for this article is available online.

Disclosure of informed consent

We obtained written and verbal consent from the subject and her legally authorized representative (biological parent).

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