

Case study

Neurovegetative dysautonomia revealed by blindness in the cold

Abstract:

The autonomic nervous system (ANS) is responsible for regulating heart rate and blood pressure and maintaining homeostasis during physiological stresses. Nervous dysautonomias are often observed in patients presenting cardiovascular symptoms. Despite that, other important but less known conditions can be encountered. We report the case of a 19-year-old young who presented for blindness occurring in the cold and improving with exercise. Investigations revealed neurovegetative dysautonomia with significant vagal hyperactivity associated with central B-sympathetic hyperactivity. Our patient received treatment based on hygienic and dietary measures with a combination of an anti-hypotensive drug such as Etilefrine. 2 months later, a re-evaluation showed a clear improvement, confirmed during the control neurovegetative exploration but also during the ophthalmological examination.

Comment [HSY1]: ...youngperson

Keywords:

Autonomic nervous system, nervous dysautonomia, blindness, vagal hyperactivity, sympathetic hyperactivity.

Introduction:

The autonomic nervous system (ANS) is responsible for regulating heart rate and blood pressure and maintaining homeostasis during physiological stresses. Nervous dysautonomias are often observed in patients presenting cardiovascular symptoms. In addition to the most common, high blood pressure, other important but less known conditions can be encountered. Our case presented in this article is a typical example of the rare manifestations revealing neurovegetative dysautonomia.

Case presentation:

Mr. Y.H. a 19-year-old young man without cardiovascular risk factors, has had monophthalmus of the left eye since birth. The patient presented to our ANS exploration unit for blindness occurring in the cold and improving with exercise, evolving for 5 months before his consultation without other associated signs. Ophthalmological exploration found a drop in visual acuity rated 7/10, while the anterior segment, electroretinography, and fundus examinations did not reveal any abnormalities. The patient also received a neurological examination without abnormalities, and a brain MRI returned normal. Furthermore, a basic cardiovascular examination coupled with a transthoracic ultrasound was also without abnormalities.

Comment [HSY2]: The presentation was not included the manifestation in other systems e.g. the gastrointestinal system, genitourinary system, sweating system (hypo-anhidrosis), and sleep disturbance.

Given the clinical orientation, in particular the improvement of symptoms during exercise, neurovegetative dysautonomia was suspected. We conducted a clinical exploration of ANS, carrying out several tests.

At rest, our patient had a minimum heart rate (HR) of 48 beats per minute (bpm) and a maximum rate of 60 bpm (Figure 1), with a systolic blood pressure (SBP) of 120 mmHg.



Figure 1: Resting ECG

During the deep breathing test, we noted a 33% vagal response (HR max at 100 bpm; decrease in SBP from 120 to 106 mmHg). The hand grip test demonstrated a vagal response identical to that of the previous test, while the sympathetic response did not exceed 24% (Figure 2).



Figure 2: hand grip test ECG

The third test consisted of hyperventilation, during which the rate increased from 74 bpm to 94 bpm with a drop in SBP from 120 mmHg to 94 mmHg (Figure 3).



Figure 3: hyperventilation test ECG

As for the mental stress test, the B-sympathetic response was frankly high, with a percentage reaching 39%; however, the α -sympathetic response was limited to 12%.

We finished our exploration of ANS with the orthostatic test, which also revealed a very significant vagal response, reaching 60% (Figure 4).





Figure 4: orthostatic test ECG

At the end of our exploration, we concluded that there was significant vagal hyperactivity associated with central B-sympathetic hyperactivity, which could be consistent with the clinical presentation that our patient presented.

A treatment based mainly on hygienic and dietary measures was initiated: avoidance of triggering factors, wearing compression stockings, and rehydration (>1.5 l/d), a high salt diet (>6 g of sodium/d), and also the combination of an anti-hypotensive such as Etilerfrine.

A re-evaluation was carried out after 2 months, showing a clear improvement, confirmed during the control exploration but also during the ophthalmological examination (figures 5-7).

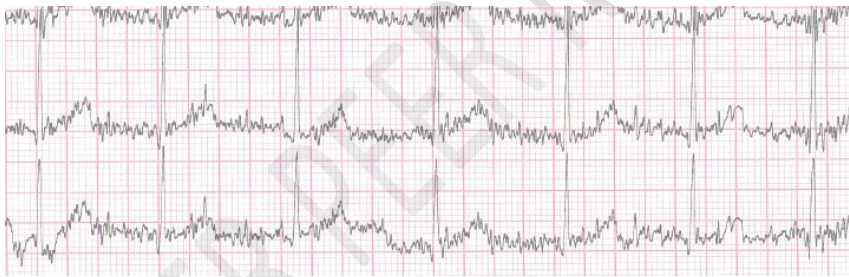


Figure 5: hand grip test, control ECG



Figure 6: hyperventilation test, control ECG

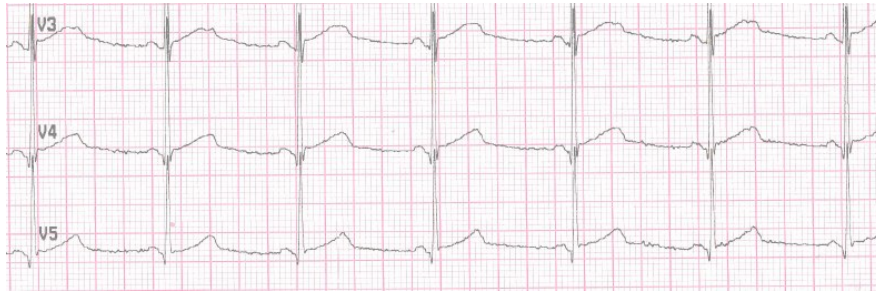


Figure 7: orthostatic test, control ECG

Discussion:

The autonomic nervous system (ANS) is made up of sympathetic, parasympathetic, and enteric components. The cardiovascular branch of the ANS is responsible for regulating heart rate and blood pressure and maintaining homeostasis during physiological stresses, including exercise and standing. The two crucial elements controlled by the ANS are the heart and the vessels (1–3).

Generally, autonomic dysfunctions are of multifactorial origin, accompanying diseases with central or peripheral involvement of the ANS and/or treatment with neurotoxic or cardiotoxic drugs. As a primary disease, it can be found in rare cases of primary autonomic failure. The disruption of autonomic nervous regulation in any reflex chain manifests itself in the corresponding organic system (4).

Autonomic symptoms can be divided into orthostatic, non-orthostatic, and diffuse. Orthostatic symptoms are generally associated with cerebral hypoperfusion. Common non-orthostatic symptoms include constipation, bladder problems, intolerance to cold or heat, excessive sweating or loss of sweat, and erectile dysfunction. Common complaints also include fatigue, headaches, and insomnia (2).

The process of cardiac dysautonomia can be paroxysmal or chronic. Disorders of the sympathetic or parasympathetic systems can result in one action without opposition from the other. The most common cardiac manifestations of dysautonomia include reflex syncope, inappropriate sinus tachycardia, and orthostatic intolerance syndromes: orthostatic hypotension and postural orthostatic tachycardia syndrome (1).

Clinically, parasympathetic function is tested by measuring changes in heart rate in response to deep breathing, the Valsalva maneuver, and active standing. Sympathetic function is typically tested by measuring changes in blood pressure in response to the Valsalva maneuver, tilt table test, and active standing (5).

There are many additional tests of autonomic function that occur in other organ systems, such as urodynamic testing in urology, gastric emptying studies in gastroenterology, and pupillometry in ophthalmology (5).

The rare clinical presentation of our patient with a drop in visual acuity occurring during cold weather may be linked to orthostatic dysautonomia secondary to cerebral hypoperfusion. More likely an orthostatic cerebral hypoperfusion syndrome without orthostatic hypotension, bradycardia, hypocapnia, and excessive tachycardia (2).

In general, the therapeutic approach to orthostatic cerebral hypoperfusion syndrome is the use of calcium channel blockers or angiotensin-converting enzyme inhibitors for patients suffering from hypertension; and volume expansion with salt, fluids, Fludrocortisone, or the use of vasopressor medications in patients with hypotension (2).

The progression of autonomic dysfunction is associated with increased cardiovascular risk. Thus, regular screening and management of the risks of progression of neurovegetative dysautonomia are necessary. Clinical exploration of ANS is recommended once every two years. However, additional studies are needed to clarify the precise mechanisms underlying the association between the progression of autonomic dysautonomia and cardiovascular diseases (6).

Cardiologists are not always aware of cardiac dysautonomia because they typically focus on primary cardiac disorders such as heart failure, coronary artery disease, arrhythmias, and hypertension. However, being alert to the possibility of cardiac dysautonomia is very important, especially when dealing with patients with unusual symptoms and apparently normal vital parameters and ECGs.

Conclusion:

Besides the most common arterial hypertension, conditions such as postural orthostatic tachycardia syndrome, orthostatic hypotension, reflex syncope, and chronotropic insufficiency should be known by cardiologists. Cardiac dysautonomias vary in prevalence across the lifespan and may overlap, thus posing a challenge for a clinician. It is therefore important to recognize their manifestations, the diagnostic methods, and the prognostic and therapeutic consequences.

References:

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Comment [HSY4]: 2019 April

Comment [HSY5]: (4) :204-214

Comment [HSY6]: Yun, JS., Park, YM., Cha, SA. et al. Progression of cardiovascular autonomic neuropathy and cardiovascular disease in type 2 diabetes. *CardiovascDiabetol* 17, 109 (2018).