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Paraneoplastic autoimmune autonomic ganglionopathy as a debut of a bladder urothelial carcinoma: A case report

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Background and aims

Autoimmune autonomic ganglionopathy (AAG) is a rare immune-mediated disorder associated with anti-α3 subunit of the ganglionic-type nicotinic acetylcholine receptor (anti-gAChRα3) antibodies, which bind to the acetylcholine-receptor in autonomic ganglia (parasympathetic and sympathetic), leading to autonomic failure. This disorder is mostly associated with viral infections, but it can also be associated with systemic malignancies. Here, we report the first case of a paraneoplastic autonomic ganglionopathy as a debut of a bladder urothelial carcinoma.

Methods

Case report.

Results

A 44-year-old man, without medical history of interest, went to the emergency department because of blurry vision, eye and mouth dryness, constipation and orthostatic hypotension, for the last 2 weeks. A drop in blood pressure (BP) upon standing was documented [100/60 mmHg (decubitus) vs. 80/50 mmHg (sitting)]. Blood tests, chest x-ray, brain-MRI, cerebrospinal fluid and electroneuronography were unremarkable. Electrochemical skin conductances were decreased in the feet. Serological examinations were positive for anti-gAChRα3 antibodies. A full-body computed tomography showed a bladder tumor, which was surgical treated by transurethral bladder resection. The pathology showed a low grade non muscle invasive bladder urothelial papillary carcinoma. After tumor resection and treatment with intravenous immunoglobulins and corticosteroids, the patient gradually improved.

Conclusions

Subacute panautonomic failure can be the debut of systemic malignancies. As far as we know, this is the first case report showing a paraneoplastic autonomic ganglionopathy as debut of bladder urothelial carcinoma. This case highlights the importance of a systemic study to rule out the presence of cancer when AAG is present.

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Infectious lesions of the insula associated with extreme sinus bradycardia: Report of three cases

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Background and aims

The relationship between ischemic lesions in the insula and alterations of the heart rhythm due to autonomic nervous system dysfunction is known. However, the association with infectious (viral or bacterial) lesions has been poorly reported in the literature. Here, we described 3 cases of sinus bradycardia associated with infectious lesion of the insula.

Methods

Case series.

Results

Case 1: A 36-year-old man went to the emergency department because of 6 repetitive transient-losses-of-consciousness on the same day along with headache. During a new syncope, a sinus bradycardia at 35 bpm with asystole (> 5 s) was documented, and a transitional pacemaker was implanted. Due to fever, a lumbar puncture was performed, showing herpes-simplex-virus-DNA. MRI showed linear uptake in the right insula. After acyclovir treatment, the electrocardiographic alterations disappeared. Case 2: A 54 year-old man went to the emergency department because of headache and fever and a sinus bradycardia of 40 bpm was documented. A lumbar puncture showed herpes-simplex-virus-DNA. MRI showed linear uptake in the right insula. After acyclovir and foscarnet treatment, the heart rate returned to normal. Case 3: A 54 year-old woman, diagnosed with acute pneumococcal meningitis caused by bilateral otitis media, developed a bradycardia of 33 bpm, without any other alteration in ECG. Echocardiogram was normal. A brain MRI showed purulent material in the context of cerebritis at right insular level. After antibiotic treatment, the bradycardia disappeared.

Conclusions

Clinicians should be aware that viral or bacterial lesions in the insula could lead to cardiac arrhythmia due to central autonomic nervous system dysfunction.

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Relationships between resting and reflexive measures of heart rate variability (HRV)

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Background and aims

Resting HRV is an important diagnostic and prognostic tool in the evaluation of autonomic dysfunction. Resting and reflexive HRV are markers of the autonomic nervous system, however, their relationship has largely been described only in healthy and diabetic patients. This relationship has not been explored in patients with neurologic disease or in medically complex patients. This study aims to investigate the demographic, medical, neurologic, and autonomic correlates of resting HRV in order to further understand factors that may be considered potential confounders in studies of resting HRV.

Methods

In this retrospective study of a diverse, urban population (50.2% female, 45% HIV+, median age 50 (IQR 41,57)), results from autonomic function testing (AFT) of 209 patients were analyzed.

Results

Moderate correlation was found between both root-mean square of differences of successive RR intervals (rMSSD) and percent of differences of adjacent RR intervals >50 ms (pNN50) and heart rate during deep breathing (HRDB) ($r_s = 0.53$, $p < 0.0005$), Valsalva ratio (VR) ($r_s = 0.45$, $p < 0.0005$), and age ($r_s = -0.25$, $p < 0.0005$). No correlation was found between pNN50/rMSSD and the motor portion of the Unified Parkinson's Disease Rating Scale (m-UPDRS). pNN50 was significantly different in patients with and without diabetes mellitus (DM), hypertension (HTN), distal sensory polyneuropathy (DSP), chronic obstructive pulmonary disease (COPD), and kidney disease ($p < 0.05$).

Conclusions

Resting HRV is positively correlated with reflexive HRV and negatively correlated with age; DM, HTN, DSP, COPD and kidney disease are associated with less heart rate variability and are potential confounders in studies of resting HRV.

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Therapeutic effect of OnabotulinumtoxinA in harlequin syndrome and episodic migraine: A case report

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Background and aims

Objective: To describe a case of Harlequin syndrome in a Movement Disorder Clinic in El Salvador. Background: Harlequin syndrome is a rare dysautonomic disorder characterized by asymmetric facial sweating and flushing in response to heat and exercise.

Methods

We performed a clinical evaluation and open application of OnabotulinumtoxinA with scheme for the treatment of Harlequin syndrome by a Movement Disorders specialist.

Results

A 47-year-old female with a 3-year history of episodes of profuse sweating and flushing on the right side of the face triggered by heat, sun exposure and exercise and episodic migraine since childhood (5 episodes per month), with chronic use of over-the-counter medications. Brain MRI was normal. Four cycles of 100 units of OnabotulinumtoxinA were applied to the right hemi face, presenting an 80% improvement in hyperhidrosis and a 90% improvement in migraine episodes and significant reduction in pain medication use. Onset of therapeutic effect was at 7 days, and duration of the maximum therapeutic effect was at 5 months.

Conclusions

In our patient, OnabotulinumtoxinA was effective in improving unilateral flushing and facial hyperhidrosis and showed a preventive therapeutic effect in episodic migraine.

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High through put method to clean, and iteratively filter raw ECG data using open source software

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Background and aims

Heart Rate Variability (HRV) provides a non-invasive quantification of autonomic nervous system function[1]. Studies of HRV often involve large data sets and preparing the raw ECG data for analysis is labor intensive[2]. Proprietary software exists to mitigate this, [1] however open-source software packages still depend on manual edits and filtering. We developed a package in R to speed this process. [1] Juha Pekka Niskanen and others, 'Software for Advanced HRV Analysis', Computer Methods and Programs in Biomedicine, 76.1 (2004), 73–81 [2] Hussein Al Osman, Mohamad Eid, and Abdulmotaleb El Saddik, 'A Pattern-Based Windowed Impulse Rejection Filter for Nonpathological HRV Artifacts Correction', IEEE Transactions on Instrumentation and Measurement, 64.7 (2015), 1944–57.

Methods

Using window filters, an iterative process was used to filter each heart rate (HR). Likely artifactual HRs are removed ($40 > HR > 180$ & $HR > 10\%$ mean previous 50 beats). The Hampel filter removes HRs > 4 median absolute deviations (MAD) in a 5-beat window. Smoothing splines were used to interpolation for time analysis measures. We developed and tested the package using patient HRV data ($n = 334$).

Results

Following processing with the package <3% of the HR data warranted any manual edits. RMSSD was reduced by ~38% on average and pNN50 by ~9% after filtering. Efficient use of iterative filtering was approximately 6 iterations.

Conclusions

The open-source "tidyrhrv" package helps to automate the calculation of HRV markers from raw ECG, a process which is otherwise expensive and/or time consuming for large datasets.

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Cardiology meets neurology: Clinical presentation and management of patients with primary neurogenic disorders and orthostatic intolerance

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Background and aims

Patients with primary neurogenic disorders such as pure autonomic failure or multiple system atrophy may present first with cardiocirculatory symptoms such as orthostatic intolerance or fluctuations in heart rate with symptomatic tachycardia. It is therefore clinically important to identify such patients since circulatory manifestations are only one of a series of symptoms resulting from autonomic dysfunction in various organ systems. These patients need a multimodal diagnostic and therapeutic approach and should undergo extensive evaluation in a specialized autonomic nervous system (ANS) outpatient unit.