ELECTROPHYSIOLOGY
Case Report
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Atrioventricular Nodal Reentrant Tachycardia in Kartagener's Syndrome

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ABSTRACT

Single case report of a middle-aged patient with Kartagener's syndrome who experienced recurrent paroxysmal supraventricular tachycardia. Despite optimized beta-blocker therapy, only cardioversion in emergency departments provided symptomatic relief. The unique electrocardiographic features of this condition and their significance in transaortic electrophysiological studies for diagnosis and treatment are highlighted.

KEYWORDS: Tachycardia, supraventricular; Catheter ablation; Kartagener Syndrome; Radiofrequency ablation; Electrophysiology.

INTRODUCTION

Kartagener's syndrome, a subset of primary ciliary dyskinesia, affects approximately[1] in 20,000 to 40,000 individuals[1]. It presents a distinctive triad of situs inversus, chronic sinusitis, and bronchiectasis. Arrhythmias are uncommon in this context, and the added complexity of situs inversus totalis challenges diagnosis and treatment[2,3,4]. In this case report, the utility, and complexities of electrophysiologic study to establish the diagnosis and guide radiofrequency ablation of atrioventricular nodal reentrant tachycardia in Kartagener's syndrome are presented.

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CASE REPORT

History of Presentation

A 40-year-old man was referred for evaluation and treatment of recurrent paroxysmal supraventricular tachycardia. He often visits emergency departments for these episodes, usually requiring cardioversion for resolution. Optimized oral beta-blocker therapy has been ineffective in managing his condition.

Physical Examination

Physical examination was unremarkable.

Past Medical History

Having been previously diagnosed with Kartagener's syndrome, he experienced chronic debilitation due to frequent episodes of bacterial sinusitis. He had no other comorbidities. No other family members with suspected or confirmed Kartagener's syndrome was identified.

Investigations

The patient arrived with a 12-lead electrocardiogram (ECG) taken during previous symptoms, following standard lead placements. It showed a regular, narrow complex tachycardia with a short RP-interval, suggestive of nodal reentrant tachycardia (Fig. 1). To enhance anatomical clarity, a non-contrast chest computed tomography (CT) and further ECGs were obtained, including additional right-sided lead positions (Fig. 2). The CT scan confirmed complete thoracoabdominal situs inversus, and thickened, dilated, and irregular bronchial walls, and ruled out other cardiac or respiratory abnormalities.

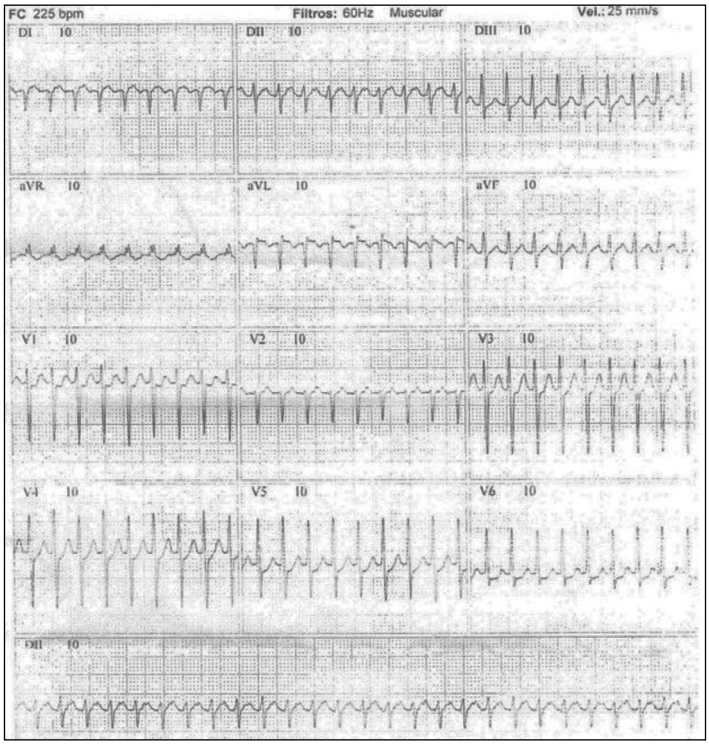

Figure 1. Surface electrocardiogram during a palpitation crisis: A supraventricular tachycardia with an undetectable retrograde P wave at 225 beats per minute was recorded.
Source: Elaborated by the authors.

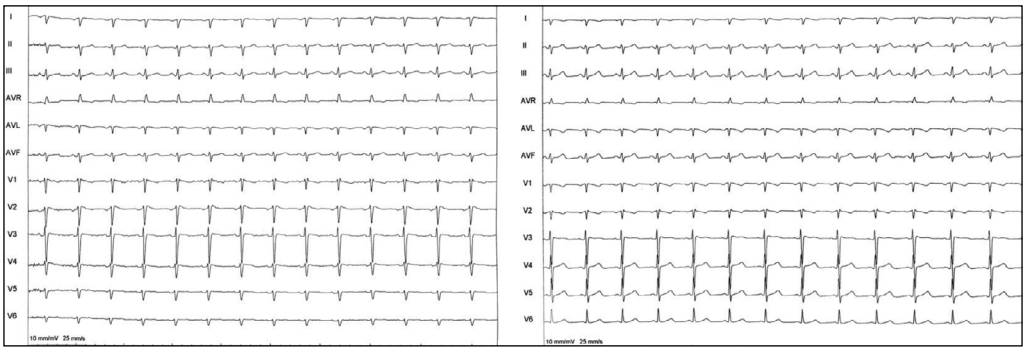

Figure 2. Resting electrocardiogram: Left recording (Conventional/left thoracic electrode positioning) - V1 electrode positioned on the fourth intercostal space to the right of the sternum, V2 on the fourth intercostal space to the left of the sternum, V3 between leads V2 and V4, V4 on the fifth intercostal space at the midclavicular line, V5 at the level of V4 at the anterior axillary line, V6 at the level of V5 at the midaxillary line. Right recording (right thoracic electrode positioning - V1R to V6R) electrodes positioned in a mirror-like fashion as described in the left thoracic electrode positioning. In Situs Inversus, the heart is positioned in the right thorax, and when electrodes are placed on the left, surface ECG will reveal a negative P wave in DI and aVL, a negative P wave in aVR, and an inferior axis in the frontal leads. These changes can be explained by the depolarization direction of the atrial resulting vector in Situs Inversus from left to right as opposed to expected in Situs Solitus. A reversed R wave progression from V1 to V6 with a decreasing amplitude, typically of Situs Inversus, is also shown in the left recording. All these changes are normalized when electrodes are reversed (right recording).
Source: Elaborated by the authors.

Under conscious sedation and local anesthesia, the patient underwent an electrophysiology study. The following catheters were percutaneously inserted through the right femoral veins and advanced into the right heart chambers: (1) a 6 Fr quadripolar diagnostic catheter was placed through the tricuspid valve, slightly rotated counterclockwise, within the interatrial septum to capture the His bundle electrogram; (2) a 7 Fr decapolar diagnostic catheter was guided through the tricuspid valve into the right ventricle, using a 30°-left anterior oblique view under fluoroscopy. It was then retracted into the right atrium while maintaining a slight counterclockwise rotation, allowing access to the coronary sinus ostium; (3) a 4 mm therapeutic catheter. Additionally, an intracardiac echocardiogram probe was positioned in the right atrium through an 11 Fr left inguinal puncture, confirming the proper positioning of the catheters (Fig. 3).

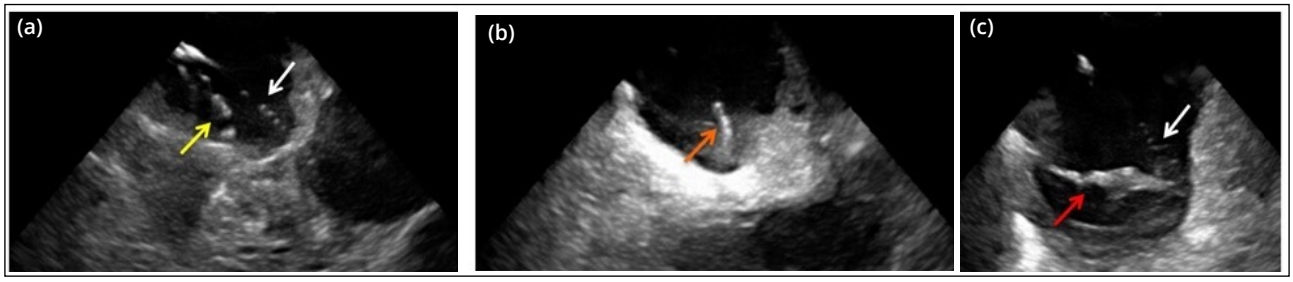

Figure 3. Intracardiac echocardiogram (ICE) guided catheter positioning: (a) ICE imaging showing a diagnostic decapolar catheter positioned in the coronary sinus (CS- yellow arrow) and a 4mm catheter ablation in the mapping zone. (b) ICE view of catheter ablation in the point of the Triangle of Koch where the slow pathway was ablated. (c) A more posterior ICE view shows the relation between the decapolar catheter in the CS (white arrow) and the tricuspid valve (red arrow).
Source: Elaborated by the authors.

First, normal intracardiac conduction intervals were identified (intra-atrial, 25 ms; atrial-His, 85 ms; His-ventricular, 42 ms). Second, decremental valvular pacing unveiled a 1:1 ventriculoatrial conduction, sustained until pacing frequency was lowered to 360 ms. Third, using a coronary sinus bipole, decremental atrial stimulation exposed an anterograde Wenckebach point at 310 ms. Fourth, programmed atrial stimulation did not reveal "dual physiology," but instead, it consistently induced regular tachycardia with short RP intervals (Fig. 4). During tachycardia, the following intervals were noted: cycle length, 330 ms; atrial-His, 190 ms; His-atrial, 140 ms; and septal ventriculoatrial, 90 ms. Fifth, tachycardia entrainment through right ventricular pacing resulted in a corrected post-pacing interval of 110 ms, consistent with nodal reentrant tachycardia (Fig. 5). Lastly, apex-base and para-Hissian pacing maneuvers were conducted, confirming the absence of a septal accessory pathway (Figs. 6 and 7).

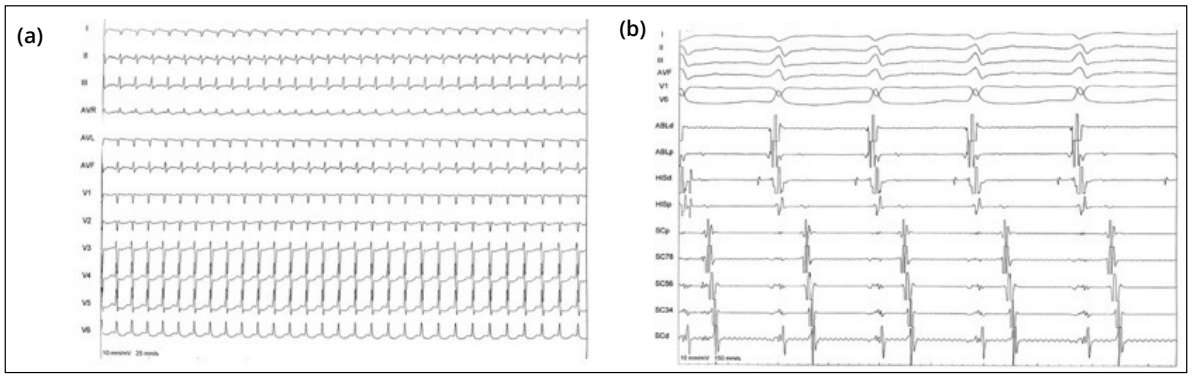

Figure 4. Tachycardia characteristics: (a) Surface electrocardiogram of the narrow and regular QRS supraventricular tachycardia (SVT). (b) Intracardiac electrograms showing a short RP (RP<PR) SVP with a concentric atrial depolarization and the following intervals: cycle length: 330 ms; atrial-His: 190 ms; His-atrial: 140 ms; and septal ventriculoatrial: 90 ms. An ablation catheter was positioned in the right ventricle (ABL), a decapolar catheter was positioned in the coronary sinus (from proximal to distal: SCp —> SC78 —> SC56 —> SC34 —> SCd), and a quadripolar catheter in the bundle of His (HIS).
Source: Elaborated by the authors.

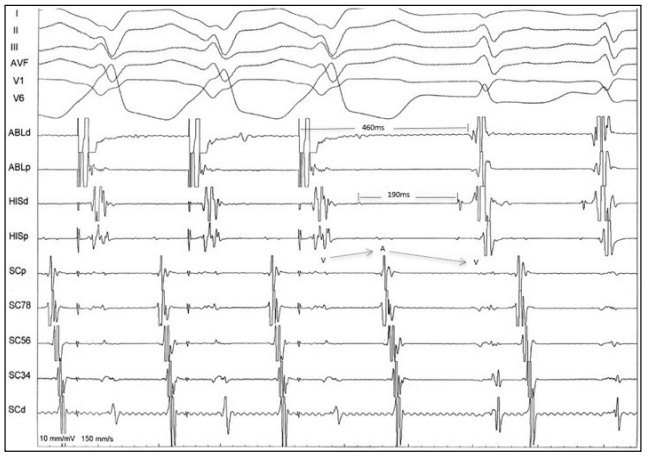

Figure 5. Ventricular entrainment of supraventricular tachycardia: The exit of ventricular entrainment by the distal pole of the catheter ablation (ABLd) with a ventricle-atrium-ventricle (V-A-V) response is shown with a PPI of 460ms and a corrected PPI – TCL of 110ms. An ablation catheter was positioned in the right ventricle (ABL), a decapolar catheter was positioned in the coronary sinus (from proximal to distal: SCp —> SC78 —> SC56 —> SC34 —> SCd), and a quadripolar catheter in the bundle of His (HIS). (PPI: post pacing interval, TCL: tachycardia cycle length).
Source: Elaborated by the authors.

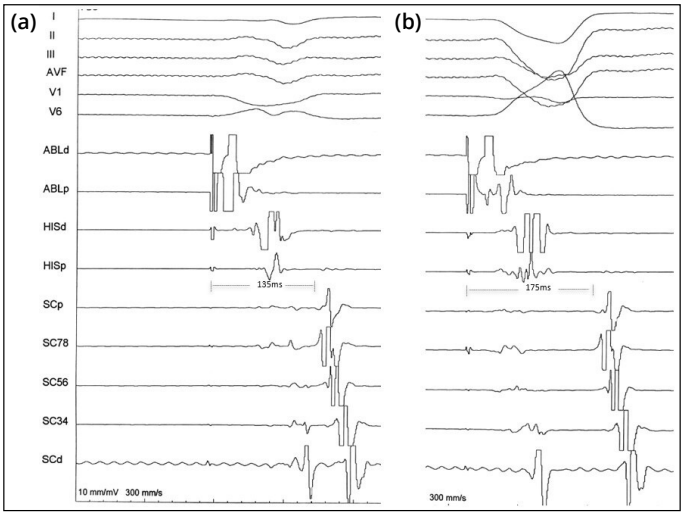

Figure 6. Apex-Base pacing maneuver: The catheter ablation was paced by its distal dipole (ABLd) in the right ventricle apex and base, and the intervals from the stimulation artifacts to the atrium were respectively measured (a) 135ms and (b) 175ms. These results pointed against a concealed septal accessory pathway. A decapolar catheter was positioned in the coronary sinus (from proximal to distal: SCp —> SC78 —> SC56 —> SC34 —> SCd) and a quadripolar catheter in the bundle of His (HIS).
Source: Elaborated by the authors.

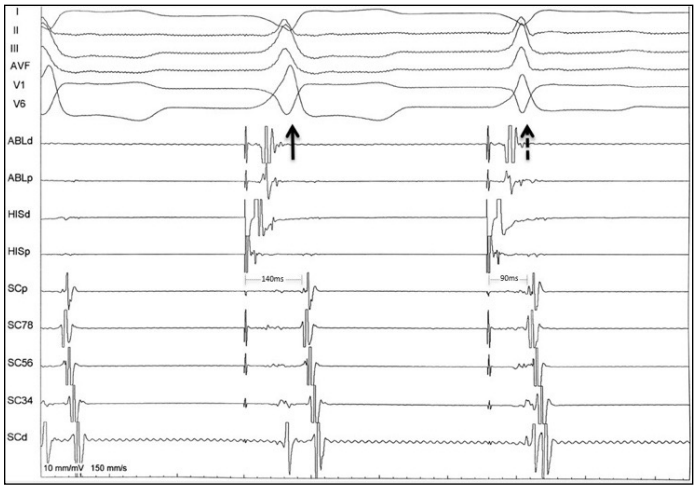

Figure 7. Para-hissian pacing maneuver: The catheter ablation was positioned in the His bundle and paced by its distal dipole (ABLd) with different output intensity until His bundle and muscle captures were observed (straight and dashed arrows respectively), and the stimulus artifact to the atrium was measured (140ms and 90ms, respectively). This response was consistent with the absence of a concealed septal accessory pathway.
Source: Elaborated by the authors.

Management

Guided by both echocardiogram and 30°-right anterior oblique fluoroscopy view, the slow pathway in the triangle of Koch was identified and ablated. A radiofrequency application at 35 Watts was enough to initiate a junctional rhythm, indicating effective disruption of the slow pathway. Following the ablation procedure, further electrophysiological testing was done, and tachycardia was no longer induced.

Follow-up

Over the in-hospital follow-up, no other complaints of palpitations, dizziness, lightheadedness, or dyspnea were presented.

DISCUSSION

Primary ciliary dyskinesia (PCD) is characterized by an inherent ciliary aberration, resulting in ciliary immotility, dysmotility, or outright absence. Diagnostic definitiveness remains elusive, notwithstanding cardinal clinical hallmarks including neonatal respiratory distress antecedents, persistent early-onset cough, chronic nasal congestion and rhinorrhea, recurrent chronic otitis media, and lateralization anomalies (such as situs inversus or ambiguities), all of which strongly indicate PCD. In adult males, observation of dyskinetic spermatozoa or respiratory compromise due to impaired mucociliary clearance should elicit suspicion. Kartagener's syndrome, a PCD subset, manifests as a triad encompassing situs inversus, persistent sinusitis, and bronchiectasis[1,5].

Pertinent diagnostic nuances warrant elucidation. First, while situs inversus contributes value, its presence is discernible in merely half of the PCD cohort, and its manifestation does not uniformly signify PCD. Second, bronchiectasis, though it may ensue in younger PCD individuals, does not manifest at birth. Consequently, the complete Kartagener's triad may manifest belatedly, challenging proper diagnosis and management. Third, virtually all afflicted individuals experience rhinosinusitis, often accompanied by nasal polyps, and in cases of absent frontal sinus, a nasal timbre ensues. Chronic sinusitis predominantly implicates certain sinuses, with frontal and sphenoid sinuses underdeveloped, a feature evident in sinus CT scans routinely conducted for refractory nasal symptoms[1].

PCD patients typically exhibit outcomes akin to the general population. However, congenital anomalies, notably cardiac malformations, pyloric stenosis, and epispadias, exhibit an elevated prevalence in those patients[5]. Despite arrhythmias are rare,

clinical suspicion for PCD mandates cardiac evaluation due to heightened propensities for congenital heart disease, especially in concurrence with heterotaxy, culminating in potential sinus and atrioventricular node perturbations alongside ventricular conduction anomalies.

Atrioventricular nodal reentrant tachycardia represents the preeminent subtype of paroxysmal supraventricular tachycardia. It predominantly ensues within structurally unremarkable cardiac substrates, underpinned by dual electrical conduits to the atrioventricular node. Although precise anatomical demarcations of these conduits remain enigmatic, they intricately interplay within Koch's triangle. Advances in catheter ablation methodologies have accentuated the imperative of grasping the anatomical nuances, particularly in individuals with congenital cardiac irregularities[6]. This case emphasizes how cardiac imaging assumes a pivotal role in the armamentarium of electrophysiologists, furnishing precise anatomical insights that optimize the safety and efficacy of ablation procedures and augment comprehension of intricate anatomical configurations and optimal catheter placement strategies.

ETHICS STATEMENT

Written informed consent was obtained from the involved patient. The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate.

CONFLICT OF INTEREST

Nothing to declare.

AUTHOR CONTRIBUTIONS

Substantive scientific and intellectual contributions to the study: Oliveira LH, Luize CM; **Conception and design:** Carvalho RS, Dietrich O; **Article writing:** Nogueira A, Lago DM; **Critical revision:** Carvalho MFS, Cirenza C; **Final approval:** Oliveira LH, Luize CM.

DATA AVAILABILITY STATEMENT

The data will be available upon request.

FUNDING

Not applicable.

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