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Right atrial appendage tachycardia: A rare cause of tachycardia induced cardiomyopathy in a 4-year-old child

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ABSTRACT

We present a rare case of tachycardiomyopathy in a 4-year-old girl. The child had incessant atrial tachycardia (AT) and refractory heart failure. Right atrial appendage (RAA) was localised as the source of the ectopic tachycardia. The child underwent successful radiofrequency ablation (RFA) using 3-D electroanatomical mapping. Fluoroscopy was used sparingly only to rule out underlying anomalies. The left ventricular functions returned to normal by one month after the procedure. RAA AT is rare in very young children and usually necessitates surgical appendectomies. RFA is a challenge in such age groups and there are very few published literature on RAA AT in very young children.

1. Introduction

Pediatric arrhythmias leading to tachycardiomyopathy (TCMP) are usually incessant ectopic atrial tachycardias (ATs), persistent junctional reentrant tachycardias (PJRT) and atrial flutters [1]. Right atrial appendage (RAA) ATs constitute 0.6–8% of all ectopic atrial tachycardias [2]. These forms of ATs are usually incessant and are known to cause TCMP in older children and in adults [3]. However, it is unusual that ectopic ATs originate from the RAA in very young children and there are only anecdotal reports of the same. We describe a rare case of right atrial appendage tachycardia leading to TCMP in a 4-year-old child.

2. Case report

A 4-year-old girl from Kenya, weighing 18 kg and with a height of 104 cm, presented to us with clinical features of heart failure. The mother of the baby also complained of persistent pounding of the precordium in the baby since the last 6 months. The baby was already being treated for heart failure with diuretics, digoxin and beta blockers. Transthoracic echocardiography (Supplementary video 1) showed a dilated heart and severe left ventricular dysfunction. Obvious anatomical causes of left ventricular dysfunction were ruled out by trans-thoracic echocardiography. The 12-lead ECG showed a regular narrow QRS tachycardia with a very rapid ventricular rate of 280 beats per minute with a RP interval of 90 m s and upright P waves in the inferior leads (Fig. 1A). Adenosine, given during the tachycardia, resulted in continuation of the tachycardia with transient atrio-ventricular block (AV) block seen as rapid P waves without QRS complexes. The differential diagnoses were atrial tachycardia with 1:1 atrio-ventricular conduction and a clockwise atrial flutter. As the drugs were ineffective in controlling the arrhythmia and the heart failure, radiofrequency ablation was considered.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.ipej.2018.07.001.

The baby was electively intubated for the procedure. She was in persistent tachycardia since the start of the procedure. Three venous accesses were obtained. A quadri-polar catheter (2 mm inter-electrode spacing) was used to map the His-bundle. A deca-polar catheter (2-5-2-mm inter-electrode spacing) was placed in the coronary sinus (CS). Another deca-polar catheter (2-5-2 mm inter-

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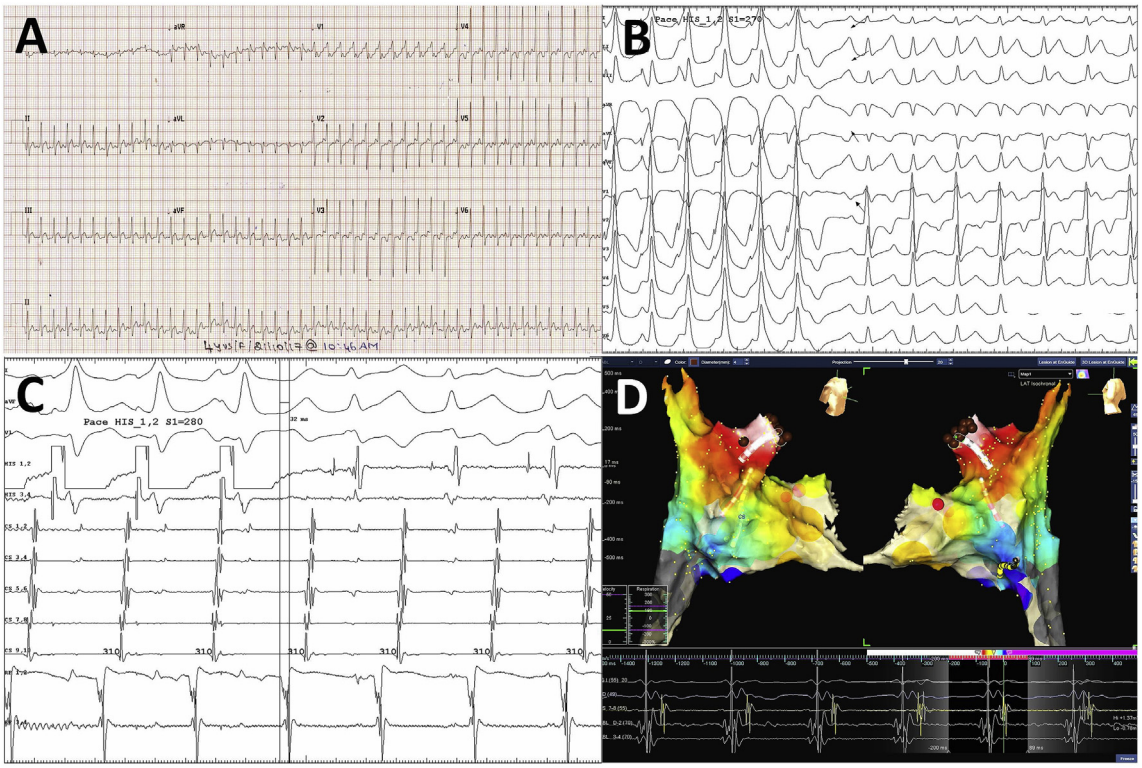

Fig. 1. **(A)** Shows a regular narrow QRS tachycardia noted in the 4-year-old child with features of heart failure; **(B)** shows the positive P waves in leads II, III (downward arrows) and negative P waves in leads avR and V1 (upward arrow), also seen is the slow P wave progression across the chest leads. The ventricle was overdrive paced as seen in the first half of the ECG; **(C)** shows the intra-cardiac signals. The HIS catheter is being utilized for ventricular overdrive pacing. Atrial signals in the CS catheter show an atrial tachycardia cycle length of 310 m s. The atrial bipolar signals at the RF catheter, which is placed near the right atrial appendage (RAA), reveals early signals with respect to the onset of P wave on the surface ECG; **(D)** shows 3D signals and ablation of RA appendage tachycardia using NavX Precision system (St. Jude Medical, St. Paul, MN, USA). Isochronal activation map has been constructed during tachycardia using proximal CS atrial signal as a reference. The RAA and the right atrium is seen in the RAO view (first half) and the left lateral view (second half). The color-coded display of the activation time in the right atrium measured relative to the reference point with red being early and blue being late. Activation map shows a very early target (white) at the base of the right atrial appendage. Ablation sites are colored brown.

electrode spacing) was used to map the right atrium (RA). Three dimensional (3-D) electro-anatomical mapping using NavX Precision system (St. Jude Medical, St. Paul, MN, USA) was utilized for mapping of the tachycardia. Radiofrequency ablation (RFA) was performed using an open-irrigated, 7F, 4 mm tip, quadri-polar catheter (Coolflex, St. Jude). No fluoroscopy was used for placement of catheters or for mapping of the atrium or even for the ablation.

Intra-cardiac signals revealed an atrial cycle length (CL) of 310 m s (Fig. 1C). A 1:1 atrio-ventricular conduction resulted in ventricular signals of the same CL. The V-V intervals (Fig. 1C) showed subtle variations due to changing A-H intervals. The activation time of bilateral atria from the RA free wall to CS- distal was only 40 m s. Entrainment from the different sites of the tricuspid annulus, the proximal and distal CS electrodes resulted in overdrive suppression and reinitiation of the tachycardia at long post-pacing intervals (390–480 m s) with variable V-A linking. All these evidences had effectively ruled out a reentrant mechanism of the tachyarrhythmia.

Ventricular overdrive pacing resulted in suppression and reinitiation of the tachycardia with an ectopic P wave (Fig. 1B). The P wave morphology was studied to help in locating the ectopic origin of the atrial tachycardia. Positive deflections in the inferior leads, negative deflection in V1 and progressive increase in p-wave amplitude across the chest leads suggested a right atrial appendage or crista terminalis (CT) or a superior tricuspid annulus as the probable origin of the tachycardia. Mapping near the RA appendage (RAA) showed early signals in the region of RAA. The 3-D electro-anatomical map suggested a centrifugal pattern of right atrial activation spreading from the base of the RAA (Supplementary video 3). The His-signals were tagged (Fig. 1D).

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Further mapping was carried out using the ablation catheter. The base of the RAA showed the earliest atrial activation signals (−32 m s) with respect to the onset of P wave on the surface ECG (Fig. 1C). Pacing at this site also resulted in short post-pacing intervals (330 m s). RFA was performed targeting a power of 25 watts (W), temperature of 40^{O} C, lasting for 10–20 seconds using an open-irrigation catheter with a flow rate of 17ml/minute. After 2–3 lesions, the power was uptitrated to 30 W. Within 10 seconds of the RF burn the tachycardia terminated after initial prolongation of cycle length (Fig. 2A), after which the burn was continued for 60 seconds. At the end of the procedure, an angiogram of the RAA had ruled out anomalies of the appendage (Fig. 2B). The 12-lead ECG, after ablation, revealed a sinus origin of the rhythm (Fig. 2C) which was eventually confirmed by recording the atrial activation pattern at the high RA. Acute procedural success was achieved, as defined by the inability to induce tachycardia 30 min after ablation despite aggressive burst atrial pacing and the use of isoproterenol. The baby was extubated on the same day. The left ventricular functions had returned to normal by the fourth week of the procedure (Supplementary video 2). The baby was discharged without any cardiac medications. The baby was followed-up at 9 months after the discharge and there was no recurrence.

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