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A Rare Case of Atrioventricular Node Cystic Tumor

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Abstract | **Case Report**

Introduction: Cystic tumors of the atrioventricular (AV) node are exceptionally rare benign cardiac neoplasms with the potential to disrupt cardiac conduction, leading to complete heart block or sudden cardiac death. Due to their rarity and nonspecific clinical presentation, diagnosis often relies on multimodal imaging and histological confirmation. ***Case Presentation:*** We reported a case of 38-year-old woman who presented with an incidental complete heart block during hospitalization for community-acquired pneumonia. Imaging studies revealed a well-defined right atrial mass adjacent to the interatrial septum. Cardiac magnetic resonance imaging suggested a fibroma; however, intraoperative findings revealed a cystic, multi-septated mass in the triangle of Koch. The patient underwent successful minimally invasive right atrial mass excision. Histopathological examination confirmed a benign cystic tumor of the AV node. The patient recovered uneventfully and remained stable pending permanent pacemaker implantation. ***Conclusion:*** AV node cystic tumors, though benign, carry a high risk of conduction abnormalities and sudden cardiac death. Early detection and surgical intervention are crucial. This case highlights the utility of multimodal imaging and supports the feasibility and safety of minimally invasive cardiac surgery for tumor resection.

Keywords: Atrioventricular node, Cystic Tumor, Cardiac Neoplasm, Right Atrial Mass, Complete Heart Block, Minimally Invasive Cardiac Surgery.

INTRODUCTION

Primary cardiac tumors are rare, occurring in 0.0017% to 0.03% of all autopsies, with benign tumors being more common than malignant ones [1]. A cystic tumor of the atrioventricular (AV) node, also referred to as mesothelioma of the AV node, is a benign congenital growth found in the triangle of Koch within the AV nodal region of the atrial septum of the heart [1]. This unusual lesion typically behaves benignly, but due to its proximity to crucial conduction pathways, it can lead to sudden cardiac death [1]. Due to its severe prognosis, surgical removal (which necessitates the placement of a permanent pacemaker) is advised if detected through clinical imaging [2]. Most affected patients are female, with a 3:1 female-to-male ratio, and the condition is typically diagnosed in the fourth decade of life [3].

Patel J *et al.,* reported three cases of sudden death where autopsy revealed microscopic cystic tumors in the AV node, with the cysts and cells infiltrating the AV node itself [4]. Velasco *et al.,* described a case where a patient with Mobitz type 1 second-degree AV block and 2:1 AV block with narrow QRS complex improved and reverted to normal sinus rhythm after the tumor was excised [5].

We reported a rare case of atrioventricular node cystic tumor.

CASE PRESENTATION

A 38-year-old Indian lady with underlying bronchial asthma and large complex cystic ovarian mass was admitted to a district hospital for acute exacerbation of bronchial asthma secondary to community acquired pneumonia. Incidentally noted complete heart block on electrocardiogram (ECG) with a heart rate of 50-60 beats per minute. Her blood test and electrolytes were unremarkable.

She was referred to a tertiary center with cardiology services. A transthoracic echocardiogram (ECHO) showed normal left and right ventricular contractility with an echogenic mass (1.77cm^2 - 2cm^2) seen in the right atrium (RA) at the interatrial septum.

Cardiac magnetic resonance (CMR) showed that the RA mass measured 18mm x 10mm x 22mm x 13mm. It had a well-defined margin, not mobile and not attached to the tricuspid valve. The mass appears isointense in T1TSE and early gadolinium images, hypointense in T2TSE and T2STIR images, no contrast uptake on first pass perfusion images, and homogenous hyperenhancement in late gadolinium images. CMR features were suggestive of a cardiac fibroma.

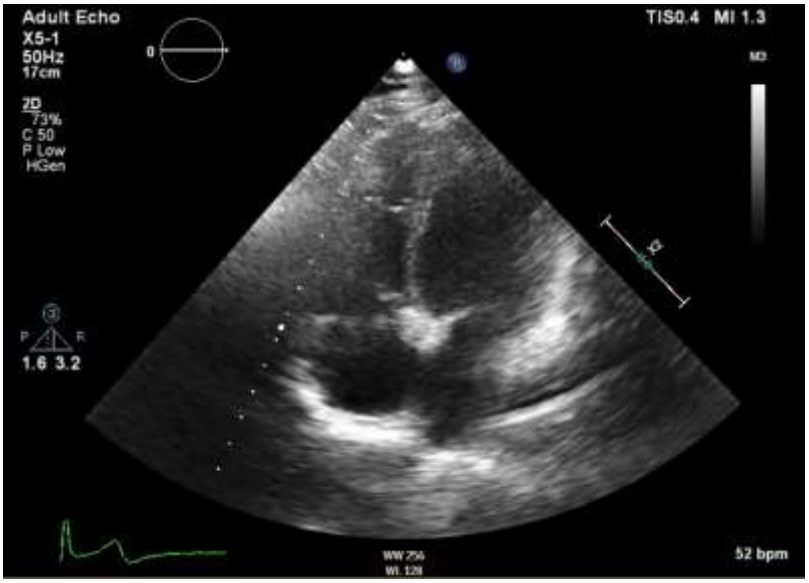

Figure 1: Transthoracic echocardiography shows an echogenic RA mass

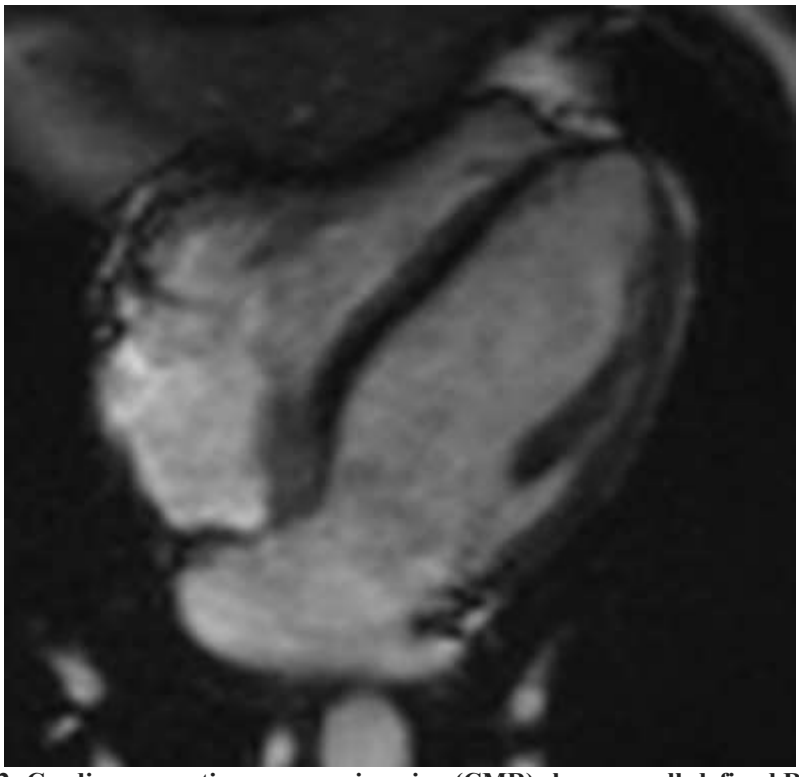

Figure 2: Cardiac magnetic resonance imaging (CMR) shows a well-defined RA mass

She was then referred to Cardiothoracic Surgery Department in Hospital Pulau Pinang, where she underwent a successful minimally invasive right atrial mass excision in November 2024. Surgical approach was via a right anterolateral mini-thoracotomy at the 4th intercostal space, a camera port was created 1 intercostal space above at anterior axillary line using a 30-degree 10mm scope. After systemic heparinization, cardiopulmonary bypass was achieved via right femoral vessels cannulation. Core temperature was lowered to 32'c. Aorta was cross-clamped and Del Nido cardioplegia was given via the antegrade cardioplegia needle at the aortic root to achieve diastolic arrest. The right atrium was then opened up, the mass was located in

the triangle of Koch in between the coronary sinus opening and septal leaflet of the tricuspid valve. It measures ~1.5cm x 1.5cm and has a cystic nature with thick mustard yellow color fluid within. The mass was multi-septated, excised and sent for histopathological examination (HPE). The remaining edges of the cyst were marsupialized. Patient was rewarmed, the right atrium was closed up, the aortic cross clamp released, and the patient was weaned off cardiopulmonary bypass with a bipolar ventricular pacing wire in-situ. After femoral decannulation and meticulous hemostasis, a right pleural drain was inserted, and finally all incisions were closed up. Patient was transferred to the cardiothoracic intensive care unit where she had an uneventful recovery post operatively. An ECHO prior to discharge no longer showed the RA mass.

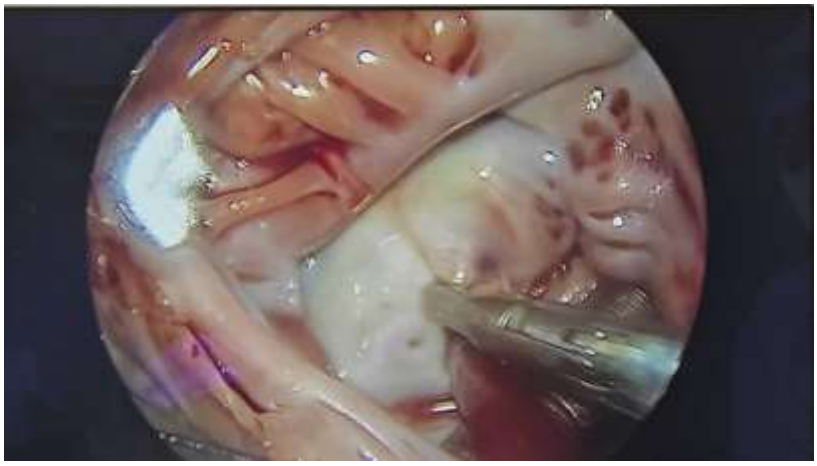

Figure 3: Intraoperative image of the AV node cyst

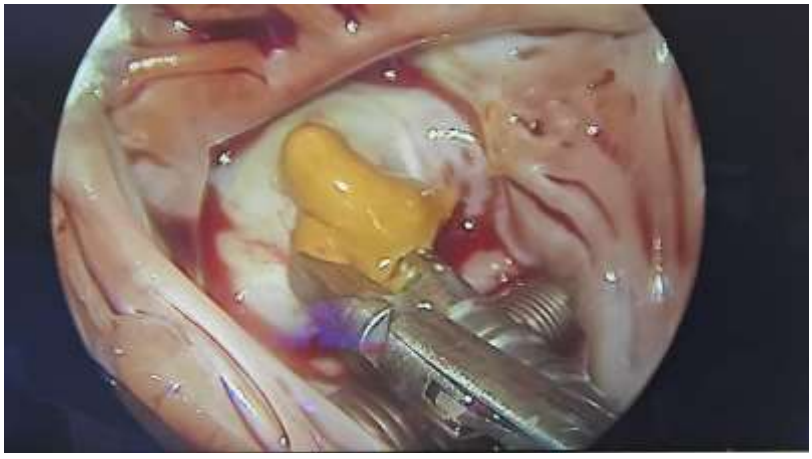

Figure 4: Thick mustard yellow AV node cystic fluid

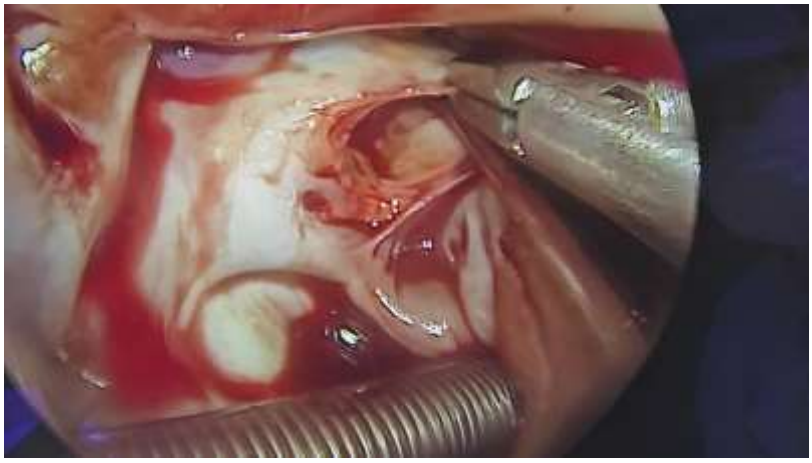

Figure 5: Multiseptated AV node cyst

Few weeks later, the histopathology examination (HPE) report showed fragments of fibrous

tissue partly lined by benign stratified squamous epithelium to simple columnar and cuboidal epithelium. A few cystic spaces lined by similar epithelial layer with scanty intraluminal secretory material is also present within the stroma. The stroma shows vascularized fibrous tissue with minimal inflammation and focal calcification. No mitotic activity, cytological atypia or teratomatous element identified. Immunohistochemistry (IHC) studies showed that the epithelial layer is positive for CKAE1/AE3 and CK5/6, and negative for calretinin, WT1, and CD34. Histological features were suggestive of cystic tumor of atrioventricular node.

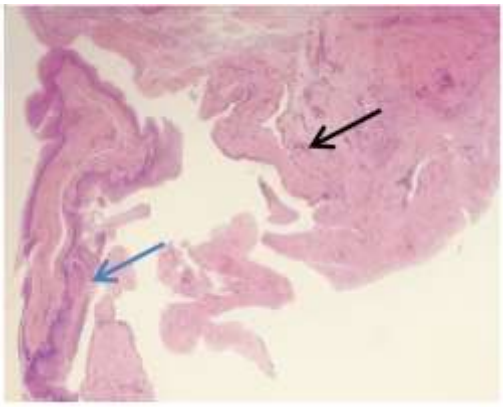

Figure 6: Fibrocollagenous cyst wall lined by benign stratified non-keratinizing squamous epithelium (blue arrow) and single layered cuboidal epithelium (black arrow). (x40)

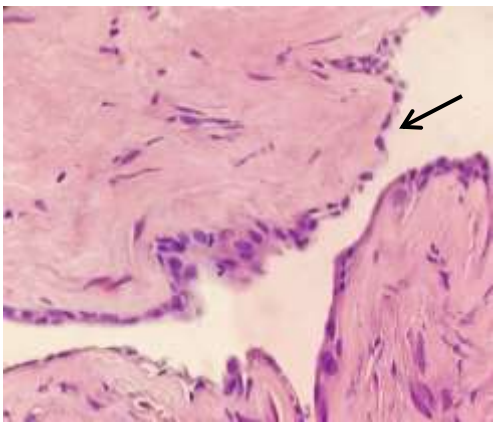

Figure 7: The single layer cuboidal epithelium in higher magnification (x400), showing bland looking, round nuclei. No overt nuclear atypia or mitoses seen.

A follow up transthoracic ECHO at 6 months post operation showed no recurrence of RA mass.

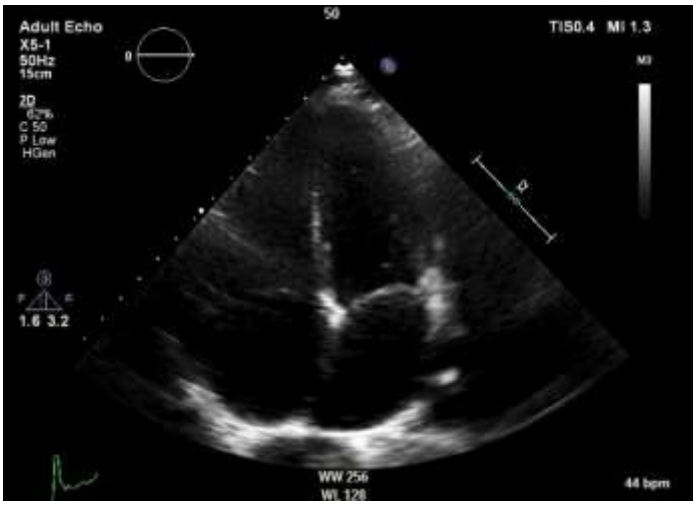

Figure 8: Trans-thoracic echocardiography at 6 months post operation did not show any recurrence of RA mass.

Discussion

Cystic tumors of the atrioventricular (AV) node are rare primary cardiac tumors located at the base of the atrial septum [6,7]. They can be associated with congenital abnormalities, including complex congenital heart disease, ovarian cysts, breast cysts, and thyroglossal duct cysts [1,3]. These tumors are more commonly found in females, with an average age of incidence being 38 years old (ranging from birth to 89 years) [1,8]. It has been reported that this tumor can disrupt AV node conduction, leading to 65% of cases resulting in complete heart block and 15% causing partial AV block [1,9]. In the case we present, the patient is 38 years old with a large ovarian cyst and incidental findings of complete heart block.

Accurate diagnosis requires multimodal imaging and histological evaluation. Echocardiography (ECHO) plays a key role in identifying intracardiac masses, which prompts further investigation through computed tomography (CT) and/or cardiac magnetic resonance (CMR) [10,11]. However, there has been a reported case where no mass was visible on ECHO [12].

Treatment options for cystic tumors of the atrioventricular (AV) node are controversial due to the rarity of this condition, and there is no clear consensus or established guideline. One concern is the risk of sudden cardiac death even after the implantation of a permanent pacemaker or implantable cardioverter-defibrillator (ICD), as the tumor may invade the AV conduction system or the proximal His bundle, potentially leading to ventricular fibrillation [4,12,13,17]. As a result, surgical resection is generally recommended [1,13,14]. In the modern era, minimally invasive cardiac surgery (MICS) is preferred over the conventional sternotomy approach [7,15,16]. The decision regarding partial versus complete tumor excision remains debated. Some experts recommend complete excision and the implantation of a pacemaker or ICD if necessary [1,10,12] while others advocate for partial excision to avoid pacemaker implantation [1,18]. Notably, there have been no reports of recurrence following partial resection [1].

In our case, we performed MICS via right anterolateral thoracotomy with cardiopulmonary bypass through femoral cannulation, followed by partial resection of the AV node cyst and marsupialization of the cyst edges. Despite the partial resection, the patient remained in complete heart block, and is planned for the implantation of a permanent pacemaker prior to her ovarian cyst removal surgery.

We recommend a regular follow up to monitor any recurrence, that is ECHO 6 monthly for a year, followed by ECHO annually for 3 years.

Conclusion

Special attention should be given when diagnosing a young female with AV node heart block, as this could indicate the presence of this rare entity, and prompt surgical intervention may help prevent sudden cardiac death. Minimally invasive cardiac surgery (MICS) is a safe approach that allows for a quicker recovery. Partial resection of the tumor is also considered safe and may help avoid the need for pacemaker or ICD implantation.

Ethical Approval: This case report has been registered under National Medical Research Register (NMRR).

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Author Contribution: KM Cheng is the primary author who did the literature research and case write up. PS Law assisted in writing and provided intra-operative images for the case report. TM Chen and NHM Amin are the referring cardiologists and provided investigational images and proof reading. FW Lee is the pathologist who reported the HPE specimen and provided the HPE images. AY Khamis is the supervisor and consultant cardiothoracic surgeon who performed the operation..

Conflict of Interest Disclosure: The authors declare no conflict of interest with regards to the content of this report.

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