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An incidental finding of an atrioventricular node tumor during standard hospitalization years after pacemaker implantation

Short title: Rare atrioventricular node tumor diagnosed incidentally

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A 64-year-old woman with complete atrioventricular (AV) block observed from early childhood was admitted to the hospital for elective replacement of a pulse generator of dual-chamber pacemaker (PM), which had been implanted ten years before. A PM implantation was postponed for many years due to lack of symptoms, however patient developed worsening bradycardia with presyncope. Transthoracic echocardiography (TTE) performed at the time of implantation showed no significant abnormalities in the right atrium nor interatrial septum.

During hospitalization for generator replacement, TTE revealed a hyperechoic tumor (22 × 16 mm) in the right atrium, adjacent to the lower portion of the interatrial septum (Figure 1A), and moderate mitral valve regurgitation due to prolapse of the A1 and 2 segments

(Supplementary material). Further studies included transesophageal echocardiography and computed tomography (CT) (Supplementary material). Computed tomography scans were highly suggestive for a cystic tumor of the AV node (CTAVN) (Figure 1 B–D). Patient was scheduled for elective tumor excision and mitral valve repair. During surgery tumor filled with caseous masses and located at the Koch's triangle of the interatrial septum was revealed (Figure 1E). The tumor was then partially excised, and the obtained material was sent for further testing. Subsequently, the patient underwent a mitral valve repair with the implantation of the Gore-Tex Sutures (W. L. Gore & Associates, Inc.; Newark, DE, US) and a 30-mm Carpentier–Edwards Physio annuloplasty ring (Edwards Lifesciences, Irvine, CA, US). Recovery was uneventful except for an increase in the atrial pacing threshold. VDD pacing mode was successfully applied. The cultures of the caseous masses were negative, and the pathological examination confirmed diagnosis of CTAVN (Figure 1F). Transthoracic echocardiography performed 6-months later was negative for the presence of a right atrial tumor.

Cystic tumor of the AV node is an extremely rare primary cardiac tumor, accounting for approximately 3% of all cardiac tumors. According to current knowledge, CTAVN is not a malignant tumor but rather a congenital heterotopia resulting from abnormal migration of cardiac neural crest cells [1]. The presence of concomitant congenital defects in patients with CTAVN supports the theory that the tumor originates in utero. It is more common in women and may lead to advanced AV conduction disorders and sudden cardiac death. Only few cases of premortem diagnosis have been reported in the literature [2]. Cystic tumor of the AV node should be considered in young patients with complete AV block. Multimodality imaging is a helpful aid in differentiating atrial masses [3, 4]. Thus approach incorporating multimodality imaging may increase chances of appropriate diagnosis and effective treatment in this population.

To our knowledge, no cases of mitral valve prolapse associated with CTAVN have been reported in the literature. Given the relatively small number of cases identified to date, it is challenging to determine the optimal therapeutic approach. In published cases, cardiac surgery was performed with either complete or partial tumor resection. There was no recurrence during follow-up, and in a few cases, AV conduction disorders completely resolved. Persisting AV block requires permanent pacing, which was the case in our patient.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal

Article information

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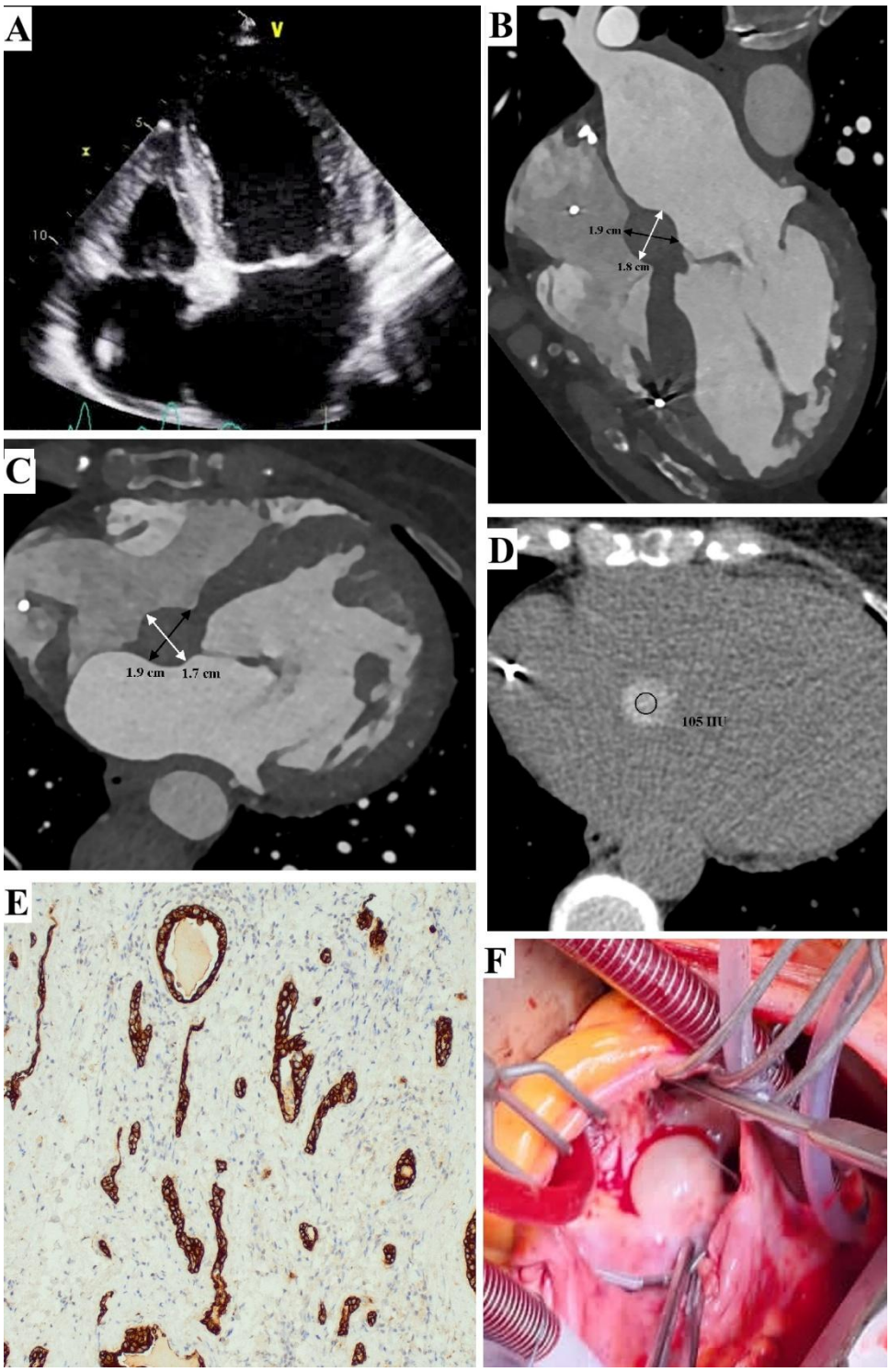

Figure 1. A. Apical 4-chamber 2-dimensional transthoracic echocardiography image showing a round echogenic mass in the right atrium, in the lower part of the interatrial septum, measuring 22 × 16 mm; **B.–D.** cardiac computed tomography (CT) scan showing a smoothly contoured oval lesion in the interatrial septum above the atrioventricular valves, measuring approximately 1.9 × 1.8 cm on long-axis, 4-chamber multiplanar reconstruction images (**B**) and approximately 1.9 × 1.7 cm on axial images (**C**); the native density of the lesion is approximately 80–110 HU (**D**), with postcontrast enhancement; the CT image suggests a cystic tumor of the atrioventricular node; **E.** the cell lining of cystic structures showed cytokeratin (AE1/AE3)

positivity (100× magnification); **F.** intraoperative image showing the location of atrioventricular node tumor within the Koch's triangle in the region of the interatrial septum