

A Giant Eustachian Valve Protruding Into the Right Ventricle: A Case Report

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Received: May 4, 2014; Revised: March 5, 2014; Accepted: May 18, 2014

Introduction: The Eustachian valve (EV) remnant, when present in adults, is usually rudimentary. However, in echocardiographic examinations, it may appear as a mobile long structure in the right atrium, and it rarely protrudes into the right ventricle. When it is quite large, the EV remnant could be misdiagnosed as a right atrial tumor, thrombus, or vegetation.

Case Presentation: An 83-year-old patient was referred to the surgical ward for the excision of a gastric adenocarcinoma. In the course of preoperative assessment, transthoracic echocardiography showed a right atrial mobile filamentous mass that was protruding into the right ventricle. Differential diagnosis included a tumor or thrombus. After a precise evaluation through multiple views, the mass was demonstrated to be a giant EV, 7.3 cm in length.

Conclusions: The giant EV remnant can persist in adults and is often diagnosed incidentally via echocardiography. Transthoracic echocardiography is a reliable noninvasive method for the diagnosis of the EV remnant and could help avoid its misdiagnosis as a tumor or thrombus. Nevertheless, sometimes transesophageal echocardiography is necessary to confirm the diagnosis or to demonstrate the existence of an additive clot on it.

Keywords: Eustachian Valve; Right Atrial Mass; Right Ventricle; Transthoracic Echocardiography

1. Introduction

The Eustachian valve (EV), which is referred to as a valve of the inferior vena cava (IVC), is an embryonic remnant of the right valve of the sinus venosus. Embryologically, the EV has the role of transferring the oxygenated blood from the IVC through the patent foramen ovale (PFO) into the systemic circulation. In adults typically, this structure may be completely absent (1) or it may appear as a thin flap originating from the anterior rim of the IVC orifice (2, 3). However, it may persist as a mobile long structure in the right atrium (RA). Occasionally, it protrudes into the right ventricle (RV) (4); this condition is usually found in infants (5, 6) and children (7) but rarely in adults (8).

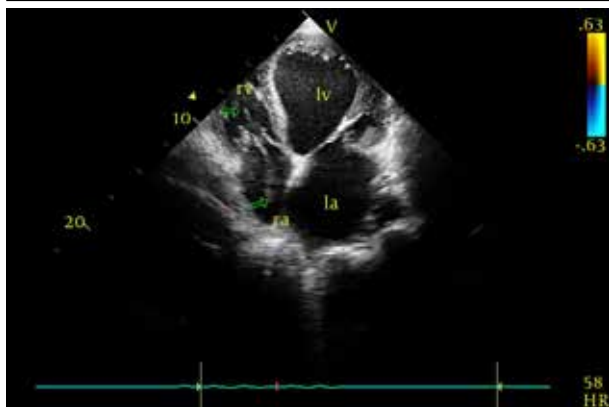
The persistence of the EV remnant can be visualized incidentally by transthoracic echocardiography (TTE) in only a minority of adults (9). Nonetheless, when it is elongated and large, it could be misdiagnosed as RA tumors, thrombi, or vegetations (2). Here we describe a rare case of a giant EV remnant, protruding into the RV in an adult patient with a gastric adenocarcinoma, which was incidentally detected by TTE. We also discuss the echocardiographic characteristics of the EV remnant, which may provide clues for its differential diagnosis from other RA masses.

2. Case Presentation

An 83-year-old man was admitted to the surgical ward for the excision of a gastric adenocarcinoma. He had a 10-

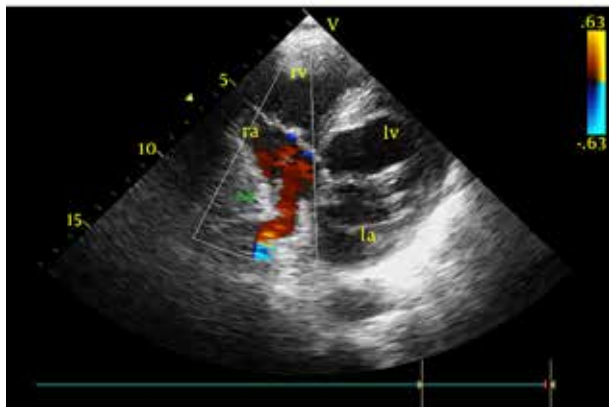
year history of diabetes mellitus but had not experienced transient ischemic attacks or stroke hitherto. Coronary angiography showed moderate stenosis (70%) in the left anterior descending artery. On physical examination, there was arrhythmic heart rhythm, systolic murmur at the apex, clear lungs, and no edema in the extremities. The body temperature was normal. The patient had progressive appetite and weight loss. Electrocardiography demonstrated atrial fibrillation with a heart rate of 100/minutes and blood pressure of 110/90 mmHg. Ultrasound scanning of the abdomen showed no presence of metastasis in the liver, kidney, or lymph nodes. TTE, performed as a part of preoperative assessment, revealed a dilated left ventricle with a mildly reduced systolic function (ejection fraction of about 50%). Also, moderate mitral valve regurgitation was present. The RV had a normal size with a preserved systolic function. A filamentous RA mass was observed in the RA in multiple views. The RA mass appeared as a thin, filamentous, highly mobile, long structure that was approximately 7.3 cm in length and was attached beneath the IVC orifice. This structure was protruding towards the tricuspid valve (Figure 1, Video 1). The tricuspid valve was anatomically and functionally intact (Figures 2a and 2b, Video 2). The differential diagnoses of the mass were firstly a tumor and then a thrombus. According to the patient's clinical condition vegetation was not

Figure 1. Apical Four-Chamber View during Diastole: A Giant Eustachian Valve Remnant (7.3 cm), Protruding into the Right Ventricle through the Tricuspid Valve (Green Arrows)



The mass was described as a mobile, large, filamentous RA structure. RA, Right atrium; RV, Right ventricle; LV, Left ventricle; LA, Left atrium

Figure 2. Tricuspid Valve, Anatomically and Functionally Intact



A) Modified apical four-chamber view during systole: The EV remnant can be visualized as a thin, mobile, filamentous structure (green arrow) in the RA that extends from the anterior rim of the IVC orifice (asterisk) into the RA cavity, without any interference during the closure of the tricuspid valve. By identifying its attachment to the orifice of the IVC, we can differentiate it from a tumor or a thrombus. RA, Right atrium; LA, Left atrium; LV, Left ventricle; VCI = IVC, Inferior vena cava. B) Color Doppler demonstrates a normal inflow from the IVC with no interference from the EV remnant (green arrow). RA, Right atrium; RV, Right ventricle; LV, Left ventricle; LA, Left atrium

considered. However, a thorough evaluation via multiple echocardiographic views led to the diagnosis of this RA mass as a normal anatomic structure: the EV remnant. The diagnosis was borne out by the morphology of the mass as a filamentous structure and its typical location at the junction of the RA and IVC. The finding of this giant EV remnant had no apparent clinical consequences, so the patient underwent the surgical procedure for adenocarcinoma with an uneventful postoperative course.

3. Discussion

The EV in the fetus directs the oxygenated blood from the IVC through the foramen ovale into the systemic circulation. The valve gradually regresses in the newborn or childhood period, so the echocardiographic prominence of the EV remnant depends on the subject's age. The valve is usually visible by TTE in 85% in newborns, in 62% in children, and only in a minority of adults (9), usually in the subcostal four-chamber, parasternal short-axis, or apical four-chamber views (2). However, the persistence of the EV remnant is associated with large variability in size, shape, texture, and thickness. In adults, it is completely absent or is represented only by a thin ridge arising from the anterior rim of the IVC orifice. But sometimes, it may persist as a mobile elongated structure in the RA cavity, demonstrating an undulating motion in real-time echocardiography (3). Occasionally, it protrudes into the RV through the tricuspid valve (4); this condition is usually found in infants (5, 6) and children (7) but rarely in adults (8). Although the length of the EV remnant is not frequently reported, a recent echocardiographic study (10) reported the average length of 2.3 cm in the range of 0.7-4.7 cm. In our patient, the EV remnant was extremely large (length of approximately 7.3 cm), and it extended from the IVC orifice into the RA and protruded into the RV.

A persisting EV can indirectly predispose to paradoxical embolism by directing the blood from the IVC to the interatrial septum and then through a PFO in the left atrium and subsequently into the systemic circulation. Moreover, a persisting EV may prevent the spontaneous closure of the PFO after birth. Therefore, the persistent EV may predispose to the patency of the foramen ovale, especially in patients with stroke. Moreover, when the EV remnant is prominent and large, it can mimic RA masses such as tumors, thrombi, and vegetations (1, 3). The echocardiographic criteria which are pathognomonic for the EV remnant are the typical location and morphology. The typical location is the origin at the junction between the IVC and the RA (1, 6). The specific morphological feature of a normal EV remnant is its thin, filamentous, dense structure (1, 6, 11) and it has a predictable oscillating motion (9). In our patient, both, i.e. the typical location and morphology, suggested an EV remnant.

Tumors of the RA are unusual. Myxomas are the most common primary benign tumors inasmuch as they occur in the RA in about 15 to 20% of the cases. Myxomas are typically attached to the interatrial septum, at the border

of the fossa ovalis (12, 13). Echocardiographically, they have a globular or spherical shape with a friable surface, have heterogeneous internal echogenicity (4, 12), and are typically pedunculated (13). The most important echocardiographic clue to the diagnosis of myxomas is the attachment site to the mid-portion of the atrial septum (13). In our patient, echocardiography clearly demonstrated that the mass was not attached to the atrial septum.

Metastatic tumors that arise from the abdomen and pelvis are a more common cause of the RA involvement (13). A specific type of cardiac involvement by a metastatic tumor is tumor extension along the IVC and into the RA as a finger-like projection (4). This is most frequently described in renal cell carcinomas, hepatomas, Wilm's tumors, and uterine leiomyomas (13). This should be recognized by the echocardiographer and should be differentiated from the EV remnant by careful tracing of the origin of the mass (12). Our patient had a primary gastric adenocarcinoma, without the presence of metastasis in the liver or kidneys. Most thrombi that may be found in the RA are thromboemboli in transit that arise in the low extremity veins. Migrating thrombi echocardiographically are presented as snake-like structures that are highly mobile and thicker than the EV remnant. Additionally, the end of the thrombi is not attached around the IVC orifice (12). In our patient, the attachment site to the IVC orifice was clearly visualized.

When clinical condition suggests infectious endocarditis, right-sided vegetations should be differentiated from the EV remnant. Typically, vegetations are attached to right-sided valves as an oscillating mass with an irregular shape and a motion independent to that of the valve (4, 14). Vegetations are also found on right-sided prostheses and on foreign bodies such as pacemaker lead and indwelling catheters (14). In our patient, there were no such conditions. In addition, the EV remnant should be differentiated from the other normal anatomic structures such as Chiari's network, a highly mobile fenestrated network of tissue, which is not attached to the IVC (2). Chiari's network can be differentiated from a large EV remnant by looking for attachments to the different parts of the RA (15). Furthermore, it is much more mobile and thinner than is the EV remnant (12). The EV remnant should not be termed a Chiari's network, even if it is mobile and fenestrated (16). Since these echocardiographic criteria were absent in our patient, Chiari's network was not considered as a possible diagnosis.

A giant EV remnant is often considered clinically insignificant. However, it may be associated with the persistence of a PFO, usually found in patients with stroke (17). The fact that our patient had not suffered stroke so far ruled out the PFO. Sometimes a giant EV remnant may interfere with cardiac catheterization (2). Occasionally, higher insertion of a giant EV into the lower portion of the interatrial septum can mimic the echocardiographic appearance of a divided right atrium. This type of abnormality may be confused with cor triatriatum dexter (3,

12). Although extremely rare, the EV can also serve as a nidus for myxomas (18), fibroelastomas (19), endocarditis (3, 20), or thrombi (3, 21).

Interestingly, a prominent EV remnant is usually found in association with congenital heart disease (e.g. Ebstein's malformation of the tricuspid valve and tetralogy of Fallot). In the absence of other significant structural heart disease, usually no treatment is required in adults (3, 6). As much as the EV remnant was large in our patient, it had no functional impact on the right heart function. Therefore, in this case, the finding of a very prominent EV remnant was clinically insignificant and required no management.

A giant EV remnant can persist in adults and is often diagnosed incidentally with echocardiography. TTE is a reliable noninvasive method for the diagnosis of the EV remnant when the typical echocardiographic characteristics are identified, and can potentially help avoid diagnostic misinterpretations as a tumor or thrombus. Nevertheless, sometimes transesophageal echocardiography is necessary to confirm the diagnosis or the existence of an additive clot on it. In adults, even when a persisting EV remnant is very large, it is considered to be a benign finding in the absence of associated cardiac anomalies.

Authors' Contributions

Darko Angjushev was involved in patient care and wrote the manuscript, Marija Kotevska-Angjushev performed the echocardiographic examinations, and Miroslav Lazarevski critically revised the manuscript.

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