Functional paraganglioma in the right atrium

A 39-year-old male presented to our clinic because of nocturnal intermittent palpitation for 2 years. His medical history showed a high blood pressure of 200/100 mm Hg. Transthoracic echocardiography revealed a 54×46-mm mass in the right atrium (Figure 1a, 1b). In addition, cardiac magnetic resonance imaging confirmed the tumor and highly suggested it to be a malignant tumor (Figure 1c, 1d). An almost 50-fold increase in the blood catecholamine metabolites was detected. Further, positron emission tomography/computed tomography found no other tumor. Given the symptoms and the functional intra-atrial tumor, surgical removal was agreed by our surgical team and the family. During the surgery, we found that the tumor had invaded into the right atrial wall and inter-atrial septum (Figure 1e, 1f). Following the complete removal of the tumor, we reconstructed the right atrium and inter-atrial septum with bovine pericardium. Intraoperative echocardiography showed no shunt between the atria and reflow of the pulmonary veins were clear. Pathological analysis revealed the tumor to be a paraganglioma with negative CK and EMA expressions and positive CD56, Syn, and S-100 expressions



Figure 1. Transthoracic echocardiography images showed a tumor in the right atrium (a, b). Cardiac magnetic resonance imaging showed an equal T1 and long T2 signal shadow in the right atrium, which was suspected as a malignant tumor (c, d). Intraoperative images and the gross anatomy of the tumor (e, f). Immunohistochemical study of the tumor for CD56 expression (g). Immunohistochemical study of the tumor for Syn expression (h). Immunohistochemical study of the tumor for S-100 expression (i)

(Figure 1g–1i). The patient fully recovered and was discharged on postoperative day 7 with a normal blood pressure. At the 6-month follow-up, the patient was still free of symptoms and catecholamine metabolites were normal. Intra-atrial paraganglioma is an extremely rare and functional primary cardiac neoplasm, and our experience suggests multimodality evaluations and complete surgical removal be performed.

Jingxiu Fan*, Zhong Wu*, Yan Kang¹ Departments of Cardiovascular Surgery, and ¹Department of Critical Care Medicine, West China Hospital/West China School of Medicine, Sichuan University; Chengdu-*People's Republic of China*

*These authors contribute equally to this article.

Address for Correspondence: Yan Kang, M.D,

Department of Critical Care Medicine, West China Hospital/West China School of Medicine, Sichuan University, Chengdu 610041, *People's Republic of China*. Address: Guo Xue Alley 37, Chengdu, Sichuan, *People's Republic of China*, 610041 Phone: +86 028 85422234

E-mail: kang_yan_123@163.com

©Copyright 2018 by Turkish Society of Cardiology - Available online at www.anatoljcardiol.com DOI:10.14744/AnatolJCardiol.2017.8241

Spontaneous giant pseudoaneurysm in the upper segment of the aortic arch

A 57-year-old male patient was referred to the Department of Emergency with a chief complaint of a 2-month history of back pain and short of breath for 1 day. Meanwhile, he developed hypertension 1 month ago that was not treated. The patient denied any history of trauma, operation, allergy, any types of hepatitis, or other potential infectious diseases. Thoracic computed tomography angiography on admission indicated a giant pseudoaneurysm in the upper segment of the aortic arch with a maximum



Figure 1. (a) Thoracic computed tomography angiography (CTA) showed a giant pseudoaneurysm in the initiating part of the upper segment of the aortic arch. (b) Three-dimensional reconstruction thoracic CTA showed a close relationship between the aneurysm and the three main branches from the aortic arch

cross-sectional area of 93×98 mm and formation of inner arterial wall thrombus that compressed the trachea and esophagus, causing displacement of the esophagus and tracheal stenosis. Three original arterial bifurcation branches from the aortic arch were initiating from the aortic pseudoaneurysm, with the proximal part of the left common carotid artery narrowing down (Fig. 1a, 1b).

Statistically, spontaneous giant pseudoaneurysms seldom occur in patients without a history of operation, trauma, or hypertension; however, the patient in this case developed suppression of the trachea with a 2-month history of lasting thoracic pain. Because the patient lived in a remote area, he did not seek any form of regular treatment during the 2 months, which may have resulted in the huge spontaneous pseudoaneurysm. Surgery is the first-line treatment for spontaneous pseudoaneurysm, whereas interventional occlusion can also be used when surgery is not possible (1). Unfortunately, despite the large number of possible convenient and efficient therapeutic remedies, the patient and his family denied any form of treatment. During a telephonic follow-up, we found that the patient had died the same day of hospital discharge.

Acknowledgement of grant support: This work was supported by the Chengdu Science and Technology Project (2015-HM01-00587-SF).

Reference

 Xiang DK, Hu K, Luo HB. Spontaneous Giant Ascending Aortic Pseudoaneurysm in a Previously Healthy Man. JACC Cardiovasc Interv 2016; 9: e155-7.

Qian He[#], Hong-yu Jin^{#1}, Ji-wen Geng¹ Department of Emergency, West China Hospital, Sichuan University; Chengdu-*China* ¹West China School of Medicine, Sichuan University; Chengdu-*China*

#These authors contributed equally.

Address for Correspondence: Qian He, MD, Department of Emergency, West China Hospital, Si Chuan University, Guo Xue Alley 37#, Chengdu-China Zip code: 610041 Phone: +86 028 85422286 Fax: +86 028 85422286 E-mail: 20740699@qq.com ©Copyright 2018 by Turkish Society of Cardiology - Available online at www.anatoljcardiol.com D0I:10.14744/AnatolJCardiol.2017.8267

An unusual cause of aortic regurgitation: accessory mitral valve 🚳

Accessory mitral valve (AMV) is a rare congenital cardiac anomaly that has been reported in 1 in 26,000 people in an echocardiography series. Most cases of AMV are associated with other cardiac abnormalities. Although patients with AMV are

Figure 1. There is an accessory mitral valve image in TTE window



Figure 2. Post- operative photo of the accessory mitral valve

usually asymptomatic, syncope, exercise intolerance, and chest pain are described in patients with left ventricular outflow (LVOT) obstruction. Here we present the case of an asymptomatic child with a diagnosis of AMV using transesophageal echocardiography (TEE) images.

A 17-year-old boy was diagnosed with suspected AMV during the evaluation of a cardiac murmur. Because he had no other associated anomalies, aortic regurgitation (AR), or LVOT obstruction, he had been followed up for 8 years by serial transthoracic echocardiography. However, the last echocardiography revealed a mild AR. Therefore, TEE was performed to confirm the diagnosis. AMV was clearly demonstrated without LVOT obstruction (Fig. 1 and Video 1). Surgical excision of AMV was performed because of new development of AR (Fig. 2). There was no AR and mitral regurgitation in the postoperative period.

Surgical excision of AMV has some difficulties and can be complicated because after initiation of cardiopulmonary bypass,