The diagnosis and clinical importance of tumour neovascularisation from coronary artery to right atrial myxoma

Rozpoznanie i znaczenie kliniczne neowaskularyzacji śluzaka prawego przedsionka z tętnicy wieńcowej

Sadik Acikel¹, Ozlem Bozkaya¹, Ugursay Kiziltepe²

¹Department of Cardiology, Ministry of Health Diskapi Yildirim Beyazit Research and Educational Hospital, Ankara, Turkey ²Department of Cardiovascular Surgery, Ministry of Health Diskapi Yildirim Beyazit Research and Educational Hospital, Ankara, Turkey

Abstract

Myxomas are the commonest cardiac tumours and are usually localised in the atria. Neovascularisation in cardiac myxomas has been shown in previous case reports. However, the clinical importance of neovascularisation in cardiac myxomas is not well understood. In our case report, we present a right atrial myxoma in a 46 year-old woman admitted to our hospital with exertional angina and dyspnea. Coronary angiography revealed the presence of tumour neovascularisation from the right coronary artery, with no evidence of coronary artery stenosis. We thus speculate that neovascularisation of myxoma may cause typical anginal symptoms as a result of coronary steal phenomenon. Coronary angiography might help in the evaluation of the neovascularisation process and also in indicating surgery.

Key words: myxoma, neovascularisation, echocardiography, angiography

Kardiol Pol 2012; 70, 5: 501–504

INTRODUCTION

Myxomas are the commonest cardiac tumours of the heart and are usually localised in the atria [1–3]. The clinical features depend on their size, location, mobility and the fragility of the mass [2]. Patients are usually asymptomatic, presenting with nonspecific signs and symptoms such as dyspnea, palpitations, anorexia and tiredness [4]. Tumour neovascularisation in cardiac myxomas has been shown in previous case reports [5–11]. However, very little is known about the effects of neovascularisation on the pre and postoperative clinical course, or about how tumour neovascularisation affects the patient's symptoms and prognosis.

In our case report, we demonstrated a right atrial myxoma after transthoracic and transoesophageal echocardiographic examinations, and pathological examination, in a patient presenting with typical exertional angina and dyspnea. During preoperative evaluation, coronary angiography (CAG) revealed the presence of tumour neovascularisation from the right coronary artery (RCA), with no evidence of coronary artery stenosis.

CASE REPORT

A 46 year-old woman was admitted to our hospital because of typical exertional angina and dyspnea. Her complaint had begun three months previously and progressed during the past weeks. She had no coronary artery risk factors, except cigarette smoking. She had no drug history. In her physical examination, her blood pressure was 120/70 mm Hg and her heart rate 90 bpm. She had clear lungs. There was no cardiac murmur. Chest radiography, kidney-liver function tests, haemogram, thyroid function tests and anaemia markers were all unremarkable. Electrocardiography showed sinus rhythm, and rarely an extra ventricular beat, and revealed no signs of coronary ischaemia. Echocardiography revealed a large

Address for correspondence:

Sadik Acikel, MD, Ministry of Health Diskapi Yildirim Beyazit Research and Educational Hospital, Department of Cardiology, 06110 Ankara, Turkey, tel: +90 312 5962943, fax: +90 312 3186690, e-mail: sadik.acikel@tkd.org.tr

Received: 19.04.2011 Accepted: 27.04.2011 Copyright © Polskie Towarzystwo Kardiologiczne

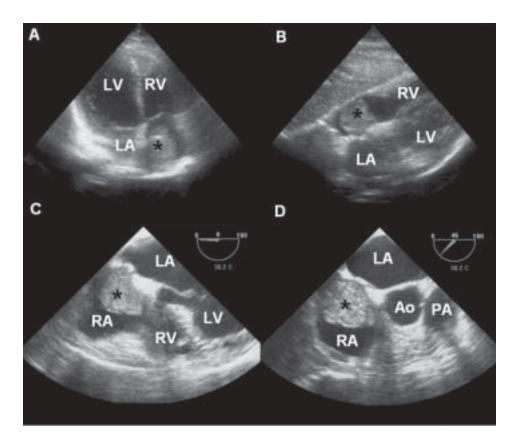


Figure 1. Right atrial mass (*) attached to the interatrial septum demonstrated by transthoracic echocardiographic apical four--chambered (**A**), subcostal (**B**) and transoesophageal echocardiographic views (**C**, **D**); Ao — aorta; LA — left atrium; LV — left ventricle; PA — pulmonary artery; RA — right atrium; RV — right ventricle

 $(4.1 \times 4.0 \text{ cm})$ and immobile mass in the right atrium (Figs. 1A, B). We performed transoesophageal echocardiography (TEE) in order to take a comprehensive look into the right atrial mass.

The TEE showed a large $(4.4 \times 4.0 \text{ cm})$, immobile, lobular, and homogeneous echogenic mass of regular shape in the right atrium, originating from the right side of the fossa ovale on the interatrial septum (Figs. 1B, C). There was no abnormal shunt from right to left with the agitated saline test on contrast echocardiography. The echocardiographic findings were consistent with right atrial myxoma. We performed CAG to rule out underlying coronary artery disease before cardiac surgery. There were no stenotic lesions in the coronary arteries, but, interestingly, we detected the presence of a tumour vessel arising from the RCA to the atrial mass (Figs. 2A, B). Complete tumour excision from the interatrial septum was achieved and the feeding artery from the RCA supplying the tumour was also ligated during cardiac surgery. After tumour excision, the atrial septal defect, approximately 1.5×1.5 cm, was closed with an intracardiac patch. Macroscopic examination of the resected mass showed a regular, smooth surface and globular shape (Figs. 3A, B). The diagnosis of myxoma was pathologically confirmed within the myxoid tissue around the typical myxoma cells, and also numerous proliferating small blood vessels were found (Figs. 3C, D). After the surgery, the patient did not have any symptoms and her transthoracic echocardiography and TEE was normal at three and six month follow-up.

DISCUSSION

In our case, typical exertional angina and dyspnea were the main symptoms of myxoma. Since significant feeding artery and intratumoural haemorrhagic fields were shown, we hypothesised that a significant amount of blood flow was drained from the patient's RCA to the tumour area, and that this may have resulted in a kind of coronary steal phenomenon. Therefore, we concluded that the coronary steal effect of neovascularisation may have caused the symptoms of typical angina and dyspnea as an angina equivalent in our case.

It has been established that angiogenesis is necessary for tumour development and proliferation, and that it is controlled by several angiogenic factors [12]. It has been also demonstrated that the neovascularisation process is closely related to vascular endothelial growth factors secreted from the tumour itself [12–14]. This evidence may explain the association between neovascularisation and increased growth of

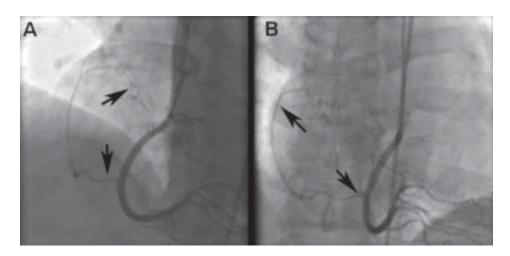


Figure 2. A demonstration of tumoural neovascularisation from the right coronary artery by right coronary artery angiography (A, B). Black arrows indicate neovascularisation of the tumour

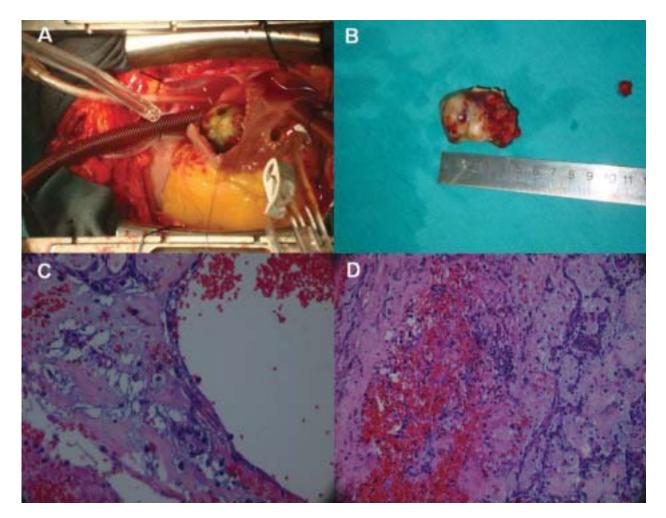


Figure 3. Gross appearance of the right atrial tumour before (A) and after (B) exision. Dilated vascular structures (C) and haemorrhaging, and typical myxoma cells around the myxoid stroma (D) with no evidence of malignancy in the sections stained with haematoxylin-eosin (\times 200)

a tumour. However, how the neovascularisation affects the patient's management, tumour recurrence, and prognosis are not well understood. Therefore, we believe that clinical decision making and the treatment of these patients in the presence of neovascularisation may also require further clinical investigations.

In the literature, neovascularisation has been reported in up to 80% of myxoma cases, and left- and right-sided myxomas were mostly supplied from the left circumflex artery or the RCA [5–11]. In our case, neovascularisation of a right atrial myxoma from the RCA was demonstrated in CAG. According to some authors, cardiac computed tomography angiography may be the choice for detecting tumour neovascularisation [5]. Some authors have suggested that CAG should be routinely performed in patients with cardiac myxoma to detect the presence of the supplying vessel [15]. However, in a large series of 28 patients who underwent excision of cardiac myxomas, Rahmanian et al. [16] concluded that CAG should only be performed on select patients, particularly those aged over 40 years and those with atherosclerotic risk factors who are undergoing surgery. In our case, we performed CAG because our patient had a coronary risk factor and had described typical exertional angina and dyspnea, which may have been an angina equivalent. We agree that CAG is important for ruling out coronary artery disease, and also possible tumour neovascularisation from coronary arteries, necessitating a surgical procedure.

In our case, preoperative demonstration of neovascularisation indicated surgery. Therefore, we believe that the diagnosis of myxoma neovascularisation may provide additional clues to indicate surgical procedures, and may also substantially change the operative strategy.

Conflict of interest: none declared

References

1. Panagiotou M, Panagopoulos ND, Ravazoula P, Kaklamanis L, Koletsis EN. Large asymptomatic left atrial myxoma with ossification: case report. J Cardiothorac Surg, 2008; 3: 19.

- Reynen K. Cardiac myxomas. N Engl J Med, 1995; 333: 1610– -1617.
- Peters PJ, Reinhardt S. The echocardiographic evaluation of intracardiac masses: a review. J Am Soc Echocardiogr, 2006; 19: 230–240.
- 4. liakos C, Alexiadou E, Metallidis S et al. Right atrium myxoma coexisting with antiphospholipid syndrome: a case report. Cardiovasc Ultrasound, 2009; 7: 47.
- 5. Fueredi GA, Knechtges TE, Czarnecki DJ. Coronary angiography in atrial myxoma: findings in nine cases. Am J Roentgenol, 1989; 152: 737–738.
- Kim YK, Yong HS, Kang EY, Woo OH. Left atrial myxoma with neovascularization: detected on cardiac computed tomography angiography. Int J Cardiovasc Imag, 2009; 25: 95–98.
- 7. Yazici M, Norgaz T, Akdemir R, Albayrak S. Asymptomatic giant left atrial myxoma supplied from right coronary artery in a 65-year-old woman. Int J Cardiol, 2005; 101:495–496.
- Ozdogru I, Duran M, Sarli B, Oguzhan A. Left atrial myxoma supplied by the circumflex coronary artery arising from the right sinus of Valsalva. Turk Kardiyol Dern Ars, 2008; 36: 549– -551.
- Reynen K, Köckeritz U, Taha M, Strasser RH. Neovascularization in left atrial myxoma. Z Kardiol, 2004; 93: 69–71.
- Acikel S, Aksoy MN, Kilic H et al. Cystic and hemorrhagic giant left atrial myxoma in a patient presenting with exertional angina and dyspnea. Cardiovasc Pathol, 2012; 21: e15–e18.
- Umeda Y, Matsuno Y, Imaizumi M, Mori Y, Iwata H, Takiya H. Right atrial myxoma with tumor vascularity originated from the left and right coronary arteries Int J Cardiol, 2009; 131: 137–139.
- 12. Amano J, Kono T, Wada Y et al. Cardiac myxoma: its origin and tumor characteristics. Ann Thorac Cardiovasc Surg, 2003; 9: 215–221.
- Kono T, Koide N, Hama Y et al. Expression of vascular endothelial growth factor and angiogenesis in cardiac myxoma: a study of fifteen patients. J Thorac Cardiovasc Surg, 2000; 119: 101–107.
- Sakamoto H, Sakamaki T, Kanda T et al. Vascular endothelial growth factor is an autocrine growth factor for cardiac myxoma cells. Circ J, 2004; 68: 488–493.
- Janas R, Jutley RS, Fenton P, Sarkar P. Should we perform preoperative coronary angiography in all cases of atrial myxomas? Catheter Cardiovasc Interv, 2006; 67: 379–383.
- Rahmanian PB, Castillo JG, Sanz J, Adams DH, Filsoufi F. Cardiac myxoma: preoperative diagnosis using a multimodal imaging approach and surgical outcome in a large contemporary series. Interact Cardiovasc Thorac Surg, 2007; 6: 479–483.