

Hypoxemia associated with right-side cardiac tumor: right atrial lymphoma with patent foramen ovale

Peter S. Y. Yu, Calvin S. H. Ng, Micky W. T. Kwok, Malcolm J. Underwood, Randolph H. L. Wong

Division of Cardiothoracic Surgery, Department of Surgery, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, Hong Kong
Correspondence to: Randolph H. L. Wong. Division of Cardiothoracic Surgery, Department of Surgery, The Chinese University of Hong Kong, Prince of Wales Hospital, 30-32 Ngan Shing Street, Shatin, New Territories, Hong Kong. Email: wonhl1@surgery.cuhk.edu.hk.

Abstract: A 66-year-old man presented with acute type I respiratory failure, and was diagnosed to have a huge mass occupying the right atrium (RA) of the heart, with no evidence of pulmonary embolism. Intra-operative transesophageal echocardiogram identified a patent foramen ovale (PFO) with right-to-left shunt apart from a huge right atrial tumor arising from the atrioventricular groove. The tumor was debulked, and the patent foramen was closed. Patient had an uneventful recovery and WAS discharged on post-operative day 7. Pathology revealed diffuse large B-cell lymphoma (DLBCL). The patient was further treated with adjuvant chemotherapy and remained well 1 year after surgery. This case illustrates the importance of meticulous echocardiographic examination to look for any intracardiac shunts in the case of hypoxemia associated with a right-sided cardiac tumor. We advocate surgery as the most important initial management to maximize survival, as it allows correction of any hemodynamic sequelae, and simultaneous attainment of definite histological diagnosis for guidance of adjuvant therapy.

Keywords: Lymphoma; patent foramen ovale (PFO); cardiac tumor; hypoxemia

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Introduction

Patients with right-sided cardiac tumor rarely present with hypoxemia unless they are associated with pulmonary embolism. We report a case of cardiac diffuse large B-cell lymphoma (DLBCL) of the right atrium (RA) causing significant right-to-left shunting via a patent foramen ovale (PFO).

Case presentation

A 66-year-old man presented with dyspnea and intermittent palpitation for 1 week with no associated chest pain, leg swelling or dizziness. He had no recent long-haul flights. Arterial blood gas showed type I respiratory failure and increased alveolar-arterial gradient, despite a chest X-ray showing clear lung fields. ECG showed sinus rhythm with no ischemic changes or right heart strain pattern. Urgent contrast computed tomography (CT) scan of thorax showed a 6.5-cm

soft tissue density in RA (*Figure 1*) and no evidence of pulmonary embolism or parenchymal lung disease.

An emergency operation was performed. On-table transesophageal echocardiogram showed a 2-cm PFO with right to left shunt (*Figures 2-4*). There was a fungating right atrial tumor arose from the atrio-ventricular (AV) groove and extended to the right ventricle surface (*Figure 5A*). Cardiopulmonary bypass circuit was achieved via right femoral arterial and venous cannulation. A right atriotomy was made distant from the tumor margin. It was noted that the right atrial tumor arose from the AV groove with a 6-cm tumor bulk within the RA. There is no tumor invasion to the tricuspid valve, the vena cava, interatrial septum and coronary sinus. As the tumor involved the AV groove and was not feasible for complete tumor resection, only the intra-atrial part of the tumor was debulked (*Figure 5B*). The PFO was closed with pleggetted 3/0 polypropylene suture. The right atriotomy was closed followed by decannulation and successful weaning from cardiopulmonary bypass.

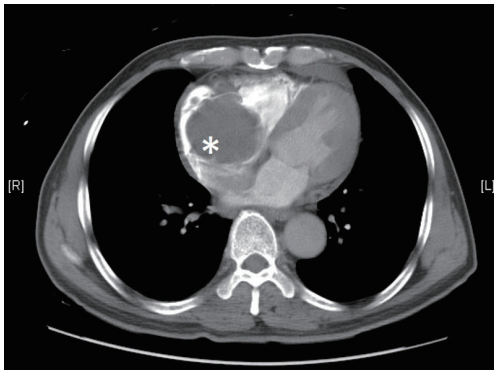


Figure 1 Computer tomography scan showing a huge mass occupying the right atrial cavity (asterisk).


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Video 1. Color Doppler in intra-operative trans-esophageal echocardiogram



Peter S. Y. Yu, Calvin S. H. Ng, Randolph H. L. Wong*, et al.
 Division of Cardiothoracic Surgery, Department of Surgery,
 The Chinese University of Hong Kong, Prince of Wales
 Hospital, Shatin, Hong Kong

Figure 2 Color Doppler in intra-operative trans-esophageal echocardiogram (1).

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Video 2. Bubble saline injection in intra-operative trans-esophageal echocardiogram



Peter S. Y. Yu, Calvin S. H. Ng, Randolph H. L. Wong*, et al.
 Division of Cardiothoracic Surgery, Department of Surgery,
 The Chinese University of Hong Kong, Prince of Wales
 Hospital, Shatin, Hong Kong

Figure 3 Bubble saline injection in intra-operative trans-esophageal echocardiogram (2).

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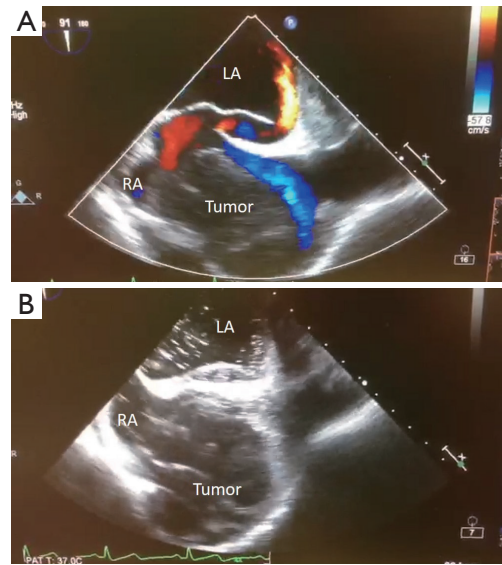


Figure 4 Intra-operative trans-esophageal echocardiogram showing the shunt from right atrium (RA) to the left atrium (LA) via a patent foramen ovale (PFO) demonstrated by color Doppler (A) and bubble saline injection (B).

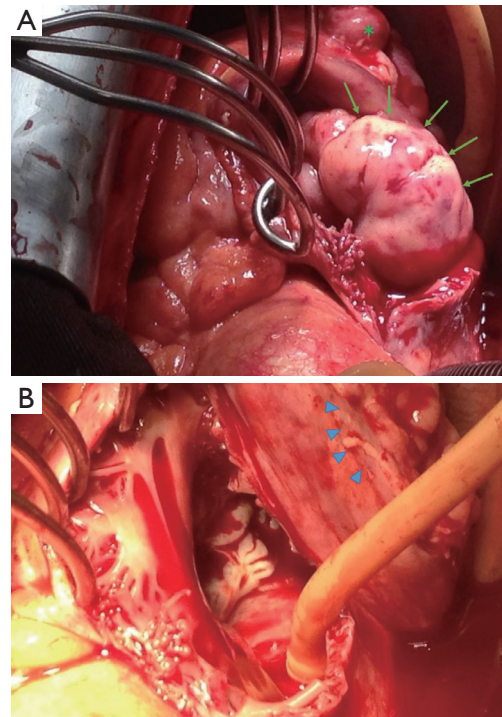


Figure 5 Intra-operative photo showing (A) the huge right atrial tumor (arrows) with extension into the right ventricle and invaded out of the right ventricular surface (asterisk), and (B) after gross debulking of the right atrial part of the tumor with residual tumor over the surface of the right ventricle (arrowheads).

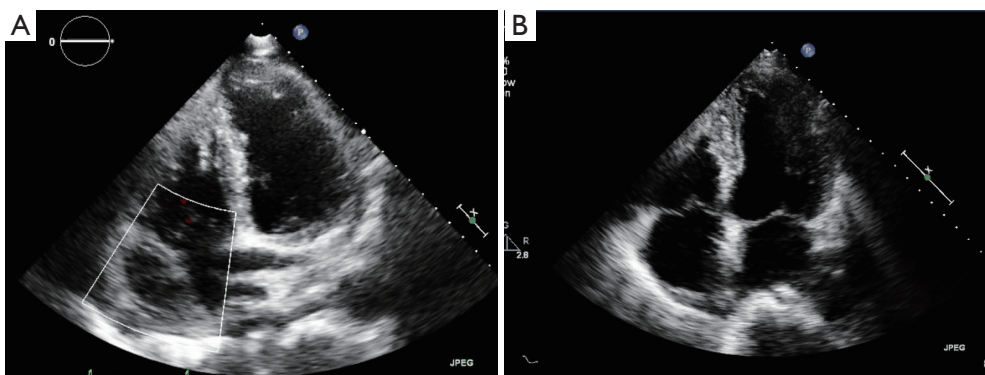


Figure 6 Echocardiogram 2 weeks after operation (A) showing the residual tumor mass in right atrium (RA), which (B) disappeared as shown in the scan 7 months after operation and 3 months after chemotherapy

Post-operative trans-esophageal echocardiogram showed no tumor mass within the right cardiac chambers and no residual intra-cardiac shunting. The total bypass time was 37 minutes, and the total operative time was 130 minutes.

Pathology of the right atrial mass showed DLBCL. Positron-emission tomography (PET) scan showed no lymphoma involvement in other organs. He was given chemotherapy (R-CHOP regimen) by Oncologist. Echocardiography 7 months after the operation showed no right atrial tumor, intra-cardiac shunt or pericardial effusion (Figure 6). He remained well 14 months after operation with no heart failure symptoms or lymphoma recurrence.

Discussion

Primary cardiac tumors had a wide range of presentations including dyspnea and palpitation. Type 1 respiratory failure, suggested by hypoxemia in blood gas analysis, was a rare encounter in right-sided cardiac tumor in the absence of embolism into the pulmonary circulation. For our patient, we postulate the hypoxemia was secondary to a forced right-to-left shunt from a huge space occupying tumor in the RA. A careful echocardiogram study is the key diagnostic tool for explaining the abnormalities.

Primary cardiac lymphoma comprises only a tiny proportion (1.3%) of all primary cardiac tumors (3). The commonest non-Hodgkin subtype was DLBCL (4). This subtype, like its extra-cardiac counterparts, was highly malignant and aggressive. Although primary cardiac malignant neoplasms were notoriously known to have poor prognosis, good clinical outcomes were commonly reported after treatments for cardiac lymphoma with overall response rate up to 79% and complete remission rate up to 59% (4). Although surgery alone

was not shown to improve survival (4,5), it palliates immediate hemodynamic disturbances, and allows definite histological diagnosis to guide oncological therapies.

Complete surgical resection is ideal but may not be feasible because of tumor extension or invasion. Moreover, it may not be a necessity for good survival. The longest survival of primary cardiac DLBCL after combined partial resection and chemotherapy was at least 13 years (6). It may be explained by the intrinsic high sensitivity of the tumor to cytotoxic agents, and further improved sensitivity after tumor debulking.

Conclusions

Hypoxemia is not a common presentation/association with right-sided cardiac tumors. Meticulous echocardiographic exam is necessary to look for any intracardiac shunts. Surgery is the single most important initial management in that it prevents or relieves any hemodynamically significant mechanical sequelae (e.g., obstruction, compression, embolism), and allows histological diagnosis which guides adjuvant therapy. Definite histological diagnosis is essential, as cardiac lymphoma is distinct from other primary cardiac malignancy in terms of a much better prognosis.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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