



# An enormous right atrial myxoma highlights the role of echocardiography in heart failure pathways: a case report

Sam Straw <sup>1,2</sup>, Anshuman Sengupta<sup>2</sup>, V. Kate Gantenby<sup>2</sup>, Dominik Schlosshan<sup>2</sup>, Antonella Ferrara<sup>3</sup>, and Klaus K. Witte <sup>1,4\*</sup>

<sup>1</sup>Leeds Institute of Cardiovascular and Metabolic Medicine, University of Leeds, Leeds, UK; <sup>2</sup>Department of Cardiology, Leeds Teaching Hospitals NHS Trust, Leeds, UK; <sup>3</sup>Department of Cardiothoracic Surgery, Leeds Teaching Hospitals NHS Trust, Leeds, UK; and <sup>4</sup> Department of Internal Medicine, RWTH Aachen University, Aachen, Germany

Received 14 September 2021; first decision 27 October 2021; accepted 25 January 2022; online publish-ahead-of-print 28 January 2022

## Background

Myxomata are rare, benign, primary tumours of the heart which can present with a variety of symptoms depending on size, location, and mobility. Here, we report a case of enormous right atrial myxoma, obliterating the right atrial and right ventricular cavities presenting with symptoms of heart failure.

## Case summary

A 66-year-old Caucasian female presented to primary care with symptoms of right heart failure and was found to have elevated N-terminal pro B-type natriuretic peptide of 2829 ng/L (normal value <125 ng/L). The patient was referred for urgent evaluation to the integrated heart failure service at our institution. Echocardiography revealed an enormous mobile mass attached to the right atrial septum, extending into the right ventricle and inferior vena cava measuring 90 × 42 mm. The patient underwent urgent surgical resection. Perioperative transoesophageal echocardiography demonstrated severe tricuspid regurgitation, which was treated with tricuspid annuloplasty ring. The patient made an uneventful recovery and was discharged. Subsequent imaging showed a reduction in right ventricular dimensions and improved systolic function.

## Discussion

This case serves to remind us of the critical role of echocardiography in the diagnosis and management of people with breathlessness and raised natriuretic peptides. Therapies for heart failure are guided by ejection fraction, therefore timely and accurate diagnosis is critical. Moreover, as in this case, echocardiography can also identify other features of critical relevance to patient care.

## Keywords

Myxoma • Tricuspid regurgitation • Cardiac tumour • Case report

## ESC Curriculum

2.2 Echocardiography • 6.1 Symptoms and signs of heart failure

## Learning points

- An enormous right atrial myxoma presented with symptoms of heart failure.
- Urgent surgical excision was undertaken due to the risk of sudden death.
- Residual tricuspid regurgitation caused by annular dilatation was successfully treated by annuloplasty ring implantation.

\* Corresponding author. Tel: +44 113 3926108, Email: [k.k.witte@leeds.ac.uk](mailto:k.k.witte@leeds.ac.uk)

Handling Editor: Piotr Nikodem Rudziński

Peer-reviewers: Jan Henzel; Christoph Sinning; Dan Octavian Nistor

Compliance Editor: Brett Sydney Bernstein

Supplementary Material Editor: Katharine Kott

© The Author(s) 2022. Published by Oxford University Press on behalf of the European Society of Cardiology.

This is an Open Access article distributed under the terms of the Creative Commons Attribution-NonCommercial License (<https://creativecommons.org/licenses/by-nc/4.0/>), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact [journals.permissions@oup.com](mailto:journals.permissions@oup.com)

## Introduction

Myxomata are rare, benign, primary tumours of the heart with an estimated incidence of 0.5 per million per year. Around three quarters of myxomata arise in the left atrial septum, are twice as common in females and usually occur in middle age.<sup>1</sup> In this report, we describe a case of massive right atrial myxoma, obliterating the right atrial and right ventricular cavities, presenting with symptoms of heart failure which was successfully managed by urgent surgical resection and tricuspid ring annuloplasty.

## Timeline

Day 1	Presents to primary care physician with symptoms of heart failure N-terminal pro B-type natriuretic peptide elevated Referred to integrated heart failure service
Day 10	Attends integrated heart failure service Echocardiogram reveals massive right atrial mass
Day 11	Computerized tomography imaging of chest, abdomen, and pelvis Coronary angiogram
Day 13	Resection of right atrial mass and tricuspid ring implantation
Day 17	Discharged from intensive care
Day 22	Discharged from hospital
Day 54	Echocardiogram showing improvement in right ventricular dimensions, no significant tricuspid regurgitation, and mildly impaired function

## Case presentation

A 66-year-old Caucasian woman presented to primary care with a 4-week history of breathlessness and ankle swelling. With the exception of hypertension, her past medical history was unremarkable. Initial investigations included measurement of N-terminal pro B-type natriuretic peptide (NT-proBNP), which was found to be elevated at 2829 ng/L (normal value <125 ng/L) and so she was referred for urgent evaluation by the integrated heart failure service at our institution.

The patient attended on Day 10 following presentation to primary care. Clinical observations were unremarkable, oxygen saturations were 98% on room air, respiratory rate 19, blood pressure 129/72 mmHg, and heart rate 86 beats/min. Physical examination revealed oedema to the mid shin, a grade III mid-diastolic murmur, raised jugular venous pressure, and clear lung fields.

The electrocardiogram showed normal sinus rhythm. Transthoracic echocardiography showed a large, mobile mass arising from the right atrium and extending into the right ventricle (*Figure 1*). The mass appeared to be attached to the intra-atrial septum,

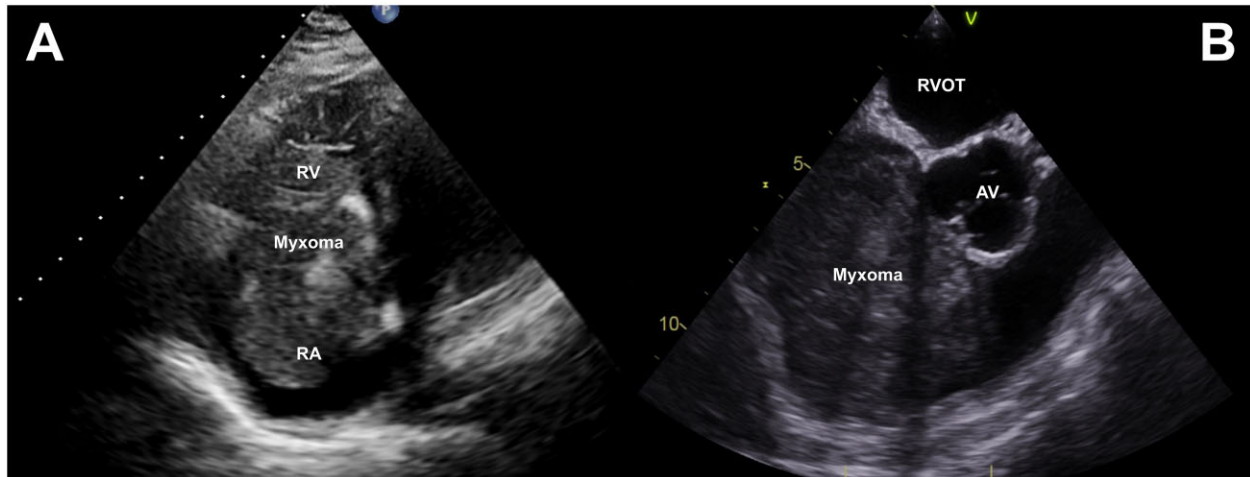
extending into the inferior vena cava in subcostal tilt view. The mass was described as echo-dense and heterogeneous, measuring 90 × 42 mm in apical four-chamber view. The tricuspid valve was stented open and so calculation of valve gradients was not possible, the basal right ventricular dimension was 43.5 mm and the tricuspid annular dimension was 49.7 mm. The inferior vena cava diameter measured 19.1 mm with minimal respiratory variation.

The patient was counselled on the differential diagnosis and in view of the risk of sudden death, was admitted directly from clinic to undergo urgent surgical resection. Prior to operation, further diagnostic evaluation was undertaken. Chest radiography showed a dilated right atrium and small left pleural effusion. Computerized tomography of the chest, abdomen, and pelvis showed an isolated mass within the right atrium extending into the right ventricle and superior vena cava, a small left basal pleural effusion, with no evidence of distant metastases or remote thromboembolism (*Figure 2*). Coronary angiography showed moderate, proximal atheroma in the left anterior descending artery, whilst the right and circumflex coronary arteries were normal.

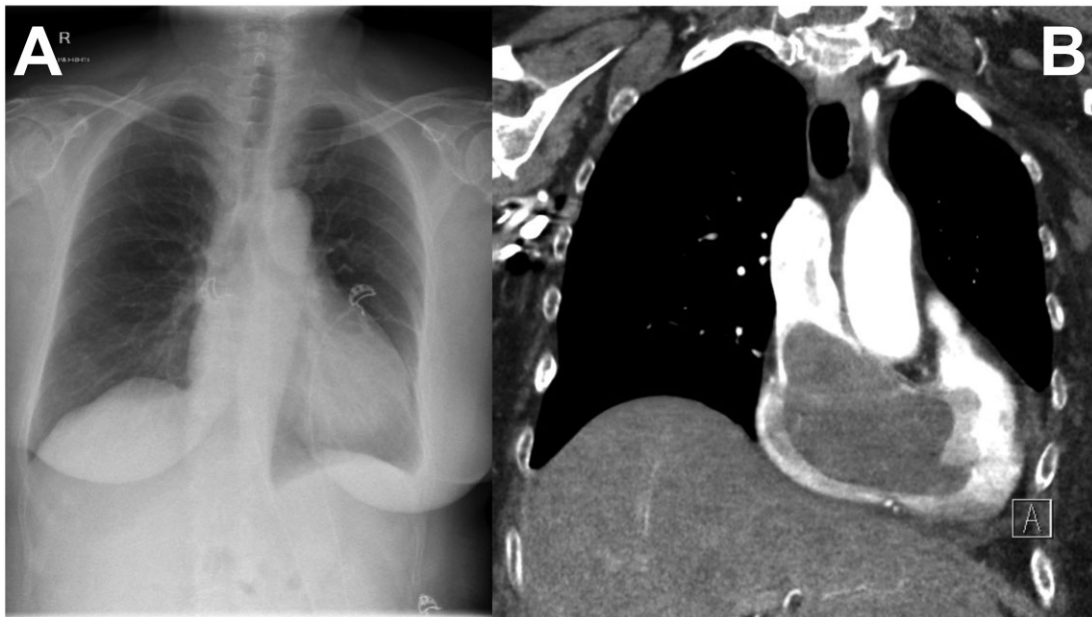
On Day 13, the patient underwent surgical resection, the pre-operative EuroSCORE II was 3.12%. The approach was via midline sternotomy, with systemic heparinization and standard aortic and bi-caval cannulation to achieve cardiopulmonary bypass with active cooling to 34°C. Following cross-clamping, the coronary arteries were perfused with cold blood cardioplegia delivered through the aortic root. Perioperative transoesophageal echocardiography again demonstrated the presence of an enormous right atrial myxoma obliterating the right atrial and ventricular cavities (*Video 1*). Right atriotomy revealed a massive right atrial mass, which was excised from the interatrial septum and removed in its entirety (*Figure 3*). The tricuspid leaflets were noted to be morphologically normal on visual inspection.

Following excision, and restoration of circulation, transoesophageal echocardiography showed severe tricuspid regurgitation due to annular dilatation (*Figure 4* and *Video 2*). Cardiopulmonary bypass was re-initiated and a 34 mm Physio II (Edwards Lifesciences, California, US) tricuspid ring was implanted. Subsequent imaging showed a central jet of mild tricuspid regurgitation and a dilated and moderately impaired right ventricle (*Video 3*). The patient was transferred to the intensive care unit, the postoperative course was uneventful, and she was discharged to her usual place of residence on Day 22.

Histopathological examination demonstrated the mass was composed of myxoid substance with stellate myxoma (lipidic) cells and variably sized vessels. There were areas of haemorrhage, both recent and remote, and scattered calcification. A small focus of glandular elements was identified (and highlighted by CK7 immunohistochemistry) (*Figure 5*), which is an uncommon but benign feature reported in the literature.<sup>2</sup> On Day 45, the patient attended for transthoracic echocardiography. There was evidence of reverse remodelling, with a reduction in right ventricular dimensions to within normal limits (34 mm). The right ventricular function was improved, reported as mildly impaired (tricuspid annular plan systolic excursion 9 mm), and only trivial tricuspid regurgitation (insufficient envelope to estimate pulmonary artery systolic pressure). No cardiovascular signs or symptoms were present.



**Figure 1** (A) Modified four-chamber transthoracic echocardiogram. (B) Short-axis transoesophageal echocardiogram showing a large, mobile mass arising from the right atrium and extending into the right ventricle.



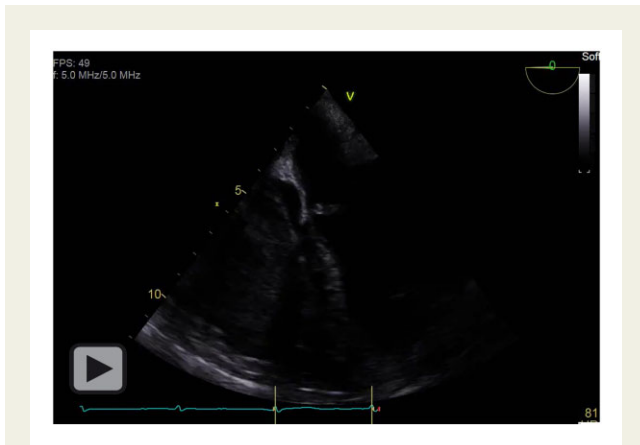
**Figure 2** (A) Chest radiograph showing a dilated right atrium and small left pleural effusion and (B) coronal plane computerized tomography of the chest showing right atrial mass obliterating right atrial and right ventricular cavities.

## Discussion

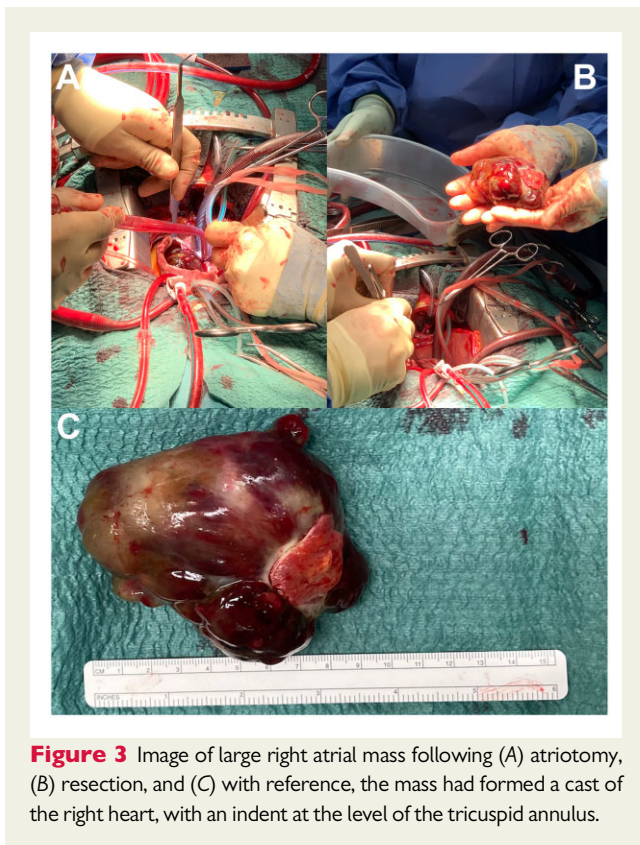
This case report described an enormous right atrial myxoma obliterating the right atrial and right ventricular cavities, resulting in symptoms of heart failure. The differential diagnosis for an intracardiac mass found during echocardiography includes primary cardiac tumours, metastatic disease, and thrombi.<sup>1</sup> Although myxomata are rare, they account for more than half of primary cardiac tumours. Other benign tumours include papillary fibroelastomas and lipomas,

whilst malignant tumours include pleomorphic sarcomas, angiosarcomas, and leiomyosarcomas.<sup>3</sup>

Myxoma may occur in any chamber of the heart and presenting symptoms vary depending on the size, location, and mobility of the tumour. Constitutional symptoms are common, including fever, weight loss or fatigue, as well as symptoms secondary to embolization. Right-sided myxomata tend to present later than left-sided tumours, due to the non-specific and benign symptoms at an early stage,<sup>4</sup> and may therefore present with invasive features due to their



**Video I** Perioperative transoesophageal echocardiography demonstrating a right atrial myxoma.



**Figure 3** Image of large right atrial mass following (A) atriotomy, (B) resection, and (C) with reference, the mass had formed a cast of the right heart, with an indent at the level of the tricuspid annulus.

location and size.<sup>5</sup> Often, however, myxomata are discovered incidentally for imaging performed for other indications.<sup>6</sup> In the present case, the mass was very large at the time of presentation, with symptoms only present for a short time prior to this.

Outcome data on myxomata are limited to case reports and small series. The optimal timing of surgery has not been evaluated in a systematic manner. However, death whilst awaiting operation has been reported and so it is logical that especially in the setting of possible

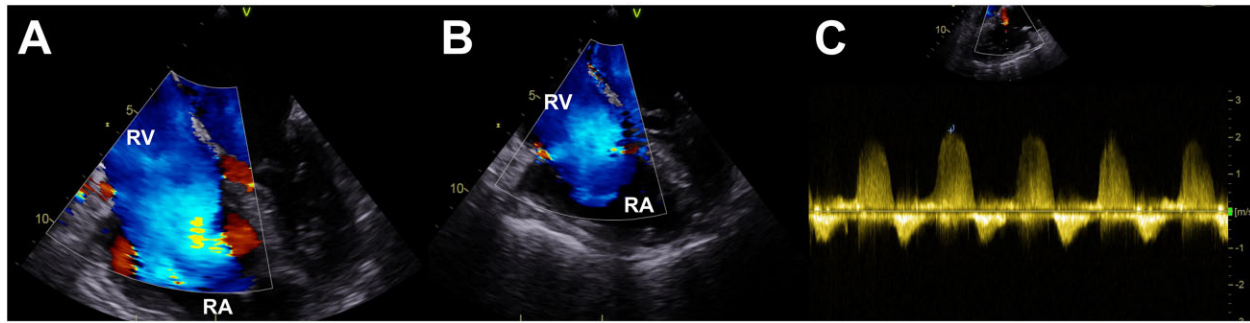
outflow tract obstruction, surgical resection should be undertaken as a matter of urgency. In this case, due to the size of the mass and the risks of sudden death due to right ventricular outflow tract obstruction, urgent surgical resection was recommended. Complete surgical resection including a sufficient margin is vital, due to reported recurrence rates of 3.3% in the event of incomplete excision.<sup>1</sup> However, following complete excision, the long-term prognosis is similar to the general population.<sup>7</sup>

This case was complicated by severe tricuspid regurgitation due to annular dilatation found on perioperative imaging. This was treated by annuloplasty ring implantation, which led to acute haemodynamic improvement, with only mild residual tricuspid regurgitation. Whilst leaflet dysfunction due to retraction, restriction, and atrophy probably due to prolonged pressure effects exerted by the mass requiring bioprosthetic valve implantation has been described,<sup>8</sup> most reports describe that the valve leaflets themselves are normal, with favourable effects of annuloplasty ring implantation.<sup>9</sup> Subsequent follow-up imaging in this case demonstrated right ventricular remodelling following correction of tricuspid regurgitation, accompanied by resolution of symptoms.

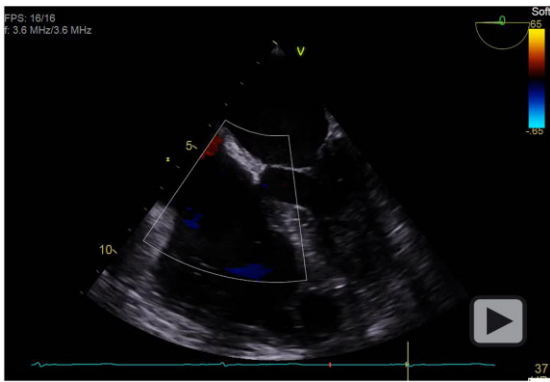
Guidelines recommend that patients presenting to primary care with signs and symptoms of heart failure are screened by testing serum NT-proBNP prior to echocardiography.<sup>10</sup> N-terminal pro B-type natriuretic peptide is a precursor to the natriuretic peptide BNP, which is elevated in response to ventricular stretch or wall tension and can stratify the pre-test probability prior to echocardiography. Low levels (<125 pg/L) allow the exclusion of important cardiac dysfunction and suggest further evaluation is not required, whereas very high levels ( $\geq 2000$  pg/L) are associated with the greatest risk of heart failure (positive predictive value around 50%) and a worse prognosis.<sup>10</sup> In the UK, national recommendations are for patients with very high levels to undergo evaluation by a specialist within 14 days.<sup>11</sup>

This case serves to remind us of the critical role of echocardiography in the diagnosis and management of people with symptoms of heart failure and raised natriuretic peptides. Echocardiography remains the most widely available imaging modality and can identify and quantify ventricular systolic and diastolic dysfunction, assess valvular function, and cardiac structure. Although cardiac magnetic resonance can provide more accurate assessment of cardiac geometry and function and avoids the pitfalls of inter-observer variability of echocardiography, it is not acceptable to all patients, contraindicated in some and less readily available, risking delays to diagnosis. Furthermore, therapies for heart failure are guided by ejection fraction, therefore timely and accurate diagnosis are critical. Often overlooked is that echocardiography cannot only *exclude* heart failure but it can also identify additional abnormalities including cardiac tumours providing diagnostic information, as well as accurately determining the size, mobility (and therefore risk of embolization), site of origin, and information on valvular dysfunction. Although definitive diagnosis requires histological examination, multi-modality imaging of an intra-cardiac mass (for instance by cardiac magnetic resonance imaging) is not required to guide management or to plan the operation, although cross-sectional imaging to rule out distant metastases is advisable.

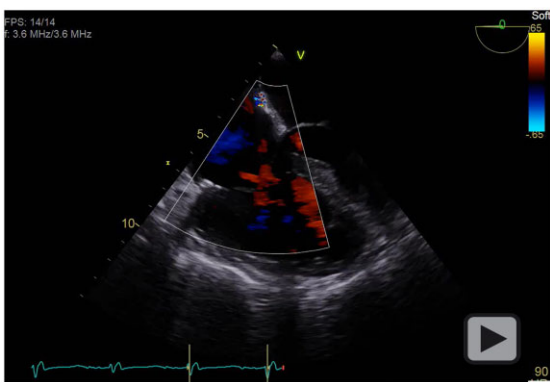




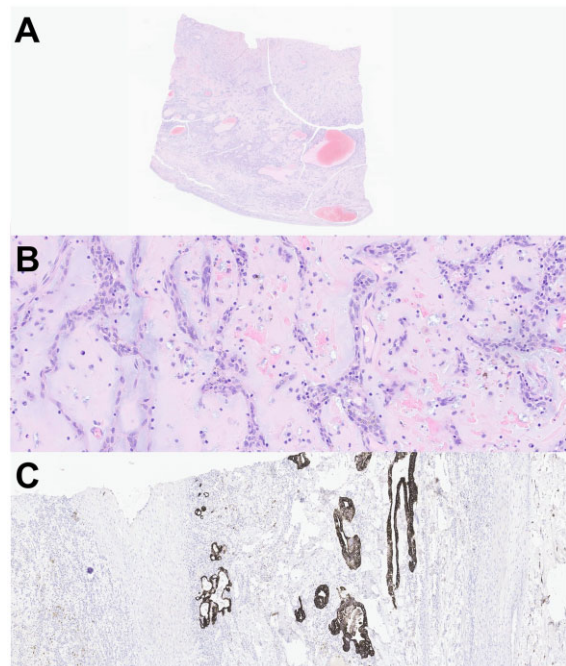
**Figure 4** (A) Two-dimensional transoesophageal echocardiography following resection showing annular dilatation and severe tricuspid regurgitation and (B) following tricuspid annuloplasty showing reduction in annular dimensions and (C) mild tricuspid regurgitation on continuous wave doppler.



**Video 2** Perioperative transoesophageal echocardiography demonstrating severe tricuspid regurgitation due to annular dilatation following excision.



**Video 3** Perioperative transoesophageal echocardiography demonstrating mild residual tricuspid regurgitation following ring annuloplasty.



**Figure 5** Histopathology imaging of myxoma following surgical resection showing (A) low power morphology, (B) lipidic cells within myxoid stroma, and (C) glandular elements highlighted by CK7 immunohistochemistry.

## Conclusion

We have described a case of massive right atrial myxoma resulting in haemodynamic compromise which was treated by urgent surgical resection. This case highlights the critical role of echocardiography in BNP-guided pathways, the role of urgent surgical excision in atrial

myxoma and of tricuspid annuloplasty ring for managing residual valvular regurgitation.

## Lead author biography



Dr Sam Straw undertook his medical training at the University of Leeds, obtaining degrees in Medicine and Surgery (MBChB Hons) and Clinical Sciences (BSc Hons). His current role is as a British Heart Foundation Clinical Research Training Fellow at the University of Leeds and is undertaking a PhD investigating the benefits of personalised pacemaker programming in heart failure.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

## Acknowledgements

The authors would like to thank Dr Foteini Malta, Dr Leslie Davidson, and Dr Kathryn J. Griffin for their help with preparation and reporting of histological images and to Dr Michael Drozd for help with manuscript preparation.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** S.S. is supported by the British Heart Foundation. K.K.W. has received speaker's fees and honoraria from Medtronic, Cardiac Dimensions, Novartis, Abbott, BMS, Pfizer, Bayer and has received an unconditional research grant from Medtronic. None of the other authors have conflicts of interest to declare.

**Funding:** This study was supported by a British Heart Foundation Clinical Research Training Fellowship awarded to S.S. (FS/CRTF/20/24071).

## References

1. Karabinis A, Samanidis G, Khoury M, Stavridis G, Perreas K. Clinical presentation and treatment of cardiac myxoma in 153 patients. *Medicine (Baltimore)* 2018;**97**: e12397.
2. Pucci A, Gagliardotto P, Zanini C, Pansini S, di Summa M, Mollo F. Histopathologic and clinical characterization of cardiac myxoma: review of 53 cases from a single institution. *Am Heart J* 2000;**140**:134–138.
3. Basso C, Rizzo S, Valente M, Thiene G. Cardiac masses and tumours. *Heart* 2016;**102**:1230–1245.
4. Khan H, Chaubey S, Uzzaman MM, Iqbal Y, Khan F, Butt S et al. Clinical presentation of atrial myxomas does it differ in left or right sided tumor? *Int J Health Sci (Qassim)* 2018;**12**:59–63.
5. Rudzinski PN, Lubiszewska B, Rozanski J, Michalowska I, Kruk M, Kepka C et al. Giant intrapericardial myxoma adjacent to the left main coronary artery. *Front Oncol* 2018;**8**:540.
6. Yuan SM, Yan SL, Wu N. Unusual aspects of cardiac myxoma. *Anatol J Cardiol* 2017;**17**:241–247.
7. Elbardissi AW, Dearani JA, Daly RC, Mullany CJ, Orszulak TA, Puga FJ et al. Survival after resection of primary cardiac tumors: a 48-year experience. *Circulation* 2008;**118**:S7–S15.
8. Chhabra A, McClung JA, Kalapatapu S, Lafaro RJ, Fallon JT 3rd, Aronow WS. Giant myxoma causing heart failure symptoms. *Am J Case Rep* 2012;**13**:29–32.
9. Schiele S, Maurer SJ, P, Salvador CV, Ivanova K, Weirich G, Meierhofer, C et al. Left atrial myxoma. *Circ Cardiovasc Imaging* 2019;**12**:e008820.
10. McDonagh TA, Metra M, Adamo M, Gardner RS, Baumbach A, Bohm M et al. 2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure. *Eur Heart J* 2021;**42**:3599–3726.
11. NICE. *Chronic Heart Failure in Adults Quality Standard [QS9]*. London, UK: National Institute of Health and Care Excellence; 2011.