

A case report: cardiac dysphagia—a ghost of the past?

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Background

Rheumatic heart disease has become rare in developed countries and physicians have grown unfamiliar with the disease and its clinical course. The mitral valve is most commonly affected leading to mitral regurgitation and/or stenosis. The chronic volume and/or pressure overload leads to atrial remodelling and enlargement, driving the development of atrial fibrillation and thrombo-embolic events.

Case summary

A 87-year-old patient with a history of rheumatic mitral stenosis and mitral valve replacement was admitted to the neurology department for vertigo. A stroke was suspected, and she underwent a transoesophageal echocardiogram (TOE) which was complicated by dysphagia. Oesophageal manometry and computed tomography revealed oesophago-gastric junction outflow obstruction due to extrinsic compression by a giant left atrium (GLA).

Discussion

Dysphagia due to a GLA is rare. Various diagnostic criteria exist and the prevalence thus depends on which criterion is used. It is mostly encountered in rheumatic mitral disease, although there are reports of non-rheumatic aetiology. When the left atrium assumes giant proportions it can compress adjacent intrathoracic structures. Compression of the oesophagus can lead to dysphagia, as in our case. A TOE in these cases is relatively contraindicated and should only be performed if there is considerable reason to believe that it may change patient management.

Keywords

Rheumatic heart disease • Mitral valve stenosis • Giant left atrium • Dysphagia • Transoesophageal echocardiography • Case report

Learning points

- Rheumatic heart disease has become rare in developed countries and the younger generation of physicians have grown unfamiliar with the disease and its clinical course.
- A giant left atrium (GLA) is rare and mainly associated with rheumatic heart disease. It can compress adjacent intrathoracic structures.
- Although perhaps self-evident, swallowing difficulties should always be inquired before performing a transoesophageal echocardiography; even in the absence of stroke and upper gastro-intestinal tract cancer or surgery.
- A transoesophageal echocardiogram is relatively contraindicated in cases of the GLA and should only be performed if it may change patient management.

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Introduction

Rheumatic mitral stenosis has become rare in developed countries due to a marked reduction in the incidence of rheumatic fever. Physicians have grown unfamiliar with the natural history of the disease. However, it remains a major cause of cardiovascular morbidity and mortality in developing countries.¹

Acute rheumatic fever is the result of an abnormal immune response to group A *Streptococcus* (e.g. *Streptococcus pyogenes*) manifesting about 3 weeks after the acute infection. It can affect joints, skin, brain, and heart. These features are outlined in the Jones criteria.² The prominent manifestation of carditis is valvulitis, predominantly of the mitral and aortic valve, causing regurgitation, and/or stenosis. The valvular dysfunction can be severe at diagnosis, requiring early intervention. Often, it is mild or moderate at diagnosis and progressive or regressive over the years.³ It is not uncommon that a preceding episode of rheumatic fever cannot be recalled.

Rheumatic mitral stenosis obstructs blood flow from the left atrium to the left ventricle, raising pressure upstream (left atrium, pulmonary vasculature, and right ventricle) and manifests as shortness of breath. The chronic pressure overload leads to atrial remodelling and enlargement, driving the development of atrial fibrillation, and thrombo-embolic events.

In this article, we report the case of dysphagia due to a giant left atrium (GLA) as a long-term complication of rheumatic mitral stenosis.

Timeline

1954	Diagnosis of rheumatic mitral stenosis for which she was treated with closed commissurotomy.
1955	Permanent atrial fibrillation.
1955–2000	Progressive mitral valve stenosis and tricuspid regurgitation.
2000	Mechanical mitral valve replacement for mitral stenosis and tricuspid valve repair with ring annuloplasty.
2011	Left-sided breast cancer for which she was treated with mastectomy and lymph node removal; no radiotherapy or chemotherapy.
August 2020	Admission to the neurology department for vertigo; a stroke is suspected. <ul style="list-style-type: none"> Day 1: Computed tomography scan is negative; adequate international normalized ratio values. Day 2: Magnetic resonance imaging brain does not show recent cerebral ischaemia. <ul style="list-style-type: none"> Day 6: Transthoracic echocardiography. Day 8: Transoesophageal echocardiography complicated by dysphagia.

Case presentation

An 87-year-old Caucasian female patient, born in Europe, was admitted to the neurology department with a complaint of vertigo. A stroke was suspected and she received a standard stroke assessment, including a cardiac evaluation given the extensive cardiac history. In 1954, at the age of 21, she underwent a closed mitral commissurotomy for rheumatic mitral stenosis. A year later she developed atrial fibrillation. In 2000, at the age of 67, she underwent mechanical mitral valve replacement (ATS M29; ATS Medical, Inc., Minneapolis, MN, USA) for mitral re-stenosis. Tricuspid valve repair with a ring annuloplasty (CE 34 mm) was performed at the same time. In 2011, she was treated for left-sided breast cancer with mastectomy and lymph node removal; no radiotherapy or chemotherapy. During the last decade, she had been followed for moderate degenerative aortic stenosis (AS), which was felt to be likely non-rheumatic in origin as it had manifested at old age.

She had been taking a vitamin K antagonist for many years with stable international normalized ratio (INR) values. Upon admission, the INR was in the therapeutic range. Chest X-ray showed cardiomegaly (Figure 1). The cardiothoracic ratio was 0.87. Transthoracic echocardiogram showed the normal systolic function of the hypertrophied left ventricle and normal functioning of the mitral valve prosthesis. The peak and mean trans-mitral gradient were 22 and 4.3 mmHg, respectively. There was no pathologic mitral regurgitation. There was a residual moderate tricuspid regurgitation. The aortic stenosis had progressed to a severe stenosis with a mean gradient of 40 mmHg. Both atria were vastly dilated, the left atrial anterior–posterior (AP) diameter measured 8 cm (Figure 2A,B). Single-plane apical four-chamber left and right atrial volume were 211 mL (143 mL/m²) and 187 mL (127 mL/m²), respectively.

A transoesophageal echocardiogram (TOE) excluded intracardiac thrombi and confirmed normal functioning of the mitral valve prosthesis. The mean trans-mitral gradient was 5 mmHg. There were closing jets but no pathologic valvular or paravalvular jets. Immediately after TOE, the patient developed dysphagia for solid foods. She lost weight due to poor oral intake. A proton-pump

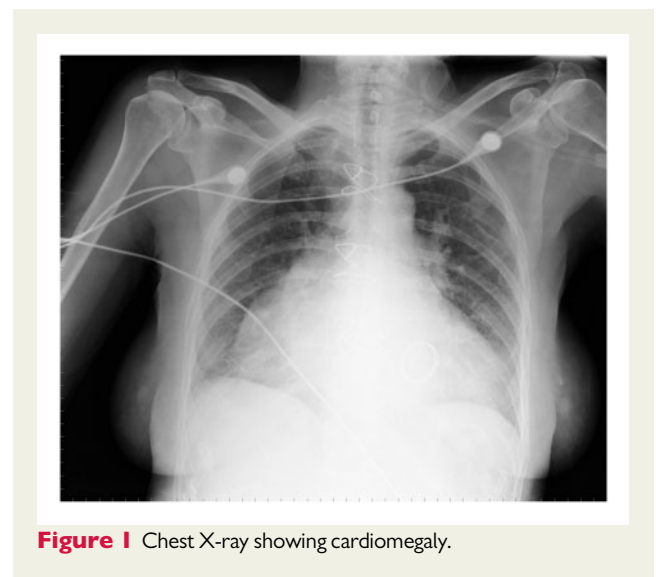


Figure 1 Chest X-ray showing cardiomegaly.

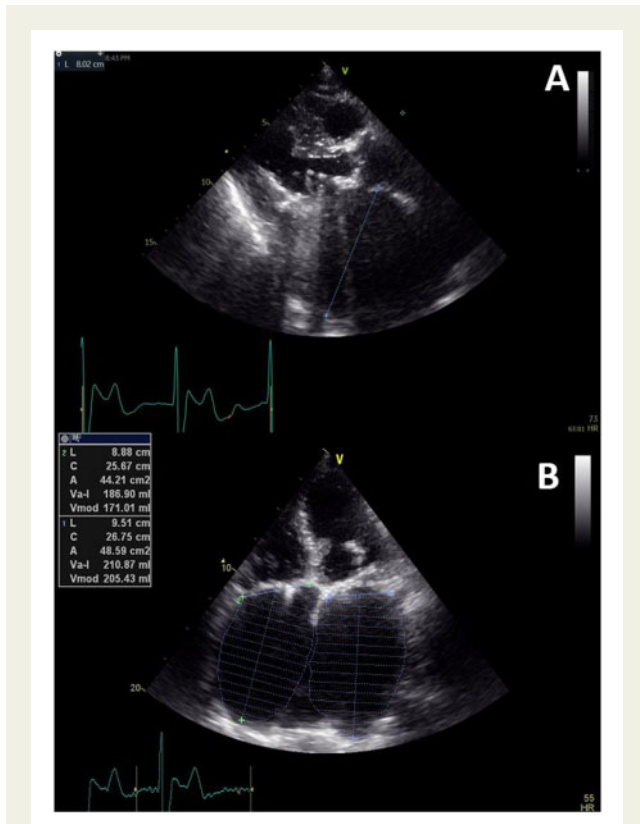


Figure 2 Transthoracic echocardiography showing extremely dilated atria. (A) Left atrial anterior–posterior diameter measures 8 cm in parasternal long axis. (B) Single-plane apical four-chamber left and right atrial volume are 211 mL (143 mL/m²) and 187 mL (127 mL/m²), respectively.

inhibitor was prescribed, and she was put on a mixed/liquid diet. Apparently, the patient had difficulty swallowing for some time. Her physician was unaware, and TOE had made it significantly worse. An unfortunate complication, all the more so since magnetic resonance imaging (MRI) of the brain was negative for recent cerebral ischaemia. Ultimately a peripheral vestibular disorder was suspected.

A barium swallow suggested achalasia (Figure 3), though oesophageal manometry rebutted this and favoured a diagnosis of oesophageal gastric outflow obstruction due to extrinsic compression. Oesophageal peristalsis was disturbed and resting lower oesophageal sphincter pressure and median integrated relaxation pressure were elevated, 90 and 43.5 mmHg, respectively. Peristalsis in the distal oesophagus was preserved, which is atypical for achalasia. During her hospital stay, a work up for transcatheter aortic valve replacement (TAVR) was initiated and a cardiac computed tomography (CT) angiography of the thorax was performed as part of the pre-TAVR protocol. Computed tomography angiography demonstrated extrinsic compression of the distal oesophagus by a massively dilated left atrium (Figure 4).

During her hospital stay, the dysphagia spontaneously regressed reaching its former level at discharge. Left atrial reduction surgery, as an attempt to alleviate the obstructive symptoms, was not considered due to age and favourable spontaneous evolution. After a

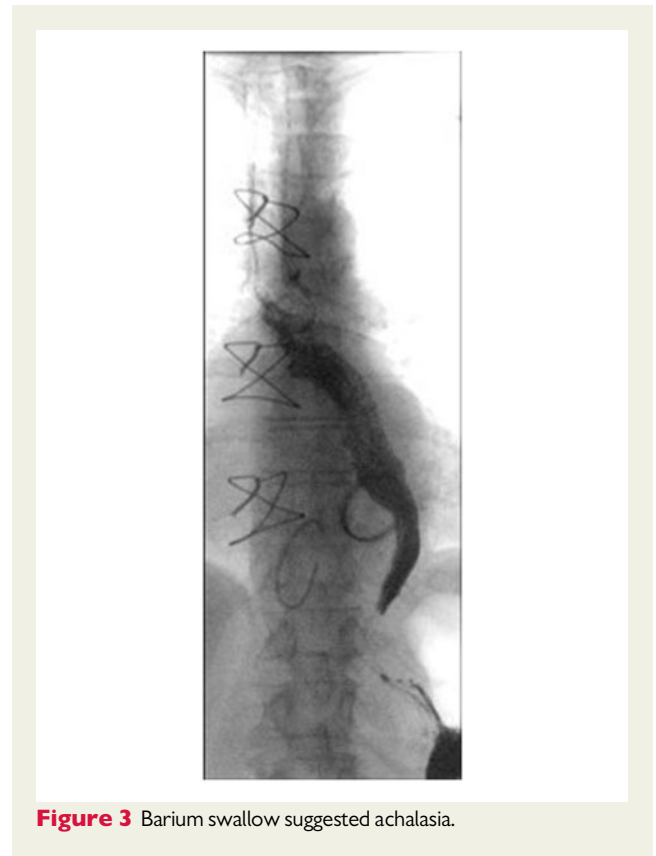


Figure 3 Barium swallow suggested achalasia.

multidisciplinary Heart Team discussion, a decision was made not to proceed with TAVR, given the paucisymptomatic clinical status, difficult vascular approach, and no expected benefit on quality of life. This decision was in line with the European Society of Cardiology guidelines for management of valvular heart disease.⁴

At a 6-month follow-up, the patient was in New York Heart Association Class II. She did not experience angina or syncope. The dysphagia had significantly improved and she had gained weight. Vertigo had not reoccurred.

Discussion

Rheumatic heart disease has become rare in developed countries, and the younger generation of physicians have grown unfamiliar with the disease and its clinical course.⁵ We presented a case of dysphagia due to GLA as a long-term complication of rheumatic mitral stenosis.

The mitral valve is the most commonly affected valve, manifesting as mitral regurgitation and/or stenosis. Both impose an overload (volume and pressure overload, respectively) to the left atrium, leading to atrial remodelling and enlargement. Some authors suggest a contribution of rheumatic inflammation ('pancarditis') to this process.⁶ The enlargement can be quite impressive, sometimes reaching up to the right lateral chest wall disguising as a right-sided pleural effusion.⁷

Various diagnostic criteria for GLA circulate due to advances in diagnostic techniques, as is often the case in medicine. A cardiothoracic ratio on chest X-ray of >0.7 is suggestive of GLA but lacks specificity.⁸ Giant left atrium has been defined in the literature based on

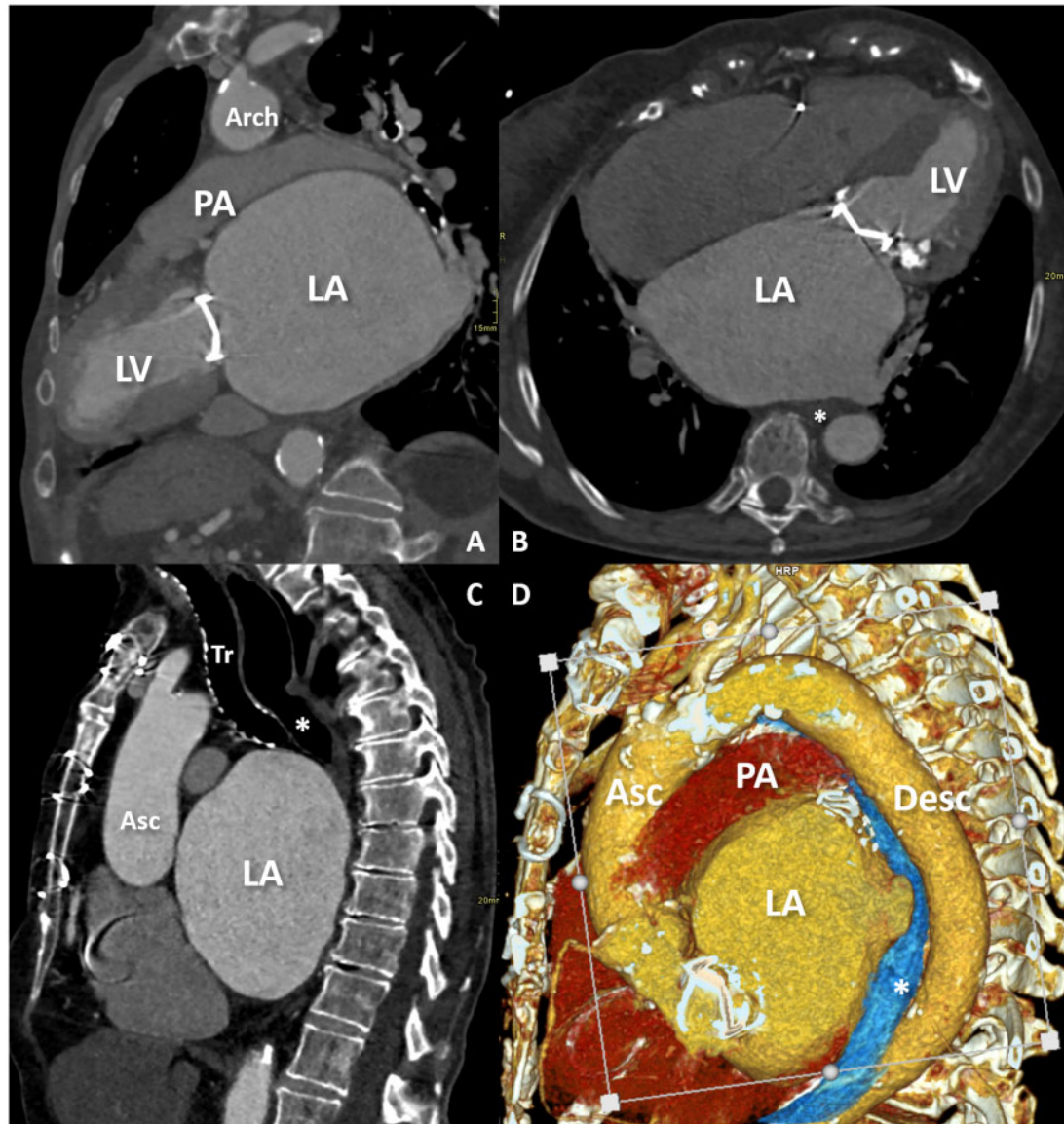


Figure 4 Computed tomography angiography of the thorax, (A) vertical long axis reconstruction, (B) four-chamber reconstruction, and (C) sagittal reconstruction showing massively enlarged left atrium, mechanical mitral valve and tricuspid valve ring. The oesophagus (*) is compressed between left atrium and descending aorta. (D) A virtual rendering technique reconstruction with an obliquely cut-away left hemi-thorax. Arch, aortic arch; Asc, ascending aorta; Desc, descending aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery.

linear AP diameter in the parasternal long-axis view, with definitions varying from 6.5 to 8 cm.^{9,10} Left atrial enlargement is often asymmetric. Relying on a single 2D parameter can lead to underestimation of true left atrial size. Although it is unlikely that when the left atrium massively dilates, it would do so in only one dimension. One could theorize that the left atrial volume (index) measurement would be the optimum echocardiographic criterion for GLA and a new definition should be considered.¹¹

In the literature, GLA is mainly reported in rheumatic heart disease. Rarely, it is associated with non-rheumatic disease.⁹ The left atrium is the most posteriorly and superiorly located of the cardiac chambers. It lays in close proximity to the descending aorta, tracheal bifurcation,

main bronchi, lungs, left recurrent laryngeal nerve, and oesophagus.¹²

This close relationship to the oesophagus is exploited during TOE and feared during atrial fibrillation ablation. When the left atrium massively dilates, it can compress the neighbouring structures leading to debilitating symptoms. Giant left atrium can splay the tracheal carina, perceivable as a widened carinal angle on chest X-ray.¹³ There are reports of partial to total lung collapse due to direct parenchymal compression or compression of a main bronchus.¹⁴ Compression of the left recurrent laryngeal nerve can lead to hoarseness of voice, otherwise known as Ortner's syndrome.¹⁵ Compression of the oesophagus can lead to dysphagia, as in our case. Decompensated heart failure with volume overload can worsen the dysphagia.

Endoscopy and barium oesophagography (barium swallow) are first-line tests in the evaluation of oesophageal dysphagia. Barium oesophagography may be performed as the initial test to map the oesophagus when the patient is judged to be at high risk of endoscopy complications. Endoscopy and barium oesophagography can both demonstrate external compression of the oesophagus, though cross-sectional imaging may be necessary for a more definitive diagnosis. Computed tomography is the preferred technique. Magnetic resonance imaging is a good alternative in case of contraindications for CT (or iodinated contrast). Compression of other intrathoracic structures can also be evaluated by CT or MRI.^{16,17}

Cardiac dysphagia—extrinsic compression of the oesophagus by a cardiovascular structure (e.g. aorta aneurysm, GLA, etc.)—is rare but should be kept in mind. It is a well-known fact among cardiologists that dysphagia is a relative contraindication for performing TOE. However, we are mainly vigilant for this contraindication in the context of stroke or upper gastro-intestinal tract cancer or surgery. We are less vigilant for dysphagia in patients lacking these clinical conditions. If dysphagia is not systematically questioned in all patients, rare causes of dysphagia, as is cardiac dysphagia, can be missed during the pre-procedural screening and TOE can worsen the pre-existing dysphagia in these patients.

We presented this case to raise awareness for this clinical entity that has become rare due to a marked reduction in the incidence of rheumatic fever in developed countries. Transoesophageal echocardiogram is relatively contraindicated in these cases and should only be performed if there is considerable reason to believe that it may significantly alter patient management.¹⁸

Lead author biography



Dr Céline Deschepper graduated in 2015 at the medical school of the Catholic University of Leuven and is currently a cardiology resident at the Ghent University Hospital.

Supplementary material

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

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

Consent

The authors confirm that 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: None declared.

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References

1. Quinn RW. Comprehensive review of morbidity and mortality trends for rheumatic fever, streptococcal disease, and scarlet fever: the decline of rheumatic fever. *Rev Infect Dis* 1989;**11**:928–953.
2. Gewitz MH, Baltimore RS, Tani LY, Sable CA, Shulman ST, Carapetis J et al.; American Heart Association Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young. Revision of the Jones Criteria for the diagnosis of acute rheumatic fever in the era of Doppler echocardiography: a scientific statement from the American Heart Association. *Circulation* 2015;**131**:1806–1818.
3. Cannon J, Roberts K, Milne C, Carapetis JR. Rheumatic heart disease severity, progression and outcomes: a multi-state model. *J Am Heart Assoc* 2017;**6**:e003498.
4. Baumgartner H, Falk V, Bax JJ, De Bonis M, Hamm C, Holm PJ et al.; ESC Scientific Document Group. 2017 ESC/EACTS Guidelines for the management of valvular heart disease. *Eur Heart J* 2017;**38**:2739–2791.
5. Watkins DA, Johnson CO, Colquhoun SM, Karthikeyan G, Beaton A, Bukhman G et al. Global, regional, and national burden of rheumatic heart disease, 1990–2015. *N Engl J Med* 2017;**377**:713–722.
6. Hurst JW. Memories of patients with a giant left atrium. *Circulation* 2001;**104**:2630–2631.
7. Dixit A, Shariff R, Yin P, Rosenberg L, Vapnyar V. Giant left atrium disguised as right-sided pleural effusion. *CASE (Phila)* 2018;**2**:276–278.
8. Piccoli GP, Massini C, Di Eusano G, Ballerini L, Iacobone G, Soro A et al. Giant left atrium and mitral valve disease: early and late results of surgical treatment in 40 cases. *J Cardiovasc Surg (Torino)* 1984;**25**:328–336.
9. El Maghraby A, Hajar R. Giant left atrium: a review. *Heart Views* 2012;**13**:46–52.
10. Oh JK. Echocardiographic evaluation of morphological and hemodynamic significance of giant left atrium. An important lesson. *Circulation* 1992;**86**:328–330.
11. Lang R, Badano LG, Mor-Avi V, Afilalo J, Armstrong A, Ernande L, et al. Recommendations for cardiac chamber quantification by echocardiography in adults: an update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. *Eur Heart J Cardiovasc Imaging* 2015;**16**:233–271.
12. Ho SY, Cabrera JA, Sanchez-Quintana D. Left atrial anatomy revisited. *Circ Arrhythm Electrophysiol* 2012;**5**:220–228.
13. Murray JG, Brown AL, Anagnostou EA, Senior R. Widening of the tracheal bifurcation on chest radiographs: value as a sign of left atrial enlargement. *Am J Roentgenol* 1995;**164**:1089–1092.
14. Tanita A, Hosokawa Y, Tomiyama T, Kato M, Matsuda J, Sawai K et al. Giant left atrium due to mitral stenosis with massive atelectasis: a successful case with perioperative approach. *Int J Cardiol* 2013;**163**:e23–5.
15. Kheok SW, Salkade PR, Bangaragiri A, Koh NSY, Chen RC. Cardiovascular hoarseness (Ortner's syndrome): a pictorial review. *Curr Probl Diagn Radiol* 2021;**50**:749–754.
16. Carucci LR, Turner MA. Dysphagia revisited: common and unusual causes. *Radiographics* 2015;**35**:105–122.
17. Liu LWC, Andrews CN, Armstrong D, Diamant N, Jaffer N, Lazarescu A et al. Clinical practice guidelines for the assessment of uninvestigated esophageal dysphagia. *J Can Assoc Gastroenterol* 2018;**1**:5–19.
18. Matta A, Elenzi K, Carrié D, Roncalli J. Dysphagia as an early sign of cardiac decompensation in elderly: case report. *Eur Heart J Case Rep* 2020;**17**:1–5.