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CASE REPORT
Other

Aortic coarctation with cardiac fibroma in a young patient: a case report

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Background Cardiac fibroma and aortic coarctation are rarely observed concomitantly in the same patient. We report a case of cardiac fibroma with aortic coarctation treated with a hybrid surgical procedure. To the best of our knowledge, this is the first case of these two abnormalities existing in one patient.

Case summary A 22-year-old female patient visited the clinic with a 10-year history of hypertension. Physical examination revealed blood pressure of the upper extremities 50 mmHg higher than that of the lower extremities. Computed tomography angiography revealed a post-ductal-type aortic coarctation at the beginning segment of the descending aorta along with a 7.7 cm × 5.1 cm left ventricular mass. Transthoracic echocardiogram showed a mass at the middle segments of the lateral wall and apex and posterior wall of the left ventricle. Cardiac magnetic resonance imaging also showed the mass with hypointense signal on T1, hyperintense signal on T2, and intense signal on late gadolinium enhancement. No evidences of metastatic lesions were observed on ^{18}F-fluorodeoxyglucose positron emission tomography. The patient underwent a hybrid surgery involving aortic stent implantation and complete left ventricular mass removal. The gradient between stenosis returned to <10 mmHg after the procedure. Pathologic findings revealed cardiac fibroma.

Discussion It is rare to encounter a patient suffering from both cardiac fibroma and aortic coarctation. No evidences indicated a single cause or syndrome resulting in the coexistence of these two abnormalities. A hybrid surgery involving aortic stent implantation and complete cardiac mass resection could optimize the treatment in such cases.

Keywords Cardiac fibroma • Coarctation of the aorta • Hybrid surgery • Case report

Learning points

- Currently, no available evidences support that the presence of cardiac fibroma and coarctation of the aorta in a single patient results from a single cause or syndrome.
- A hybrid surgery involving aortic stent implantation and cardiac mass resection is an optimized therapy for patients with both cardiac fibroma and coarctation of the aorta.

Introduction

Coarctation of the aorta (CoA) is a common congenital heart defect with an estimated incidence of 1 in 2500 newborns.[1] Cardiac fibroma is a benign but rare primary cardiac tumour, accounting for 1% of all benign cardiac tumours in adults.[2] Considering that CoA and cardiac fibroma are pathologically unrelated, it is extremely rare for a single patient to suffer these two abnormalities. In this report, we describe a case of a large cardiac fibroma complicated with CoA in a young

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female patient. To the best of our knowledge, this is the first case report of CoA with cardiac fibroma, which was successfully treated with a single hybrid procedure.

Timeline

10 years ago	Hypertension
09 April 2020	Transthoracic echocardiogram revealed the following: A large, fixed, moderate-intensity cardiac mass at the left ventricular wall A minimal pressure gradient of 46 mmHg at the beginning of the descending aorta
10 April 2020	Aortic computed tomography angiography revealed the following: A post-ductal-type coarctation of aorta at the beginning segment of the descending aorta A large soft-tissue cardiac mass with a compressed left ventricular chamber Cardiac magnetic resonance imaging revealed the following: Local thickening (82 mm × 51 mm) of the left ventricular wall toward to the heart cavity with low signals on T1-weighted imaging and uneven, high signals on T2-weighted imaging Abundant blood supply in the cardiac mass with gradual late gadolinium enhancement
14 April 2020	^{18}F-fluorodeoxyglucose positron emission tomography revealed the following: A left ventricular soft-tissue lump with annular uneven enhancement Significantly reduced uptake of ^{99m}Tc-methoxy-isobutyl-isonitrile
28 April 2020	A hybrid cardiac surgical procedure Percutaneous catheter intervention with aortic stent implantation Complete surgical removal of the left ventricular mass
11 May 2020	Discharge
4 months after discharge	No reduction in left ventricular ejection fraction and normalized blood pressure. No symptoms while being off medication.

Case presentation

A 22-year-old Chinese female patient visited the clinic with a 10-year history of hypertension. The patient did not complain of chest pain or discomfort after exercise and had normal exercise tolerance. The patient had no other history of cardiovascular disease or cardiovascular risk factors. The patient's blood pressure was 178/87 mmHg measured at the left upper extremity, 184/82 mmHg measured at the right upper extremity, 133/94 mmHg measured at the left lower extremity, and 146/97 mmHg measured at the right lower extremity, with a pressure gradient of more than 40 mmHg. Physical examination revealed that a systolic murmur appeared in the left infraclavicular area, and radio-femoral delay existed. Routine laboratory tests were negative, except for serum angiotensin-converting enzyme level of 13 U/L, which is lower than the normal range (18–55 U/L). Electrocardiogram showed an ST-segment depression in leads I, II, aVL, and V4–V6 and a T-wave inversion in leads I, II, aVL, and V3–V6. Posteroanterior chest X-ray showed an enlarged heart with a cardiothoracic ratio of 0.65 along with an S-shaped spinal curvature. Transthoracic echocardiogram revealed a large, fixed, moderate-intensity cardiac mass (76 mm × 42 mm × 62 mm) from a parasternal short-axis view, located at the apex and middle segment on the lateral and posterior walls of the left ventricle, without a clear boundary from healthy myocardial tissue (*Figure 1*). At the beginning of the descending aorta, a coarctation was observed with a diameter of 10 mm, and the minimal pressure gradient was 46 mmHg. The aortic valve was found to be tricuspid. Aortic computed tomography angiography (CTA) revealed a post-ductal-type CoA at the beginning segment of the descending aorta accompanied by multiple collateral vessels (*Figure 2A*) and a large soft tissue cardiac mass (77 mm × 51 mm) with a compressed left ventricular chamber (*Figure 2B*). Cardiac magnetic resonance imaging revealed a local thickness (82 mm × 51 mm) of the left ventricular wall toward the heart cavity with low signals on T1-weighted imaging, uneven high signals on T2-weighted imaging, and abundant blood supply in the cardiac mass with gradual late gadolinium enhancement (*Figure 3*). Positron emission tomography–CT revealed a left ventricular soft tissue lump with an annular uneven enhancement of ^{18}F-fluorodeoxyglucose metabolic activity and significantly reduced uptake of ^{99m}Tc-methoxy-isobutyl-isonitrile. Coronary angiography revealed no vascular connections between the coronary artery and left ventricular mass with myocardial bridging at the middle segment of the left anterior descending artery without stenosis in the coronary arteries.

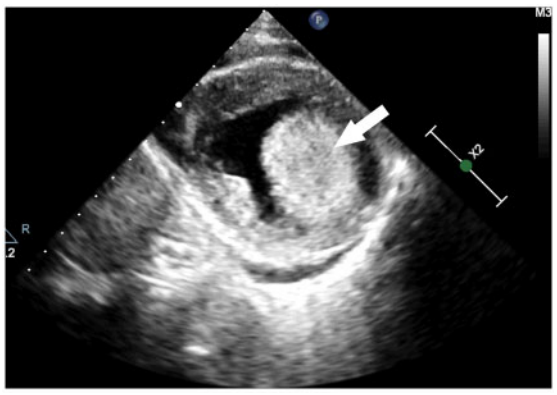

Figure 1 From a parasternal short-axis view, the transthoracic echocardiogram revealed a 76 mm × 42 mm × 62 mm cardiac mass located at the apex and middle segment of the left ventricular wall without a clear boundary with healthy myocardial tissue (white arrow).

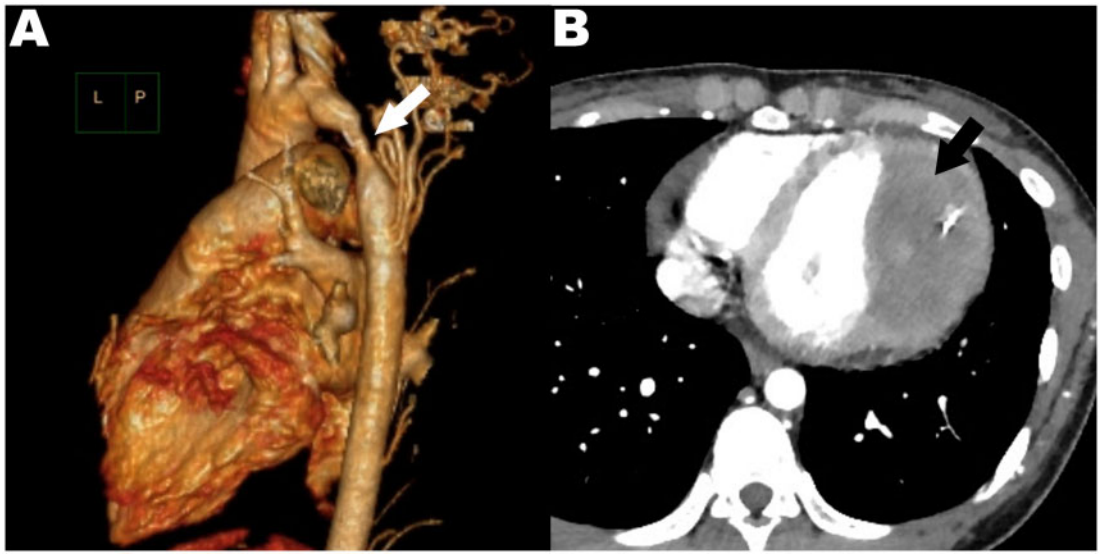

Figure 2 The aortic computed tomography angiography revealed a post-ductal-type coarctation of the aorta (white arrow) at the beginning segment of the descending aorta accompanied by multiple collateral vessels, presenting from a left posterior oblique view (*A*), and a 77 mm × 51 mm large cardiac mass with a compressed left ventricular chamber (black arrow; *B*).

The patient underwent hybrid surgery with percutaneous catheter intervention with the implantation of an aorta stent and complete surgical removal of the left ventricular mass (*Figures 4 and 5A*). The peak-to-peak gradient was lowered to <10 mmHg, and blood pressure returned to normal after aortic stent implantation. Pathological examination confirmed that the cardiac mass was cardiac fibroma (*Figure 5B*). Post-operative transthoracic echocardiogram demonstrated that the ventricular cavity remained normal without left ventricular ejection fraction reduction. At the 4-month follow-up, the patient's left ventricular ejection fraction did not decrease and blood pressure was normal. All medications were stopped.

Discussion

Coarctation of the aorta is a discrete stenosis of the aorta typically located at the insertion point of ductus arteriosus. Blood pressure is increased in the upper extremities and decreased in the lower extremities in patients with CoA. A blood pressure gradient over 20 mmHg indicates a significant CoA.[3] Our patient is presented with a blood pressure gradient of more than 40 mmHg with upper limb hypertension, and more than 50% aortic stenosis at the diaphragm level as revealed by aortic CTA, which are two indications for intervention.[3] Although surgical repair is a traditional treatment, catheter interventional treatment is becoming the first choice for adult patients with appropriate anatomical features in the native CoA.[4] For this young female patient, catheter intervention may be preferable over end-to-end anastomosis to treat CoA because catheter intervention could avoid a left posterolateral thoracotomy additional to the median sternotomy required for complete surgical removal of the left ventricular mass, which diminishes procedure-related injury, shortens the duration of surgery, enhances the safety of the procedure, facilitates post-procedural recovery, and improves long-term quality of life.

Here, we performed an interventional procedure by implanting an aortic stent and successfully lowered the peak-to-peak gradient to <10 mmHg, and the patient's systemic blood pressure returned to normal.

Primary cardiac tumours are rare, with an incidence rate of <1% in the general population. Among them, cardiac fibromas are the second most common, accounting for approximately 1% in all benign cardiac tumours in adult patients.[2] Although histologically benign, cardiac tumours (either primary or secondary) may present with locally malignant characteristics and compromise heart function.[5] Surgical resection usually results in a recurrence-free recovery and good prognosis.[6] In this case, we observed a cardiac mass with an approximately 6-cm diameter. Given the increased risks of fatal arrhythmias and potentially augmented reduction in cardiac function, the cardiac mass was resected and pathologically confirmed as a cardiac fibroma.

Although CoA is easily identified using cardiovascular imaging modalities, distinguishing cardiac fibroma from other sources of cardiac masses is clinically challenging before surgical resection and pathological examination. In this case, the characteristics of the cardiac mass have revealed inconsistencies in multiple cardiovascular imaging modalities before surgery. Nonetheless, whether the causes of CoA and cardiac mass originate from a single origin remains unclear.

Two syndromes are related to CoA and cardiac masses. One is posterior fossa anomalies, haemangioma, arterial anomalies, cardiac anomalies, and eye anomalies (PHACE) syndrome, in which CoA is presented as comorbidity with haemangioma. It is a genetic defect characterized by a constellation of systemic abnormalities, including posterior fossa anomalies, haemangioma, arterial lesions, cardiac

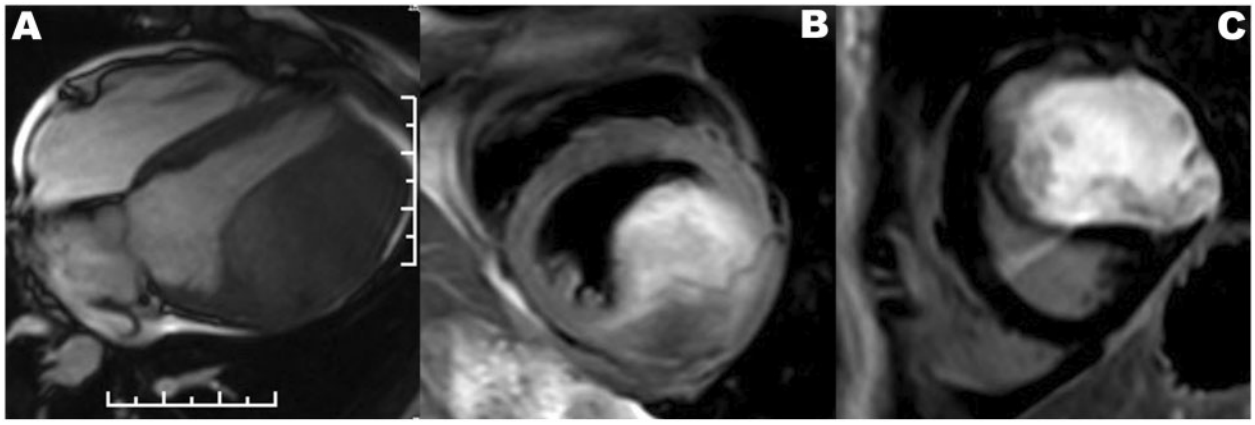

Figure 3 Cardiac magnetic resonance imaging revealed an 82 mm × 51 mm thickness of the left ventricular wall with low signals on T1-weighted imaging (*A*), uneven high signals on T2-weighted imaging (*B*), and abundant blood supply in the cardiac mass with gradual late gadolinium enhancement (*C*).

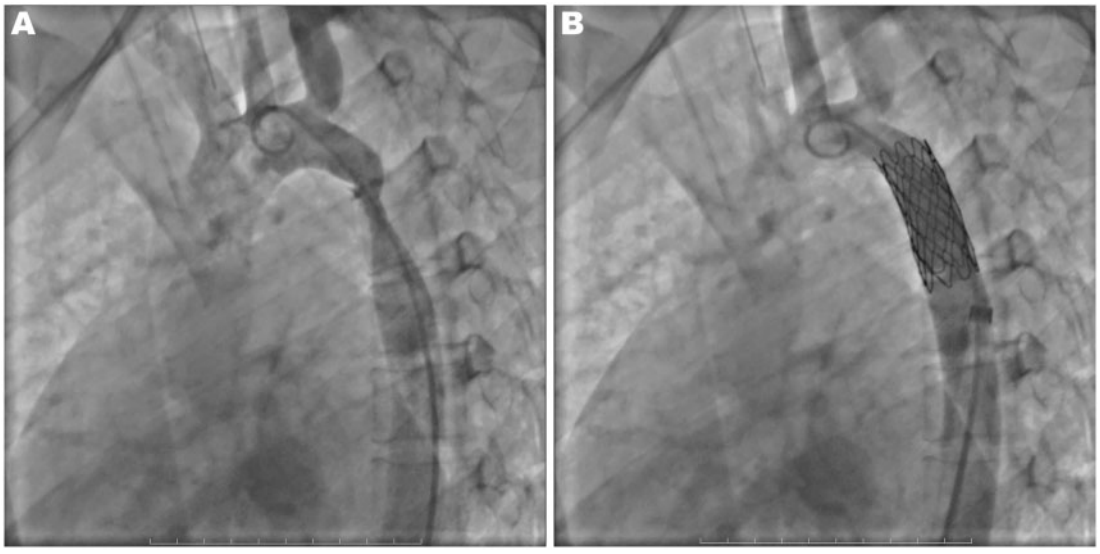

Figure 4 Before (*A*) and after (*B*) aortic stent implantation.

abnormalities/CoA, and eye anomalies, and is usually diagnosed in children.[7] Patients with PHACE syndrome typically have haemangiomas in the head, neck, upper trunk, or upper limbs. In this case, no abnormal clinical manifestations and lesions on the vessels and brain were observed, which did not support the diagnosis of PHACE syndrome. Another syndrome with the manifestation of CoA and cardiac masses is Kabuki syndrome, which is also a congenital defect disease. It might have aortic coarctation as a congenital heart defect and occasionally accompanied by a cardiac tumour.[8] However, patients who are diagnosed with Kabuki syndrome usually have a history of infantile hypotonia, developmental delay, and/or intellectual disability along with other typical dysmorphic features (e.g. long palpebral fissures; arched and broad eyebrows; short columella with a depressed nasal tip; large, prominent, or cupped ears; and persistent fingertip pads) and a pathogenic variant of KMT2D or KDM6A. None of these manifestations were observed in our patient. According to our literature review, no known syndromes could explain the presence of cardiac fibroma and CoA in a single patient. Another explanation could be that it is a new syndrome that originated from an unknown cause, which requires further investigation.

Here, we report a unique case with CoA and cardiac fibroma in a single young patient, who was treated with a hybrid procedure of aortic stent implantation and left ventricular mass surgical removal. In this case, although the cause of the disease remains unclear, the

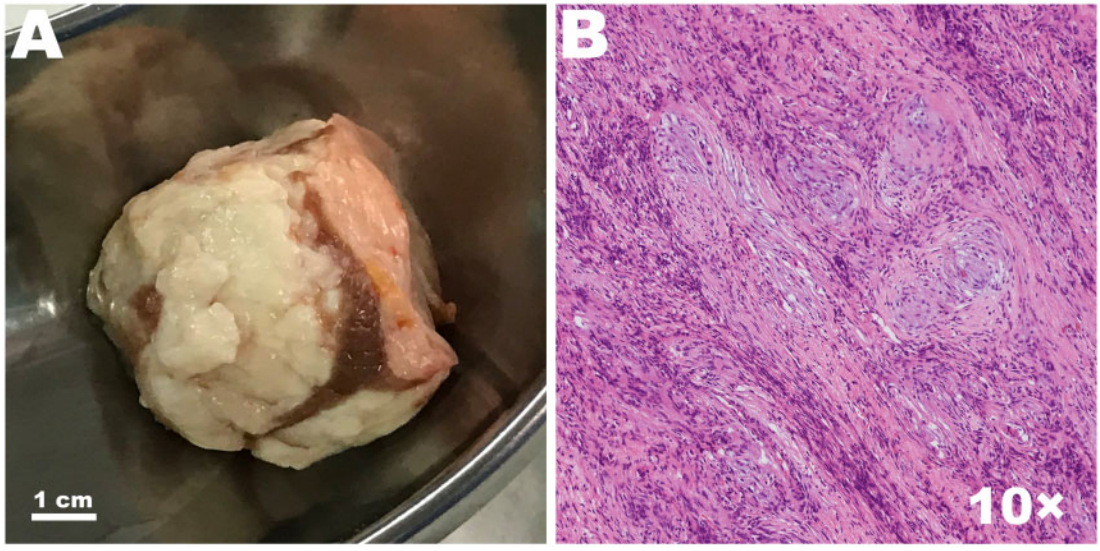

Figure 5 The cardiac tumour was removed from the anterior wall of the left ventricle. The removed tumour has a yellowish and smooth surface (*A*). Pathological examination revealed that the cardiac mass was cardiac fibroma (*B*).

patient received an optimized treatment plan finalized by a multidisciplinary expert panel with the use of multiple imaging modalities and achieved a satisfactory treatment result at the 4-month follow-up.

Lead author biography

Prof. Ben He (MD, FACC FESC, FSCAI) is the leader of First-Class discipline of Cardiology of Shanghai Jiao Tong University, Director of Heart Center, Director of Cardiology Department in Shanghai Chest Hospital affiliated to Shanghai Jiao Tong University. He is an expert and well-experienced in various interventional procedures, especially for highly challenged coronary intervention and left atrial appendage occlusion (LAAO). He got the first certified Watchman proctor by global standard in Shanghai. He has published >70 full-text SCI papers as first or correspondent author, with a total impact factor of over 400 and an H index up to 22.

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 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: none declared.

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References

1. Samanek M, Voriskova M. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: a prospective Bohemia survival study. *Pediatr Cardiol* 1999;**20**:411–417.
2. McAllister HA Jr, Hall RJ, Cooley DA. Tumors of the heart and pericardium. *Curr Probl Cardiol* 1999;**24**:57–116.
3. Erbel R, Aboyans V, Boileau C, Bossone E, Bartolomeo RD, Eggebrecht H et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases: document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). *Eur Heart J* 2014;**35**: 2873–2926.
4. Hu ZP, Wang ZW, Dai XF, Zhan BT, Ren W, Li LC et al. Outcomes of surgical versus balloon angioplasty treatment for native coarctation of the aorta: a meta-analysis. *Ann Vasc Surg* 2014;**28**:394–403.
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. Cho JM, Danielson GK, Puga FJ, Dearani JA, McGregor CG, Tazelaar HD et al. Surgical resection of ventricular cardiac fibromas: early and late results. *Ann Thorac Surg* 2003;**76**:1929–1934.
7. Garzon MC, Epstein LG, Heyer GL, Frommelt PC, Orbach DB, Baylis AL et al. PHACE syndrome: consensus-derived diagnosis and care recommendations. *J Pediatr* 2016;**178**:24–33. e22.
8. Digilio MC, Gnazzo M, Lepri F, Dentici ML, Pisaneschi E, Baban A et al. Congenital heart defects in molecularly proven Kabuki syndrome patients. *Am J Med Genet A* 2017;**173**:2912–2922.