CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 5 (2014) 1058–1060

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Right ventricular myxoma in a patient with tetralogy of Fallot

Fotios Mitropoulos[a], George Giannakoulas[b], Alexandros Kallifatidis[c], Meletios Kanakis[a], Maria Kiaffas[d], Andrew C. Chatzis[a,*]

[a] Department of Paediatric and Congenital Cardiac Surgery, Onassis Cardiac Surgery Centre, Athens, Greece
[b] Department of Cardiology, AHEPA University Hospital, Thessaloniki, Greece
[c] Department of Radiology, St Luke's Hospital, Thessaloniki, Greece
[d] Department of Paediatric Cardiology, Onassis Cardiac Surgery Centre, Athens, Greece

ARTICLE INFO

Article history:
Received 26 September 2013
Received in revised form
24 September 2014
Accepted 29 September 2014
Available online 5 November 2014

Keywords:
Myxoma
Tetralogy of Fallot
Surgical repair
Ventricle
Pulmonary regurgitation

ABSTRACT

INTRODUCTION: Cardiac myxoma is the most common primary cardiac tumour in adulthood and may present in the context of Carney's complex.
PRESENTATION OF CASE: A 32-year-old male with a history of repaired tetralogy of Fallot in childhood was admitted with severe pulmonary valve regurgitation and a mobile mass in the right ventricle. The patient underwent pulmonary valve replacement and mass excision. Pathology examination showed myxoma.
DISCUSSION: In the majority of cases myxomas originate in the atria, nevertheless they can also be found in a ventricular cavity. Myxoma is a prevalent feature of Carney's complex, an inherited, autosomal disease, characterised by multiple tumours in several organs. Tetralogy of Fallot has also been described in association with Carney's complex.
CONCLUSION: Coexistence of tetralogy of Fallot with a cardiac ventricular myxoma in a patient not affected from Carney's complex or other familial syndrome.

1. Introduction

Cardiac myxomas are the most common adult primary cardiac neoplasms, typically located in the atria, most commonly the left.[1] They often appear in a familial form as in Carney's complex.[2] Tetralogy of Fallot has been described previously in association with Carney's complex.[3] Herein, however, we report a right ventricular myxoma in a patient with a history of repaired tetralogy of Fallot not affected from Carney's complex or other familial syndrome.

2. Case

A 32-year-old male patient with a history of repaired tetralogy of Fallot in childhood was admitted with progressive shortness of breath. Transthoracic echocardiography demonstrated severe pulmonary valve regurgitation, aneurysmatic right ventricular outflow tract (RVOT) and right ventricular dilatation. A mobile mass was prominent in the right ventricular apex and an intracardiac thrombus was initially suspected (Fig. 1). Gadolinium-enhanced cardiovascular magnetic resonance imaging (MRI), however, revealed delayed heterogenous enhancement of the right ventricular mass, suggesting a myxoma (Fig. 2a and b).

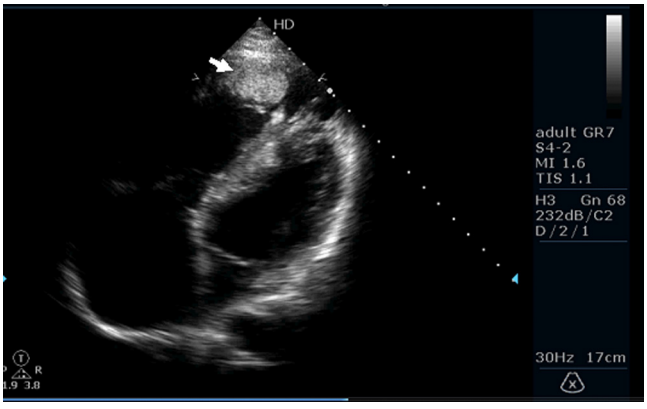

Fig. 1. Cardiac echocardiography depicting the lesion (arrow).

At operation, redo sternotomy was performed and cardiopulmonary bypass with standard bicaval and ascending aorta cannulation instituted. After cardioplegic arrest, the right atrium was incised. While the tricuspid leaflets were retracted, a gelatinous mass became visible in the right ventricular apex. The mass was excised en bloc with a segment of ventricular muscle. Subsequently, the main pulmonary artery was opened and the

* Corresponding author at: Department of Paediatric and Congenital Cardiac Surgery, Onassis Cardiac Surgery Centre, 356 Syngrou Ave, 17674 Kallithea, Athens, Greece. Tel.: +30 210 9493318.
E-mail address: achatzis@ath.forthnet.gr (A.C. Chatzis).

http://dx.doi.org/10.1016/j.ijscr.2014.09.030

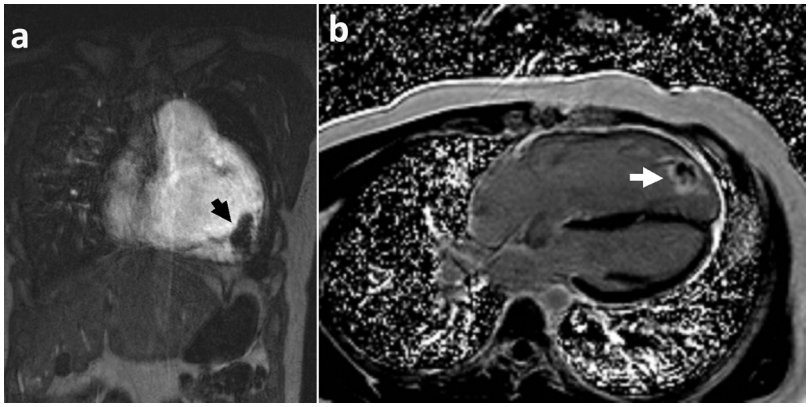

Fig. 2. Magnetic resonance angiography. (a) Showing a non-enhanced, in the arterial phase, lesion (arrow). (b) Delayed gadolinium enhanced (Phase Sensitive Inversion Recovery) sequence showing heterogeneous contrast uptake of the lesion (arrow).

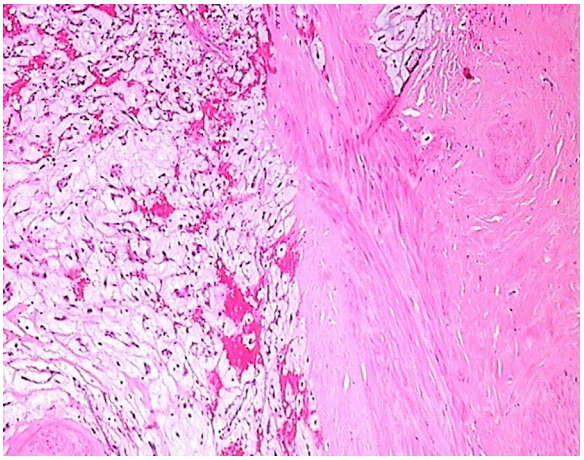

Fig. 3. Histology. Stain: Haematoxylin–Eosin 40×. The myxomatous component of the lesion on the left and the base of the myxoma on the right.

old pericardial RVOT patch and pulmonary valve remnants were excised. A bioprosthetic valve was inserted and reconstruction of the RVOT performed with a Dacron graft. A Kay type tricuspid annuloplasty was also carried out. The patient made an excellent overall recovery and was discharged home the fifth postoperative day in excellent clinical condition. Pathology demonstrated stellate cells with eosinophilic cytoplasm consistent with a final diagnosis of myxoma (Fig. 3).

At twelve months follow-up, the patient remains asymptomatic and echocardiography shows a well-functioning bioprosthetic pulmonary valve, significant decrease in right heart dimensions and no evidence of myxoma recurrence.

3. Discussion

Although rare, myxomas represent the most common tumours of the heart.[1] In the majority of cases they are located in the atria, the left in particular, yet alternate locations like the right ventricle have been reported.[1,4] Their origin as an isolated entity remains obscure and a matter of continuous research.[5] However, the familial form, more likely to be seen in younger patients, predominant male, as in the Carney's complex, research points to certain gene mutations.[6] Carney's complex is an inherited, autosomal neoplastic disease characterised by myxomas, spotty skin pigmentation and endocrine overactivity.[2,6] In this context presence of multiple foci, recurrence, local invasion and malignant transformation have been reported.[5]

Recent technological advances in imaging modalities such as echocardiography and cardiac MRI have resulted in earlier and accurate diagnosis, imperative for safe surgical intervention.[4] Currently, surgery remains the treatment of choice in patients with cardiac myxomas and the operated patients enjoy survival equivalent to that of a standard population.[7] Nonetheless, following successful surgical treatment patients should be followed closely with echocardiography for at least 4 years in order to detect possible recurrence, particularly in younger patients.[1,5]

Operative and long term results have rendered repair of Tetralogy of Fallot as one of the success stories of modern medicine.[8,9] However, progressive right ventricular volume overload from longstanding pulmonary regurgitation, a common post repair aftereffect, is associated with significant late complications; right and left ventricular dysfunction, and most importantly life threatening atrial and ventricular arrhythmias, are the most common.[10] The additional presence of a cytokine producing tumour may, therefore, aggravate clinical status and trigger troublesome arrhythmias.[5] Restoration of pulmonary valve function is consequently recommended to reduce this risk.[10,11]

Cardiac myxoma is rarely located in the right ventricle and hence Carney's complex or other family trait should be sought when encountering this tumour in an uncommon location in young patients.[4,12] Also tetralogy of Fallot has been described in association with Carney's complex.[3] However, in our case, there was no evidence of a familiar disorder. Removal of the tumour and restoration of pulmonary valve function proved successful in this patient with an excellent postoperative and midterm result.

Conflict of interest

The authors declare no conflict of interest.

Funding

None.

Ethical approval

Not applicable.

Author contribution

Study concept and design: Fotios Mitropoulos; data collection: George Giannakoulas and Meletios Kanakis; data analysis and interpretation: Alexandros Kallifatidis, Maria Kiaffas, Andrew C. Chatzis;

writing the paper: Meletios Kanakis, Andrew C. Chatzis; editing the paper: Andrew C. Chatzis

Key learning points

- Myxoma and repaired tetralogy of Fallot in the absence of a familiar trait.
- Myxoma may aggravate symptoms in patients with repaired tetralogy of Fallot.
- Combined pulmonary valve replacement and myxoma excision is feasible in this setting.

References

1. Reynen K. Cardiac myxomas. *N Engl J Med* 1995;**333**:1610–7.
2. Courcoutsakis NA, Tatsi C, Patronas NJ, Lee CCR, Prassopoulos PK, Stratakis CA. The complex of myxomas, spotty skin pigmentation and endocrine overactivity (Carney complex): imaging findings with clinical and pathological correlation. *Insights Imaging* 2013;**4**:119–33.
3. Akbas H, Kirali K, Daglar B, Kutay V, Isik O, Yakut C. Surgical treatment of left-atrial myxoma in Carney's complex. *Thorac Cardiovasc Surg* 1997;**45**:148–50.
4. Satitthummanid S, Tumkosit M, Benjacholamas V, Chattranukulchai P, Boonyaratavej S, Puwanant S. Right ventricular myxoma. *J Am Coll Cardiol* 2011;**57**:630.
5. Amano J, Kono T, Wada Y, Zhang T, Koide N, Fujimori M, et al. Cardiac myxoma: its origin and tumour characteristics. *Ann Thorac Cardiovasc Surg* 2003;**9**:215–21.
6. Bertherat J, Horvath A, Groussin L, Grabar S, Boikos S, Cazabat L, et al. Mutations in regulatory subunit type 1A of cyclic adenosine 5′-monophosphate-dependent protein kinase (PRKAR1A): phenotype analysis in 353 patients and 80 different genotypes. *J Clin Endocrinol Metab* 2009;**94**:2085–91.
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**:S7–15.
8. Lillehei CW, Varco RL, Cohen M, Warden HE, Gott VL, DeWall RA, et al. The first open heart corrections of tetralogy of Fallot. A 26 to 31 year follow-up of 106 patients. *Ann Surg* 1986;**204**:490–502.
9. Apitz C, Webb GD, Redington AN. Tetralogy of Fallot. *Lancet* 2009;**374**:1462–71.
10. Warner KG, O'Brien PKH, Rhodes J, Kaur A, Robinson DA, Payne DD. Expanding the indications for pulmonary valve replacement after repair of tetralogy of Fallot. *Ann Thorac Surg* 2003;**76**:1066–72.
11. Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. *Lancet* 2000;**356**:975–81.
12. Tamura Y, Seki T. Carney complex with right ventricular myxoma following second excision of left atrial myxoma. *Ann Thorac Cardiovasc Surg* 2013 [Epub ahead of print].