

SCIENTIFIC LETTER

Primary cardiac valve tumours

C Basso, T Bottio, M Valente, R Bonato, D Casarotto, G Thiene

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Primary cardiac tumours (CT) are rare lesions with an estimated incidence at necropsy ranging from 0.001% to 0.3%.¹ Myxoma, which is the most frequent primary CT, rarely grows on valve tissue, unlike papillary fibroelastoma, which most often arises from the valvar endocardium.^{1–3} The aim of the study was to assess the prevalence of valvar CT among a consecutive series of primary CT observed in a single cardiovascular pathology centre.

METHODS

The records of the Institute of Pathological Anatomy, University of Padua, Italy were reviewed for CT involving primarily the valve apparatus (that is, leaflets, chordae tendineae, and papillary muscles). CT involving other extravalvar structures, as well as metastatic ones, were excluded.

Clinical data, including transthoracic (TTE) and transoesophageal (TOE) echocardiographic findings and surgical procedure, were collected from clinical records.

Gross and histological examination, including immunohistochemistry and transmission electron microscopy, were performed according to a previously reported method.²

RESULTS

In the time interval 1970–2002 a series of 182 primary CT, 21 autoptic and 161 bioptic, mostly consisting of myxomas (121, 66.5%), were studied.

Fifteen (8%) primary CT were located at the valve level: three autoptic (two male and one female patient, mean (SD) age 78 (4) years) and 12 bioptic (seven male and five female patients, age ranging from 4 months to 77 years, mean 39 years, median 35).

Diagnosis of valvar CT was incidental in eight patients (53%) during either TTE or TOE investigation (six) or at necropsy (two). In the remaining seven patients (47%), clinical presentation consisted of angina or myocardial infarction (three) and congestive heart failure, right ventricular obstruction, dyspnoea, and ictus cerebri (one each, the latter diagnosed at necropsy).

The site of CT was the mitral valve in six (papillary muscle in two), tricuspid valve in five (papillary muscle in one), aortic valve in three, and pulmonary valve in one. Overall, nine were left sided: five of the affected patients were symptomatic and four of the CT were found incidentally by echocardiography (three intraoperative by TOE and one by TTE). Six were right sided: two of the affected patients were symptomatic and four CT were found incidentally (two at necropsy, one during surgical repair of congenital heart disease, and one by TTE). Both the left and right valvar CT that were detected incidentally by TTE required surgery because of the uncertain echocardiographic nature of the mass.

Among the 10 patients who had a TTE investigation, myxoma was misdiagnosed in three and unspecified valve mass was diagnosed in four. TTE did not visualise any mass in the remaining three (30%), and CT was incidentally detected by intraoperative TOE (cardiac surgery for prosthetic valve replacement, aortocoronary bypass graft, and pericardiectomy, respectively) (fig 1). Valve repair was done in all but one



Figure 1 Mitral valve papillary fibroelastoma in a 74 year old patient who underwent coronary bypass surgery. Transoesophageal echocardiogram showing a round mass 5 mm in diameter on the atrial aspect of the anterior mitral valve leaflet (not seen by transthoracic echocardiography). AO, aorta; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; RV, right ventricle.

patient, who had valve replacement because of mitral valve prolapse; quadrangular resection of the posterior mitral valve leaflet and goretex chordae implantation after tricuspid valve papillary muscle removal were also performed.

Valvar CT dimensions ranged from 4–35 mm (mean (SD) 15 (9) mm). All valvar CT were benign and at histology consisted of papillary fibroelastoma in 11 (71%), haematic cyst in two, and lipoma and angioma one each. In two cases superficial endocardial thrombus was visible (fig 2). None of the 121 consecutive cardiac myxomas was located on a valve.

Follow up data were available for all surgically treated patients but one. They were alive after a mean (SD) follow up period of 105 (88) months, except for the patient with mitral valve replacement, who died at home at the age of 64 years of unknown causes. TTE did not show CT recurrence in any of them.

DISCUSSION

Improvements in cardiac imaging have allowed the increasing recognition of even small endocavitary masses, in both symptomatic and asymptomatic patients. In most series, valvar CT constitute < 10% of total primary CT and are mostly histologically benign.^{1–4} In our series, valvar CT accounted for 8% of all primary CT and were frequently found incidentally during routine cardiac investigation. Edwards and colleagues,⁵ in a report of a multi-institutional experience

Abbreviations: CT, cardiac tumours; TOE, transoesophageal echocardiography; TTE, transthoracic echocardiography

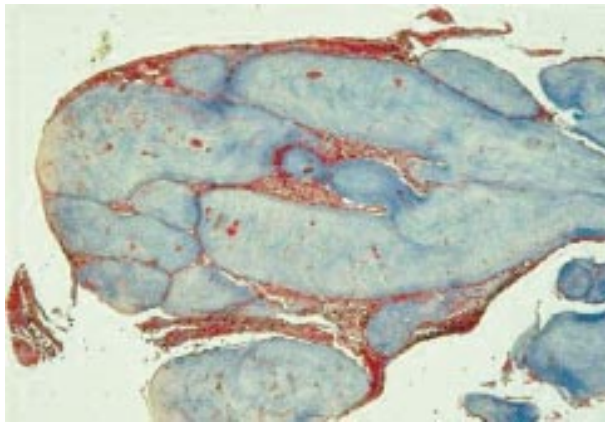


Figure 2 Mitral valve papillary fibroelastoma (same patient as in fig 1). At histology, fibrin thrombus deposition is visible among the fibroelastic fronds (Heidenhain trichrome stain).

with 56 valvar CT including malignant ones, described fibroelastoma as the most frequent (73%); more rarely, lipomas, myxomas, and haematic cysts were reported.

The anatomical location of the CT rather than its size determines the clinical findings. While large infiltrative tumours of the myocardium may be clinically silent, small strategically located endocardial masses may obstruct blood flow or cause embolisation with dramatic sequelae. Among valvar CT, papillary fibroelastoma was generally believed to be not only histologically but also haemodynamically benign since, unlike myxoma, it never reaches a size that causes obstruction and the papillae have a firm consistency, which should imply a low risk of fragmentation with embolisation. Nonetheless, review of the literature found cases symptomatic of transient ischaemic attack, stroke, angina, myocardial infarction, and even sudden death, most probably caused by systemic or coronary embolisation. Indeed, the embolic source may not only be the tumour itself, but also thrombus deposition on the fibroelastic fronds.^{3,5}

The differential diagnosis of papillary fibroelastomas includes other endocardial masses, either neoplastic or non-neoplastic (such as bacterial and non-bacterial vegetations). A definitive response derives from a fine histological

study including immunohistochemical investigation of the excised mass. In this regard, 30% of our patients had a preoperative TTE misdiagnosis of myxoma that was later found to be a fibroelastoma. Moreover, no case of valvar myxoma was found in our series. Nowadays TTE is the best tool in evaluating cardiac masses and especially valvar ones.⁴ However, TOE appears to be superior to TTE when valvar CT are small, as shown in our series by the fact that three valvar CT that were smaller than 1 cm were first undetected by TTE.

Because of the potential haemodynamic “malignant” behaviour of left sided valvar CT, the recommended treatment is surgical excision with preservation of the targeted valve apparatus. Meanwhile, anticoagulation is always advised because of the predisposition for endocardial thrombus deposition and the risk of embolisation.

Authors' affiliations

C Basso, M Valente, G Thiene, Institute of Pathology, University of Padua Medical School, Padua, Italy

T Bottio, D Casarotto, Institute of Cardiac Surgery, University of Padua Medical School

R Bonato, Institute of Anaesthesiology, University of Padua Medical School

Correspondence to: Dr G Thiene, Institute of Pathology, Via A Gabelli, 61, 35121 Padua, Italy; cardpath@unipd.it

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REFERENCES

- 1 **Burke A**, Virmani R. *Atlas of tumor pathology. Tumors of the heart and great vessels*. Washington DC: Armed Forces Institute of Pathology, 1996.
- 2 **Tazelaar HD**, Locke TJ, McGregor CGA. Pathology of surgically excised primary cardiac tumors. *Mayo Clin Proc* 1992;**67**:957–65.
- 3 **Basso C**, Valente M, Poletti A, *et al*. Surgical pathology of primary cardiac and pericardial tumors. *Eur J Cardiothorac Surg* 1997;**12**:730–8.
- 4 **Sun JP**, Asher CR, Yang XS, *et al*. Clinical and echocardiographic characteristics of papillary fibroelastomas. A retrospective and prospective study in 162 patients. *Circulation* 2001;**103**:2687–93.
- 5 **Edwards FE**, Hale D, Cohen A, *et al*. Primary cardiac valve tumors. *Ann Thorac Surg* 1991;**52**:1127–31.



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