Echocardiographic Approach to the Diagnosis of Cardiac Tumors

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Paolo Voci and Francesco Pizzuto

Introduction

Before the clinical introduction of echocardiography, the in vivo diagnosis of cardiac tumors was virtually impossible, and almost exclusively done at autopsy [1].

Echocardiography has definitely changed the diagnostic work-up of cardiac tumors, allowing an easy, early, fast, low-cost, and accurate diagnosis [2–12]. As a result, the prognosis improved too. In fact, before the introduction of echocardiography the prognosis was poor not only for malignant, but also for benign neoplasms, such as myxomas, fibromas, and fibroelastomas, which could not be recognized until progressive obstruction of cardiac chambers, severe arrhythmias, and/or embolic events occurred.

The ultrasound diagnosis of cardiac tumors depends on the technological level of the instrumentation and on the operator's skill and experience, which is particularly true for small and unusual lesions. The cardiologist performing the examination should know the clinical history of the patient and should be expert on the wide spectrum of cardiac masses which may be incidentally encountered during a routine examination.

Very often cardiac tumors are asymptomatic at an early stage, and in about 12% of the cases they are occasionally found during ultrasound examination requested for other reasons [13]. They often produce mild clinical signs, but sometimes their first clinical presentation is dramatic with life-threatening arrhythmias, atrioventricular block, pericardial effusion or tamponade, cardiac failure [14], severe valvular regurgitation, pulmonary hypertension secondary to left ventricular inflow obstruction, and embolic events. Sometimes general signs as fever and artralgia may also occur [15].

Prevalence

Primary cardiac tumors are much rarer than metastatic tumors (prevalence at autopsy of 0.001–0.28% and 1.5–21%, respectively) [16–18]. The "3/4 rule" may be a useful mnemonic aid: more than 3/4 of primary tumors are benign and 3/4 of those are atrial myxomas, which therefore are the most common cardiac tumors; 3/4 of malignant cardiac tumors are sarcomas [19].

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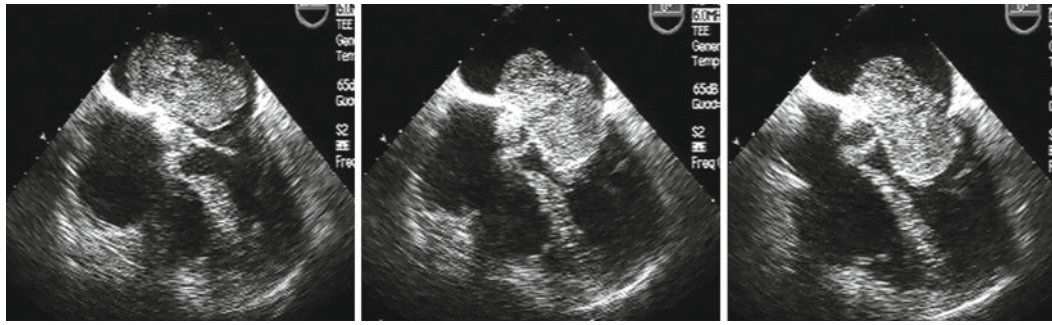

Fig. 7.1 Transesophageal echocardiography. Reprinted with permission of the Italian Society of Cardiology. Large left atrial myxoma attached to the interatrial septum, prolapsing in diastole through the mitral valve

How to Distinguish a Malignant from a Benign Lesion

The histological differentiation of cardiac masses or even the ability to simply distinguish between a malignant and a benign mass is still very difficult with ultrasound, and sometimes even at surgical inspection. Therefore, the histological diagnosis at surgery is deemed necessary to make a correct and definitive diagnosis and to optimize therapy. Despite these important limitations, some ultrasound characteristics as the number of masses, their location, their ultrasound texture, and the presence of calcification, may help suggesting the histological type.

Number. Myxomas are the most frequent cardiac tumors. They are usually made of a single intracardiac mass (Fig. 7.1) but very rarely, and particularly in young patients, they are multiple. Fibromas are made of a single mass too, but they are usually found well before the adolescence when compared to myxomas, and sometimes, albeit very rarely, even in utero (Fig. 7.2). Rhabdomyomas are often multiple (Figs. 7.3 and 7.4), they are associated with tuberous sclerosis, and may regress spontaneously.

Dimensions. Fibromas are usually very large (Fig. 7.2) whereas rhabdomyomas are of variable dimensions (Figs. 7.3 and 7.4), but most often small, and may regress spontaneously. Fibroelastoma is usually small (Fig. 7.5) and other tumors may be very different in size. Obviously, a big mass always derives from the growth of a small mass, therefore, this criterion may have a limited value.

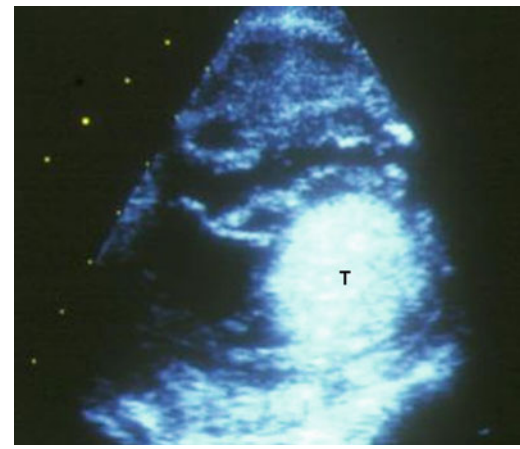

Fig. 7.2 Fetal echocardiography. Reprinted with permission of the Italian Society of Cardiology. Large right ventricular fibroma (T) occasionally seen in a fetus without signs of heart failure

Location. Myxomas are generally found in the atria, particularly in the left atrium, and almost always originate from the atrial septum (Fig. 7.1), whereas rarely involve the valvular or subvalvular apparatus [10]. They are almost never found on the posterior wall of the left atrium, which may help differentiating them from atrial thrombi or leiomyosarcomas. Fibroelastomas are almost always attached to a cardiac valve, mostly to the aortic valve (Fig. 7.5), then to the mitral valve and very rarely the tricuspid valve and the endocardial wall. Rhabdomyomas have a typical intramyocardial distribution. Lipomas are found

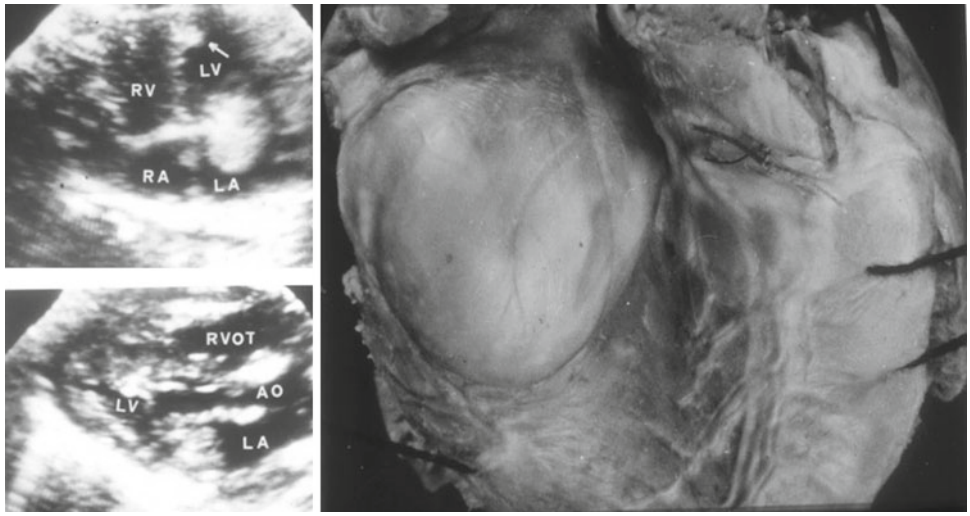

Fig. 7.3 Neonatal echocardiography. Reprinted with permission of the Italian Society of Cardiology. Two rhabdomyomas, the smaller at the apical segment of the interventricular septum (*arrow*) and the larger in the left atrium in a newborn with heart failure, confirmed at autopsy. *AO* aorta, *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle, *RVOT* right ventricular outflow tract

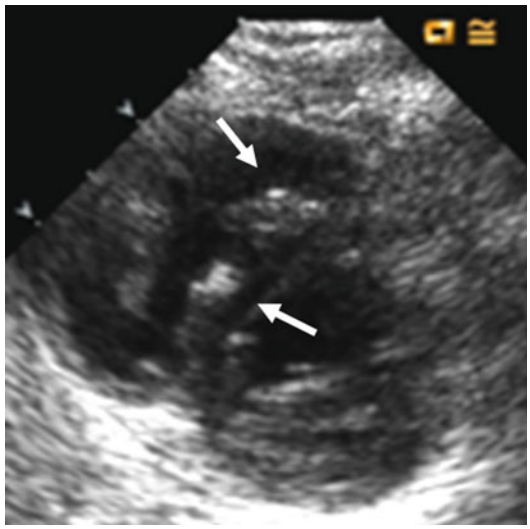

Fig. 7.4 Two-dimensional echocardiography. Reprinted with permission of the Italian Society of Cardiology. Left ventricular short-axis view in a 12-year-old child with tuberous sclerosis. *Arrows* indicate rhabdomyomas in regression

in the interatrial septum, they are round or they may produce a diffuse septal infiltration known as septal lipomatosis, but they may be observed in other structures, included the pericardium. Sarcomas (Figs. 7.6 and 7.7) may be found in any part of the heart.

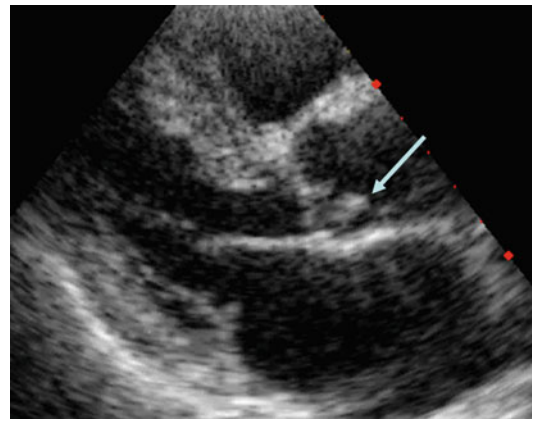

Fig. 7.5 Color-Doppler transthoracic echocardiography. Fibroelastoma of the aortic valve (*arrow*)

Ultrasound structure. The ultrasound "texture" and some peculiar characteristics of cardiac tumors may help in the differential diagnosis. Teratomas and hamartomas are often non-homogeneous and may have spotty calcifications, whereas rhabdomyomas are homogeneous (Figs. 7.3 and 7.4). Left atrial thrombi can be differentiated from myxomas when a multi-layered structure is detected. Echo-lucent or transparent areas, corresponding to hemorrhagic or necrotic spots at histology, may be also be detected in

myxomas, (Fig. 7.1) but exceptionally in vegetations and thrombi. Lipomas are homogeneous and markedly hyper-reflective masses.

Vascularization. A rich vascularization may suggest the diagnosis of malignancy. However, the paraganglioma, which is also called cardiac pheochromocytoma independently from its ability to produce catecholamines, is an exception. In this tumor, which is most often benign, the vascularization is so developed (Fig. 7.8) that coronary steal and even angina may occur [12]. Cardiac hemangioma, which is benign too, is also highly vascularized [5]. Lastly, the presence of vessels in a cardiac mass is not typical of tumors: in fact, old thrombi may be perfused by neovessels which can be imaged by high-resolution ultrasound (Fig. 7.9) [20].

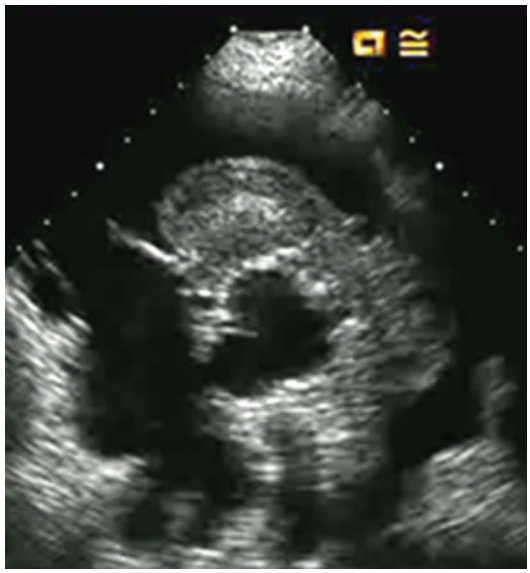

Fig. 7.6 Transthoracic echocardiography. Reprinted with permission of the Italian Society of Cardiology. Short-axis view at the level of the great arteries. Sarcoma infiltrating the right ventricular outflow tract and the pulmonary artery

Cardiac myxoma, the great mimic. The case of myxoma demonstrates how many exceptions may have these diagnostic tips. The cardiac myxoma is a mobile mass attached to the left side of the fossa ovalis by a peduncle allowing wide back and forth excursion in the left atrial chamber. The mass may obstruct in diastole the atrioventricular inflow (Fig. 7.1) thus clinically mimicking mitral stenosis, and similar to mitral stenosis it may produce embolic events secondary to fragmentation of the fragile mass. In 10% of the cases the mass is sessile.

The site of attachment, dimensions, and structure of myxomas may be very heterogeneous. In the large study of Goswami et al. [3] 84% originated from the left atrium, 12% from the right atrium, and the remaining 4% from both atria. Sixty-nine percent of left atrial myxoma originated from the fossa ovalis, 28% from the inferior

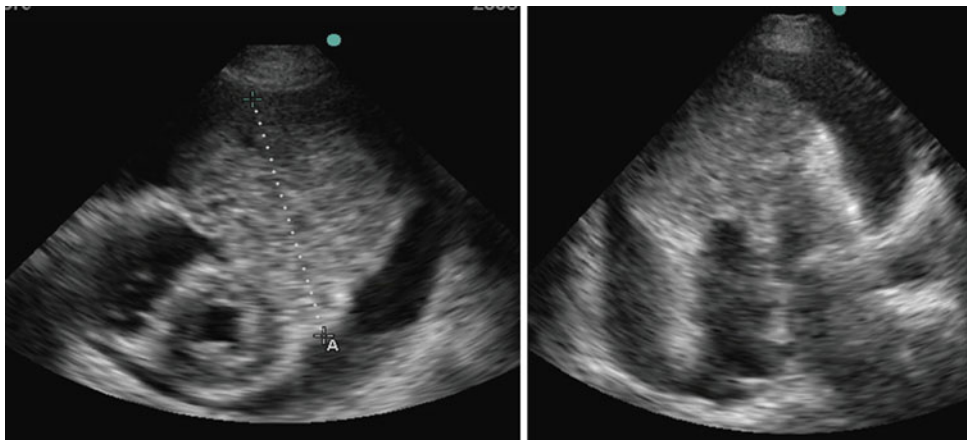

Fig. 7.7 Transthoracic echocardiography. Large sarcoma infiltrating the anteroseptal and apical wall of both left and right ventricles, with massive pericardial effusion

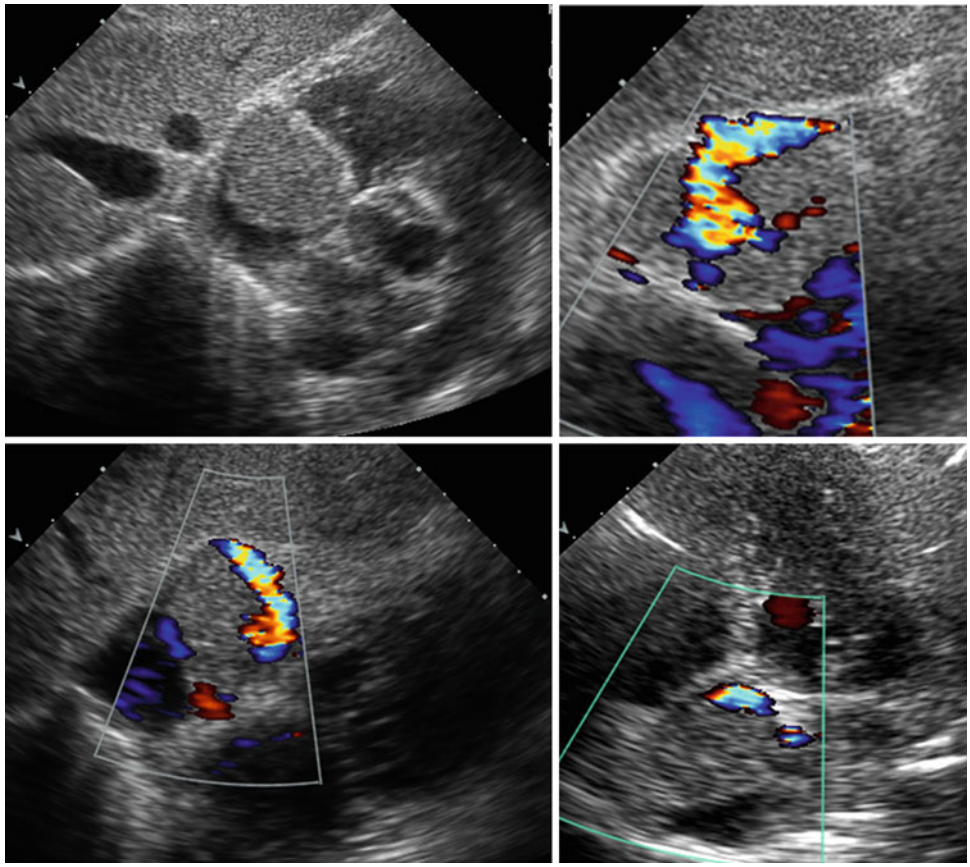

Fig. 7.8 Transthoracic color-Doppler echocardiography. Reprinted with permission of the Italian Society of Cardiology. Richly vascularized right atrial paraganglioma

border of the septum, and the remaining 3% from the lateral wall. Similarly, the great majority of right atrial myxomas originated from the right side of the fossa ovalis. All biatrial myxomas in this large series straddled the fossa ovalis. Very rarely myxomas may be found in both the right and left ventricle. The surface is most often smooth and globular, but in 15% of the cases it may be papillary and friable. Echo-lucent areas within the myxoma can be found in 70% of the cases, often as large as 1 cm [2]. In about 10% of the cases, calcifications may be also observed.

When the diagnosis is made in a young patient, one should always consider the presence of multiple and relapsing lesions and a familiar distribution.

Which Role for Transesophageal Echocardiography?

Transthoracic echocardiography has greatly improved in the last decade allowing better detection of structures in the far field, as the atria. In a recent review of 149 primary cardiac tumors in China [1] transthoracic echocardiography was diagnostic in 93.3% of the cases. In the 10 remaining cases missed at transthoracic echocardiography the mass was in the pericardium (eight cases), in the left atrium (one case), and on the posterior surface of the heart (one case).

Of course, transesophageal echocardiography [5–8] has a better resolution than transthoracic echocardiography, because the acoustic

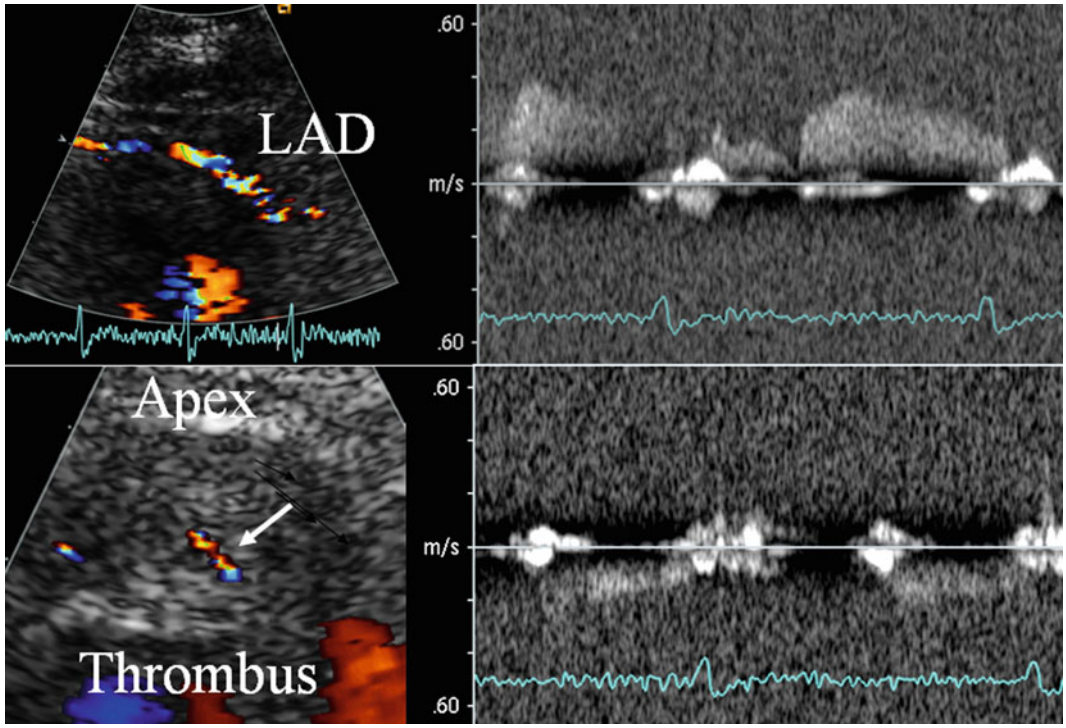

Fig. 7.9 Transthoracic color-Doppler echocardiography. Vascularized apical thrombus. *Upper panel*: epicardial tract of the left anterior descending (LAD) coronary artery visualized by color-Doppler and the corresponding pulsed Doppler tracing with the characteristic anterograde systolic and diastolic flow. *Lower panel*: vascularized apical thrombus with flow directed away from the transducer (*modified from Voci et al.* [20])

impedance is lower and the transducers have a higher spatial resolution. Despite this advantage, transesophageal echocardiography not often brings additional information over transthoracic echocardiography, relevant for surgical referral of the patient [3]. However, the superior wall of the right atrium and the right atrial appendage are only incompletely seen by transthoracic echocardiography and anatomical details of the inferior and superior vena cava may be better seen by transesophageal echocardiography [7].

Intraoperative transesophageal echocardiography is useful to guide surgery and to evaluate the results of valve repair in case of infiltration of the atrioventricular valves or to confirm the absence of intracardiac shunts after septal repair. Lastly, transesophageal echocardiography monitors weaning from cardiopulmonary bypass, particularly patients undergoing partial ventricular resection for infiltrating tumors. Sometimes transesophageal echocardiography may help guiding transvenous biopsy of right chamber masses particularly of inoperable malignant tumors, when the histology is necessary to guide chemotherapy.

Metastatic tumors

The prevalence of cardiac metastasis from tumors originated in the lung, breast, kidney, skin, and colon is 100–1,000 times higher compared to primary cardiac tumors [1]. Cardiac metastases

represent a social problem, because paradoxically the improvement in chemo- and radiotherapy, prolonging the life expectancy of these patients also increased the prevalence of cardiac metastasis, from 0.2 to 6% before 1996 to more than 10% in 2003 [2]. Cardiac metastases can be intramyocardial (Fig. 7.10) and/or pericardial (Fig. 7.11) and produce pericardial effusion and tamponade. Lymphoma may compress the cardiac chambers (Fig. 7.12) or produce the superior vena cava syndrome (Fig. 7.13). Kidney tumors may infiltrate the inferior vena cava and reach the right atrium (Fig. 7.14), even causing pulmonary neoplastic embolism.

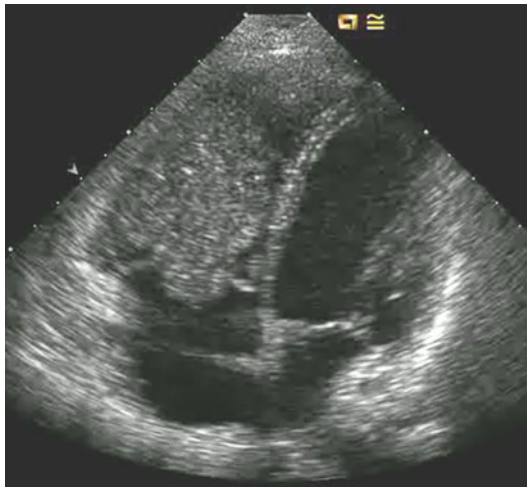

Fig. 7.10 Two-dimensional echocardiography, apical four-chamber projection. Reprinted with permission of the Italian Society of Cardiology. Metastatic melanoma infiltrating almost entirely the right ventricle, and prolapsing in systole through the tricuspid valve

Conclusions

Nowadays, the diagnosis of cardiac tumors is based mainly on transthoracic echocardiography which allows to visualize a cardiac mass and to roughly predict its nature. Transesophageal echocardiography is indicated in the rare cases when transthoracic echocardiography is non-diagnostic, and in the perioperative monitoring of complex lesions.

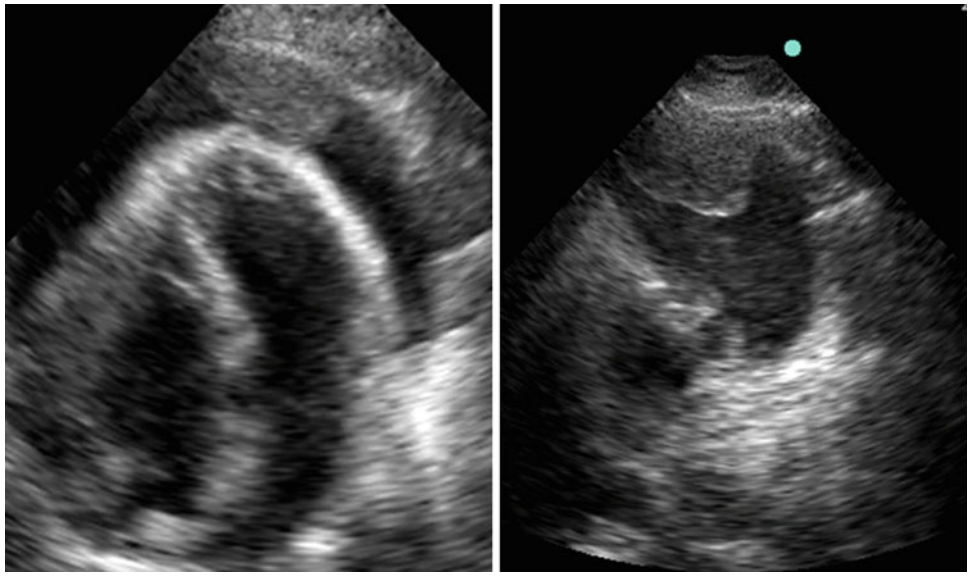

Fig. 7.11 Two-dimensional echocardiography. Pericardial metastasis secondary to a colon carcinoma. The off-axis projection in the *right panel* clearly shows that the mass is attached to the parietal pericardium and does not infiltrate the myocardium, being therefore suitable for surgical resection

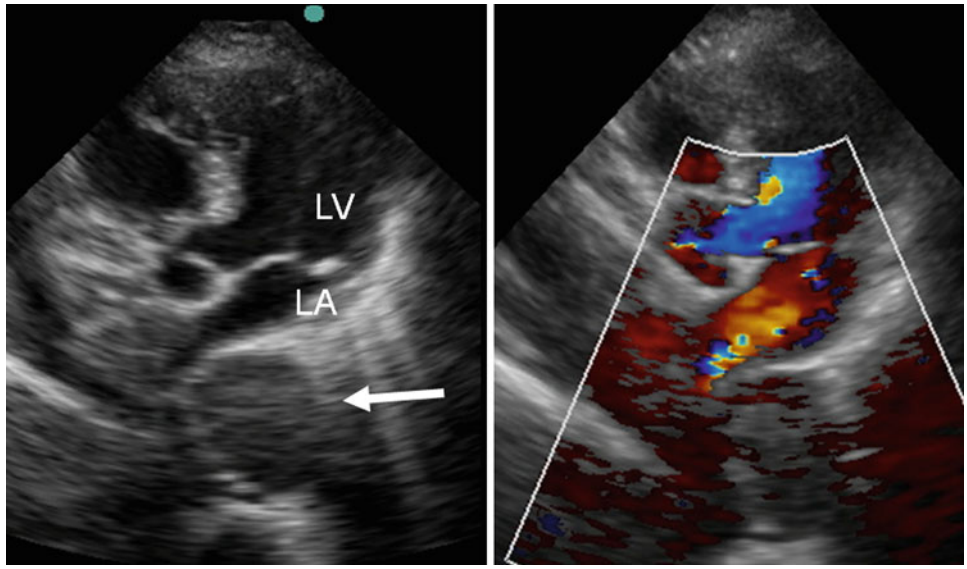

Fig. 7.12 Mediastinal lymphoma compressing the left atrium (LA). *LV* left ventricle

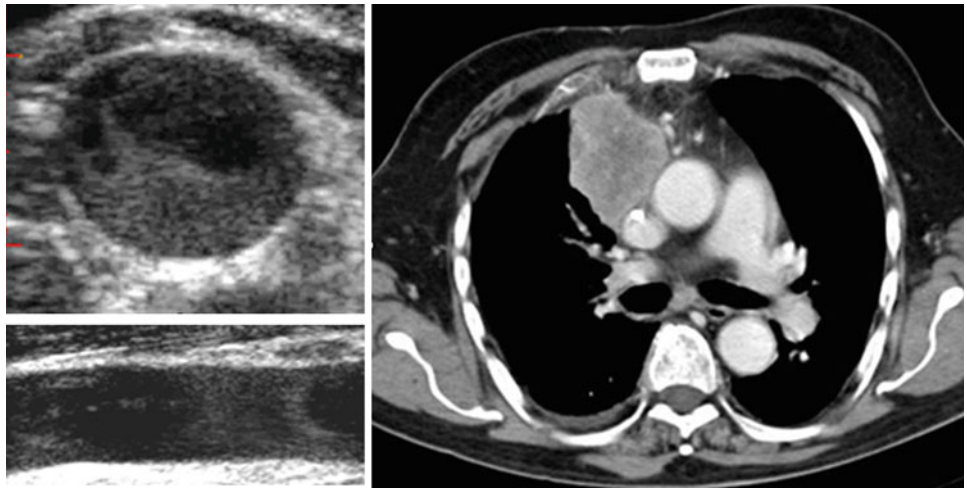

Fig. 7.13 Intrathoracic lymphoma (CT scan, *right panel*) leading to superior vena cava syndrome with marked stagnation of flow in the jugular vein (*left panel*)

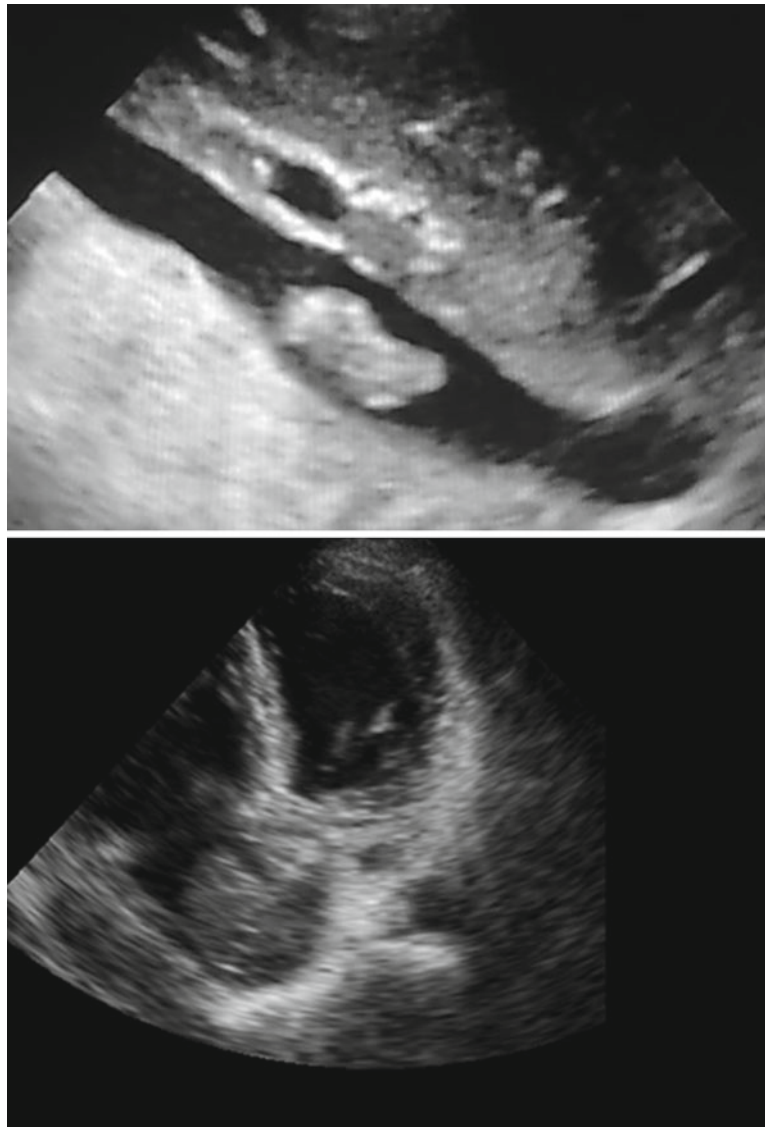

Fig. 7.14 Right kidney tumor producing a mobile mass in the inferior vena cava (*upper panel*) and right atrial metastasis (*lower panel*)

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