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Use of CT in the Evaluation of Primary Cardiac Tumors

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Abstract. Two cases of primary cardiac sarcomas diagnosed with CT are presented. CT demonstrated the origin, extent, and potential pathology of the tumors. In both cases CT provided more specific information for diagnosis and treatment planning than the 2D echocardiography. The advantages of CT in the detection of cardiac tumors as well as its potential advantages over 2D echocardiography are also discussed.

Key words: Computer tomography—Cardiac tumors—Osteogenic sarcoma—Leiomyosarcoma—Cardiac tumors, diagnosis

The use of computed tomography (CT) in the evaluation of cardiac tumors has steadily evolved over the last 7 years. Initial case reports first noted its usefulness in the diagnosis of atrial myxoma [1, 2] and metastatic intracavitary pericardial lesions [3, 4]. The superiority of CT over echocardiography and angiocardiography for the diagnosis of pericardial tumors has now been well established [4–6]. In addition, CT has proved most useful in determining the full extent of both metastatic neoplasms and primary sarcomas of the heart and great vessels [7–9]. In a recent report by FitzGerald [10], a case of primary sarcoma of the pulmonary trunk was described in which the CT appearance was considered a potentially definitive diagnostic procedure.

We present our experience with two cases of primary sarcoma of the heart and great vessels in which CT was useful in evaluating the full extent of tumor and in suggesting a more definitive diagnosis.

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Case Reports

Case 1

A 58-year-old white male developed sudden onset of dyspnea, fatigue, and nonproductive cough. Within 1 week the patient developed pedal edema and increased abdominal girth. Initial evaluation by a private physician failed to define the illness. Two months later the patient's condition dramatically deteriorated to dyspnea at rest with pedal edema and increased abdominal girth. Upon referral to a cardiologist, a 2D echocardiogram revealed bilateral atrial masses consistent with atrial myxoma. The patient was subsequently referred to our institution.

Physical examination was significant for jugulovenous distension, an S_4 gallop, and a grade 2/6 systolic "scratch" along the left sternal border. The abdomen was protruberant with shifting dullness and hepatomegaly. There was bilateral pedal edema. Two-dimensional echocardiography was performed which demonstrated large bilateral atrial masses with broad based attachment to the septum. Computed tomography with bolus injection of intravenous contrast material (Fig. 1) revealed a calcified mass involving half of the left atrium. The mass had a broad base that extended completely through the septum and was contiguous with a large calcified mass involving almost the entire right atrium. The right atrial mass was also shown to extend into the inferior vena cava with a filling defect continuing down to the level of the renal veins. A clinical diagnosis of primary cardiac sarcoma with osteosarcomatous elements was made from these findings.

The patient was taken to surgery, where a heterogeneous, multilobulated tumor arising bilaterally from the interatrial septum and occupying much of both atria was debulked. Tumor was found within the distal inferior vena cava with more proximal attachment of thrombus. Histopathology and electron microscopy demonstrated a mixed osteosarcoma and leiomyosarcoma.

Case 2

A 43-year-old white male developed progressive dyspnea on exertion, lethargy, and leg weakness over a 3-month period. The patient's symptoms progressively worsened in the next 3 months, and the development of pedal edema was noted.

Physical examination was significant for jugolovenous distension, a precordial thrill, a loud P_{2V}, and two distinct, grade 3/6 murmurs heard at the apex and upper left sternal border. There was mild pedal edema. The plain chest roentgenogram showed cardiomegaly and right costrophrenic blunting. An initial 2D echocardiogram revealed an enlarged right atrium and ventricle with paradoxical septal motion consistent with right ventricular

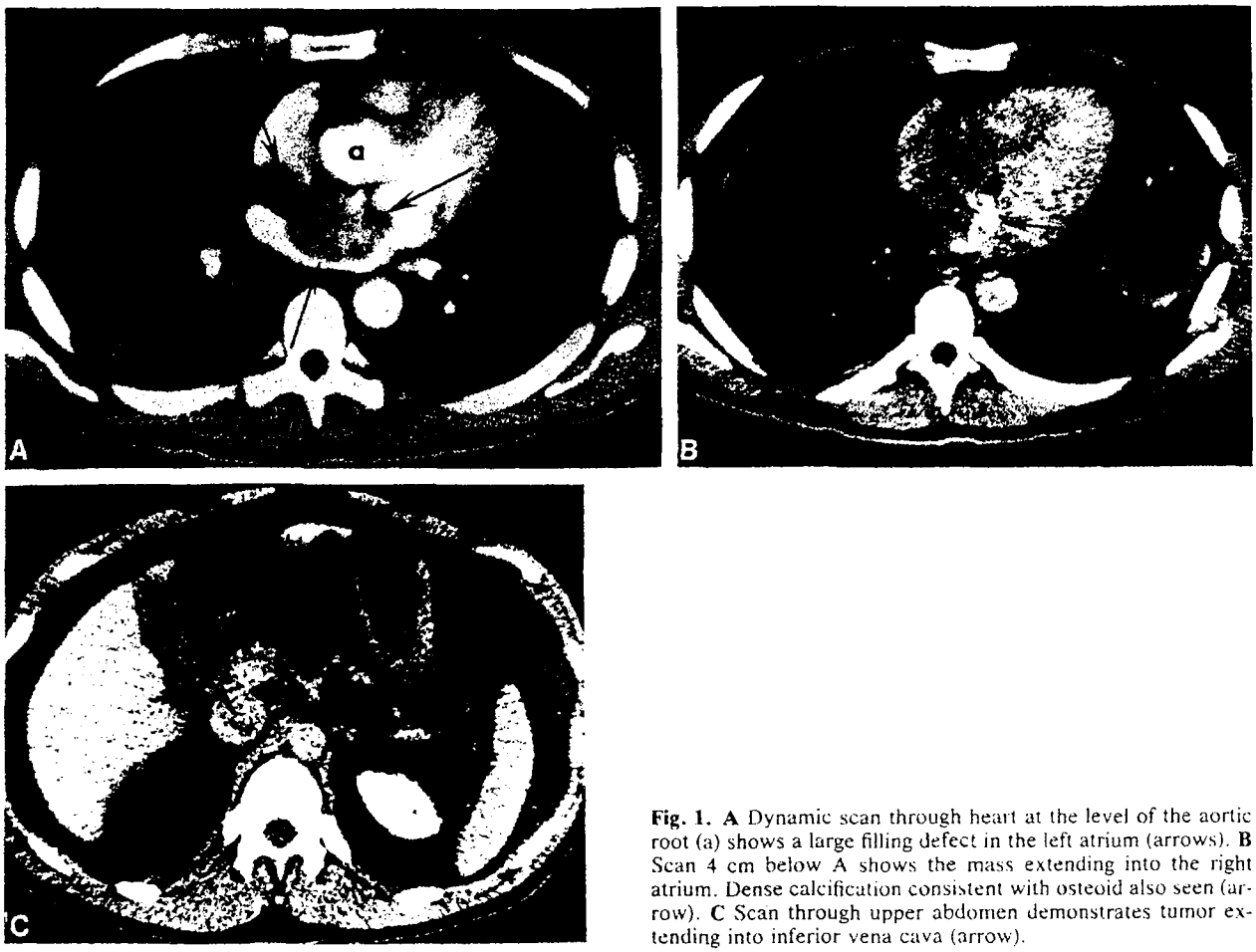

Fig. 1. **A** Dynamic scan through heart at the level of the aortic root (a) shows a large filling defect in the left atrium (arrows). **B** Scan 4 cm below A shows the mass extending into the right atrium. Dense calcification consistent with osteoid also seen (arrow). **C** Scan through upper abdomen demonstrates tumor extending into inferior vena cava (arrow).

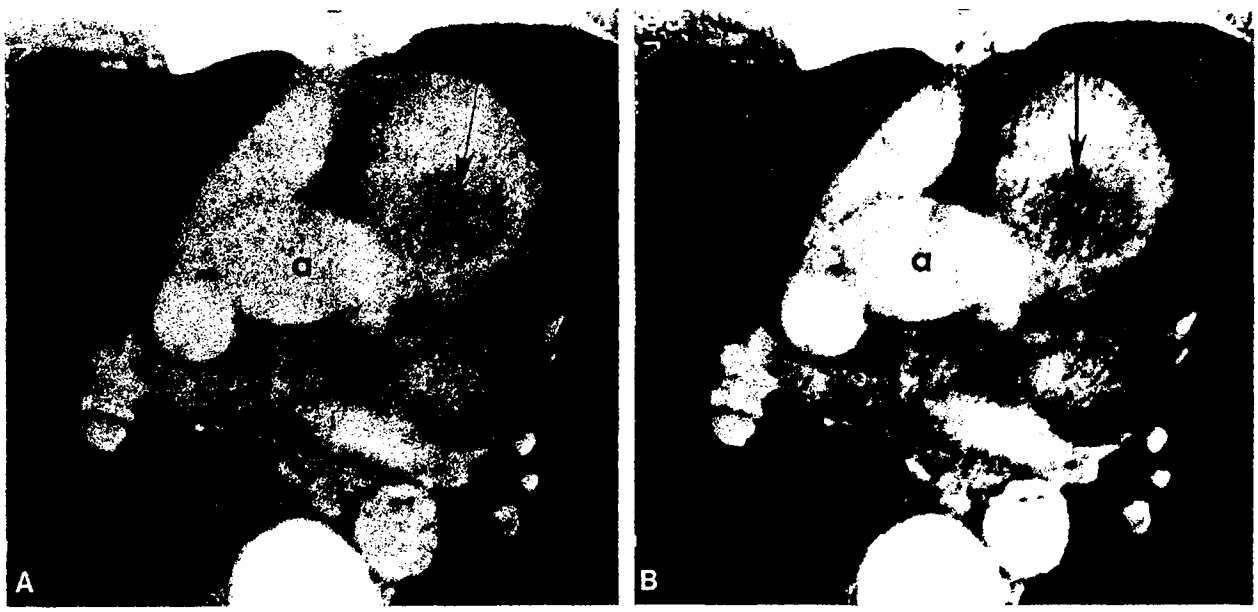

Fig. 2. **A** CT scan through level of aortic root (a) demonstrates a 2.5-cm mass in pulmonary artery (arrow). **B** Image at narrow window setting demonstrates the apparent nodularity of the mass.

overload. Angiocardiography demonstrated a functional pulmonic stenosis secondary to an apparent ill-defined intrinsic mass. A CT scan with bolus injection of intravenous contrast showed a large mass involving the main pulmonary artery and right ventricle with multiple sites of attachment (Fig. 2). A second 2D echocardiogram was performed, which this time demonstrated masses in the pulmonary outflow tract. These findings by CT and 2D echocardiography strongly suggested a clinical diagnosis of primary sarcoma of the pulmonary artery and right ventricle.

The patient was taken to surgery and found to have multiple multilobulated masses with broad bases of attachment in the proximal pulmonary trunk and right ventricle. The tumors could only be incompletely excised because of their extensive attachment. Hispathology and electron microscopy demonstrated an undifferentiated sarcoma.

Discussion

Primary tumors of the heart are relatively rare with an estimated incidence of 2–8 per 10,000 in the general population [11]. Approximately 70–80% are benign, and of these more than two-thirds are myxomas [12]. All primary malignancies of the heart are sarcomas. The two most common are angiosarcoma, which almost invariably occurs in the right atrium, and rhabdomyosarcoma, which has no characteristic localization [11, 13]. Other tumor types such as leiomyosarcomas, osteosarcomas, and undifferentiated sarcomas occur much less frequently [14]. Primary sarcomas of the pulmonary trunk are extremely rare. A report of only the 60th such known case was recently described by Bleisch and Kraus [15]. In their review of the literature it was found that although the tumor has variable histological and gross characteristics, it seems to originate in a relatively constant anatomical position within the proximal pulmonary trunk.

The development of more sophisticated imaging modalities within the last two decades has led to substantial improvements in the preoperative diagnosis of primary cardiac tumors. In particular, 2D echocardiography has received substantial attention because of its very high sensitivity in detecting intracardiac pathology while being entirely noninvasive and risk-free. It is currently well accepted that 2D echocardiography is the diagnostic imaging method of choice for the initial evaluation of suspected cardiac tumors [11, 16]. This technique, however, has certain limitations which diminish its overall diagnostic precision and accuracy. Poor image quality may be encountered when suboptimal acoustic windows result from chronic lung and pleural disease or abnormal configuration of the chest [3]. Even under ideal circumstances only a limited amount of sectional anatomy is available to scanning owing to the physical constraints imposed by standard positions of acoustic windows. A more significant limitation of 2D echocardiography is its inability to adequately assess the pericardium, great vessels, and extracardiac structures, all of which are important considerations when evaluating a potential cardiac tumor [3, 5, 6]. In addition, this imaging technique produces limited resolution of tissue planes and relatively poor tissue discrimination, which can result in underestimation of pathology and misdiagnosis [11].

The advent of high-resolution CT with short scanning times has made it possible to obtain higher-quality images of the cardiac chambers, septa, pericardium, and great vessels. Quality imaging of the thorax can be done more universally except in rare situations where too much artifact occurs from the presence of extensive metallic surgical hardware. Also the standard cross-sectional format of CT is often preferred for best demonstrating cardiac anatomy [3, 4]. This has enabled CT to fill in much of the void created by the limitations of 2D echocardiography in evaluating cardiac tumors. Thus, CT is ideally suited for demonstrating tumors of the pericardium [5] and evaluating the full extent of tumor as occurs with direct invasion of the myocardium, pericardium, and mediastinum [4, 6]. Direct extension into contiguous great vessels can also be well demonstrated [3, 10]. An additional advantage of CT is its ability to detect embolic pulmonary metastases [6, 8]. Such detailed information obtained through CT can be very useful for both better preoperative planning by the surgeons and enabling the radiologist to make a more accurate and definitive diagnosis (e.g., benign vs. malignant neoplasm).

The two cases illustrated demonstrate how information obtained by CT substantially improves the overall evaluation of suspected cardiac tumors. In case 1, 2D echocardiography demonstrated atrial masses with broad septal attachment. This finding was thought to be consistent with bilateral atrial myxomas, tumors that typically have a distinct biatrial "dumbbell" appearance [13]. CT of the lesion better defined its relationship with the interatrial septum. It was clear that there was extensive invasion through the septum instead of a narrow stalk of tumor growing through the foramen ovale, as seen with biatrial myxomas [17]. Also direct extension of tumor into the distal inferior vena cava was clearly seen. Because of these findings, not only was there more information available for surgical planning, but the correct diagnosis of primary cardiac sarcoma could be confidently suggested. Perhaps the diagnosis could have been more specific, because the calcification present indicated that at least part of the tumor consisted of osteosarcomatous elements, a finding later confirmed histologically.

In case 2, the initial 2D echocardiogram missed the diagnosis. Cardiac catheterization and angiocar-

diography only revealed functional pulmonic stenosis caused by an ill-defined mass within the proximal pulmonary trunk. CT nicely demonstrated a tumor involving both the proximal pulmonary trunk and right ventricle with multiple sites of attachment. As originally reported by FitzGerald [10], this characteristic location of tumor enabled us to more confidently suggest a diagnosis of primary sarcoma of the pulmonary trunk and right ventricle. Also the findings of no pulmonary metastases or mediastinal extension provided important information for therapeutic planning, since the cardiac surgeons were less reluctant to attempt resection in the absence of extracardiac involvement.

We conclude that CT with bolus injection of contrast material is a very valuable noninvasive imaging modality for the evaluation of suspected cardiac tumors. The technique should be used in conjunction with 2D echocardiography to provide more information for surgical planning and to increase the likelihood of making an accurate and precise diagnosis.

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