A primary osteosarcoma of the heart as a cause of recurrent peripheral arterial emboli

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Summary
The case of a 66-year-old woman with a primary cardiac osteosarcoma is described. These distinctly rare malignant tumors arise preferentially in the left atrium. Clinically, they often present symptoms of both, intramural and intracavitary neoplasm in addition to general weakness, recurrent breast pain, and dyspnea. As shown in the present case, with growing intracavitary tumor masses the risk for peripheral arterial including cerebral embolism increases. Consequently, in most patients with symptoms of systemic arterial embolism of unknown origin performance of transesophageal echocardiography seems advisable, which is presently the most convenient noninvasive imaging method to exclude or to identify intracardiac sources of emboli, irrespective of their type.

Key words: arterial embolism, cardiac osteosarcoma, transesophageal echocardiography

Introduction
Primary malignant tumors of the heart are rare and can arise in any area of the heart. The most commonly reported histologic types are angiosarcomas, usually located in the right atrium or the right ventricle [1]. In contrast, all of the distinctly rare primary cardiac sarcomas with osteosarcomatous differentiation have been found to be localised within the left atrium [2] or at the mitral valve [3]. The preference of osteosarcomas to occur in these sites is not explained. Although the most common sites for benign cardiac myxomas are identical, there is no evidence that cardiac osteosarcomas represent malignant transformation of originally benign tumors [4].

The nature of the clinical presentation may help to suggest the diagnosis, because malignant cardiac tumors often show clinical features of both intramural effects with conduction abnormalities or arrhythmia as well as intracavitary effects resulting in left ventricular inflow obstruction [5]. However, most clinical signs found in up to 50% of the patients are non-specific such as general weakness, breast pain, dyspnea, or peripheral arterial embolism. Because of this it is important to examine thoroughly any suspicious intracardiac structure irrespective of its location or the presenting clinical symptoms, always keeping in mind the possibility that it is caused by a malignant tumor. Today with transesophageal echocardiography (TEE) and magnetic resonance imaging (MRI) we have two excellent noninvasive methods available to approach this aim. According to autopsy reports the prevalence of primary cardiac tumors ranges from 0.001% to 0.28%. From these tumors about 25% were found to be malignant [6]. Until now, only 10 cases of a primary intracardiac osteosarcoma have been reported [6]. In addition, nine primary cardiac sarcomas with osteosarcomatous differentiation were histologically identified in a recent report reviewing 81 primary sarcomas of the heart [2]. Here, we present another case of this extremely rare malignant cardiac tumor.

Case report
A 66-year-old woman was presented with a nine-month history of progressive general weakness and a significant loss of body weight. Moreover, she reported recurrent episodes of dizziness and shortness of breath during this period. No fever episodes were remembered. Just prior to hospital admission, a syncopal episode with vomiting occurred. On physical examination her blood pressure was 125/85 mmHg, with a regular pulse of 88 bpm. Heart auscultation revealed a 2/6 systolic murmur over the area of the tricuspid valve indicating tricuspid valve insufficiency, probably due to an elevated right heart load. Two days later, she presented an amaurosis fugax of the right side suggesting cerebrovascular disease or an arterial embolic event, but no abnormalities of the retina were found upon ophthalmic examination.

Because of a rapidly progressive dyspnea the patient was suspected to have an acute pulmonary embolism and was transferred to the intensive care unit. Chest X-ray examination revealed borderline cardiomegaly and a left sided pleural effusion. However, scintigraphic...
examination of the lung was normal. At the third day, by transthoracic echocardiography an echo-dense structure adherent to the lateral atrial wall in the enlarged left atrium was detected. Further examination by TEE (Figure 1) confirmed an immobile polypoid structure measuring 3.8 x 3.0 cm in the left atrium with significant obstruction of transmitral blood flow. By computed tomography (CT) and MRI (Figure 2) infiltration of the neighbouring left pleura and the right atrial wall was documented. Cardiac catheterization revealed an elevated pulmonary artery pressure (60/30 mmHg) and pulmonary capillary wedge pressure (35 mmHg) with a normal left ventricular end diastolic pressure (10 mmHg) indicating either pulmonary vein occlusion or elevated left atrial pressure. After excluding coronary heart disease by coronary angiography the patient was referred for urgent surgery. Intraoperatively, a 40 cm³ nodular infiltrating mass was found originating from the posterior left atrial wall extending to the left upper pulmonary vein with infiltration of the right atrial wall. Complete excision of the tumor was not possible and, thus, operation was stopped after resection of all tumor parts extending into the left atrium. Histologic examination (Figure 3) revealed an osteoid-producing primary osteosarcoma of the heart with a high level of mitotic activity. In addition, the tumor contained some structural elements of an angiosarcoma and a chondrosarcoma. X-ray studies before and after surgery did not identify any primary lesions of the skeletal system. CT of the abdomen and abdominal ultrasound revealed metastases in the left kidney. Because of motor disturbances of the left hand, an intraoperative embolic event affecting the right arte- ria cerebri media was suspected. However, cranial CT three days after surgery only showed signs of prior ischemic events in brain areas depending on the right a. cerebelli superior and the left a. sulci postcentralis. It was tempting to speculate that these events might have been associated with the recurrent episodes of dizziness in the patient's clinical history. Following surgery the patient was in a bad condition and, thus, palliative chemotherapy was not given. The patient died five weeks
after discharge from our hospital, an autopsy revealed extensive metastatic spread of the tumor.

Discussion

As shown in the present case, the clinical signs and symptoms of intracardiac tumors are often independent of their type and are not specific. In addition, most general symptoms of malignant cardiac tumors like weight loss (25% incidence), subfebrile temperature (50% incidence), and weakness are typical and identical to the symptoms of other malignant diseases [7]. However, dependent on their localisation these malignant tumors are able to mimic symptoms and induce pathophysiological changes which are characteristic for primary cardiac disease. As an example, pathological findings upon auscultation of the heart can be obtained in up to 75% of cardiac tumors, and right heart failure occurs in up to 70% of the patients suffering from cardiac myxoma. Whereas the latter benign intracardiac tumors are the source of most tumor emboli because of their friable consistency, other types of tumors including malignant cardiac neoplasm may occasionally embolize, too [8]. The reported occurrence is in the range of 15%-20% and the emboli are mostly due to thrombi from the tumor surface. Left sided tumors preferentially embolize to the systemic circulation and, by this, may also cause neurological symptoms [9].

Taken together, tumors of the heart can simulate a variety of cardiac diseases and, thus, many other cardiac conditions which may be confused with these tumors have to be included in the differential diagnosis (Table 1). Consequently, in patients with non-specific signs of 'cardiac disease' accompanied by neurological symptoms and/or typical general symptoms of malignant tumors physical examination should be extended by non-invasive imaging methods focusing on the cardio-pulmonary system.

Compared with extracardiac sarcomas, the prognosis is poor for patients with cardiac sarcoma. This is mostly related to the difficulties in completely resecting the tumor and the proximity of the tumor to vital structures [10]. Not surprisingly, curative operability is best if the tumor is small. Thus, early detection of these tumors is essential and may offer further therapeutic options.

Cardiac tumors are often detected by noninvasive diagnostic methods such as transthoracic echocardiography (TTE), TEE, CT and MRI. Two-dimensional echocardiography can generally be used to determine the location, size, shape and mobility of the tumor. Whenever indicated and possible, TTE should be ex-

Table 1. Differential diagnosis of cardiac diseases often confused with cardiac tumors.

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<th>Structural heart disease</th>
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<td>Valvular heart disease</td>
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<td>(stenosis, insufficiency)</td>
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<td>Atrial septal defect</td>
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<td>Cardiomyopathy</td>
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<td>Infectious heart disease</td>
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<td>Endocarditis</td>
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<td>Myocarditis</td>
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<td>Other conditions</td>
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<td>Constrictive pericarditis</td>
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<td>Carcinoid heart disease</td>
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<td>Mural thrombus</td>
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tended by the transesophageal approach, the latter providing unimpeded visualisation of the atria, the atrial septum, and portions of the left and right ventricles. TEE is therefore particularly helpful in localising the site of the tumor precisely and defining the morphologic features of atrial and ventricular tumors [11]. Moreover, the limit of detection of echogenic structures by TEE is in the range of 1–3 mm in diameter, which produces a higher resolution than CT and MRI (5–10 mm) [12]. MRI, however, is helpful in differentiating tissue composition with the ability to identify solid, liquid, haemorrhagic, and fatty structures. These two imaging techniques are routinely available and should widely be applied, if there is suspicious of an intracardiac tumor. Although in our case surgery was done within 14 days of detection of the cardiac tumor, the patient died about one month after discharge from our hospital. However, dizziness and dyspnea were present nine months before hospitalization and amaurosis fugax occurred two days after admission of the patient. Therefore, TEE should be performed early in the management of systemic – including cerebral – embolic events to assure early identification of potential cardiac sources of emboli, regardless of their type.

In conclusion, TEE is actually the most convenient noninvasive diagnostic tool to exclude intracardiac thrombi or tumors as a source of embolism. A broad application of this method in all cases of arterial embolic events of unknown origin should permit early diagnosis (and then follow-up) of cardiac malignant neoplasm, which is essential for further treatment.

References


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