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Synchronous Primary Heart Liposarcoma and Papillary Renal Carcinoma - a Case Report

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A case of synchronous primary cardiac dedifferentiated liposarcoma and papillary renal carcinoma is presented. The occurrence of typical areas of round cell liposarcoma made the pathological diagnosis of the sarcoma relatively easy; however the neoplasm was not diagnosed correctly before the autopsy. Cardiac liposarcoma is a very rare primary malignant neoplasm and its diagnosis based on image procedures may be extremely difficult especially at non-advanced stage of disease.

Introduction

The human heart is relatively rare location for malignant neoplasms. Among them the most frequent are metastases of carcinomas and primary neoplasms of other chest organs infiltrating secondarily the myocardium. The development of mesenchymal malignant neoplasm originating from tissue being a component of the heart is also possible. The frequency of primary heart mesenchymal tumors found during autopsies was estimated as high as 0.0017 - 0.28% [12] and most of them (about 75%) are benign - myxomas, rhabdomyomas, lipomas, fibromas. Among malignant non-epithelial neoplasms of the heart the most frequent are teratomas, rhabdomyosarcomas and fibrosarcomas in children up to 16 years old and angiosarcomas in adults. Primary heart liposarcoma is extremely rare - about 1% of primary heart sarcomas [5, 6, 12]. Till 1996 only 18 such neoplasms were reported [8]. We present a case of primary heart liposarcoma correctly diagnosed during post-mortem procedure, synchronous with papillary renal carcinoma.

A Case Description

A 58-year-old man was admitted to Cardiology Ward of Municipal Hospital in Kraków because of lower tolerance of exercise, subfebrile condition and non-characteristic chest pain lasting for few days. In the past he had never complained

of circulatory disorder symptoms; for last 3 years he has been treated with oral hypoglycemic drugs. At admission an electrocardiogram revealed atrial fibrillation with ventricular rate of 100, additionally tachypnoe 20 per minute and normal blood pressure were observed. By auscultation bilateral crepitations over lower lung areas were found. The liver was slightly enlarged and bilateral lower leg edema was visible. Laboratory tests showed only mild elevation of glycaemia and hepatic enzymes - AspAT - 78U/l, AlAT - 139U/l. X-ray examination of the chest revealed bilateral blurring of the costophrenic angles due to the presence of pleural effusion and enlargement of the cardiac silhouette. Echocardiography showed normal atrial and ventricular volume, normal contractility of cardiac muscle as well as a pericardial effusion and dense, thicker pericardial echo. The pericardial effusion increased, so two pericardial puncture were performed. During each of that procedures about 1000ml of hemorrhagic fluid were obtained. Cytological examination of pericardial effusion and transudate pleural effusion obtained following drainage punctures did not reveal atypical cells. The patient was firstly treated with salicylates, and then with corticosteroids as viral infection of the heart was suspected. However, during hospitalization deterioration was observed - increasing symptoms of right heart failure, recurrent pericardial effusion, and chest CT revealed tumorous thickening of interatrial septum. Ultrasound and CT examinations of abdominal cavity indicated a 1cm-tumor in the left kidney. Control laboratory tests showed gradual increase of hematocrit value (from 33.6% to 48.3%), hemoglobin concentration (from 11.4g/l to 16.4g/l), erythrocyte and leukocyte count (from 3.96/mm³ to 5.67/mm³, from 8.7/mm³ to 13.9/mm³, respectively), and C-reactive protein (CRP), while blood platelets, serum levels of cholinesterase, total protein and albumins gradually decreased. As the condition of the patient deteriorated he was transferred to the Invasive Pulmonology Unit, Department

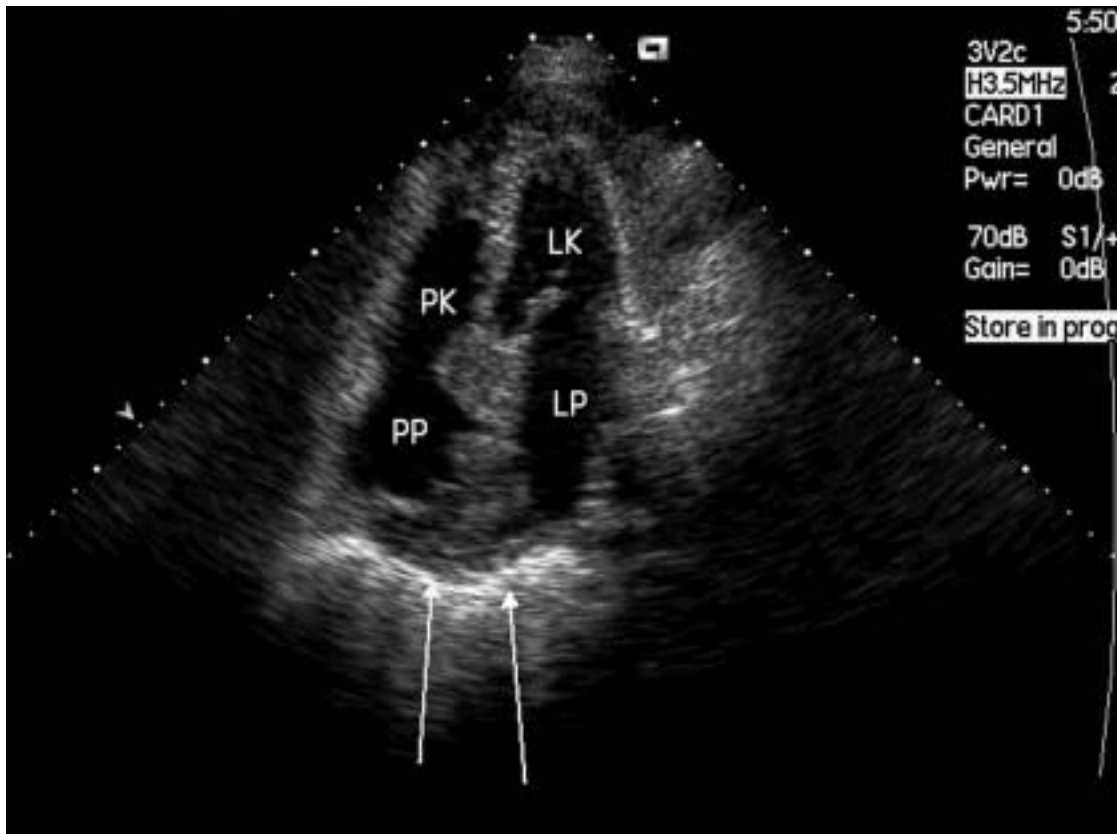


Fig. 1. Transthoracic echocardiography, apical 4-chamber view. Marked irregular thickening of interatrial septum and tissues behind the heart (arrows). PK - right ventricle, LK - left ventricle, PP - right atrium, LP - left atrium.



Fig. 2. CT scan of the chest at the level of the heart. A tumor within the right atrium adhering to the interatrial septum. A pericardial effusion is visible.

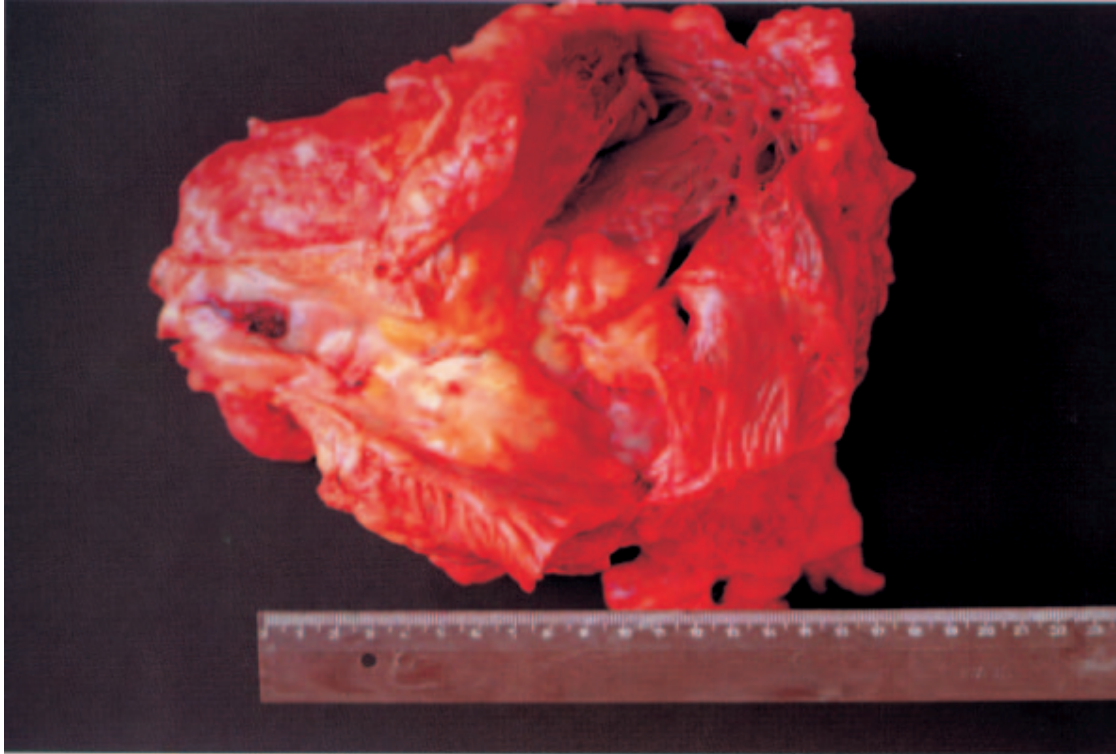


Fig. 3. Opened right heart ventricle and atrium. Yellow-whitish tumorous mass protruding into atrial lumen with distortion of the tricuspid valve (on the left - opened superior vena cava).

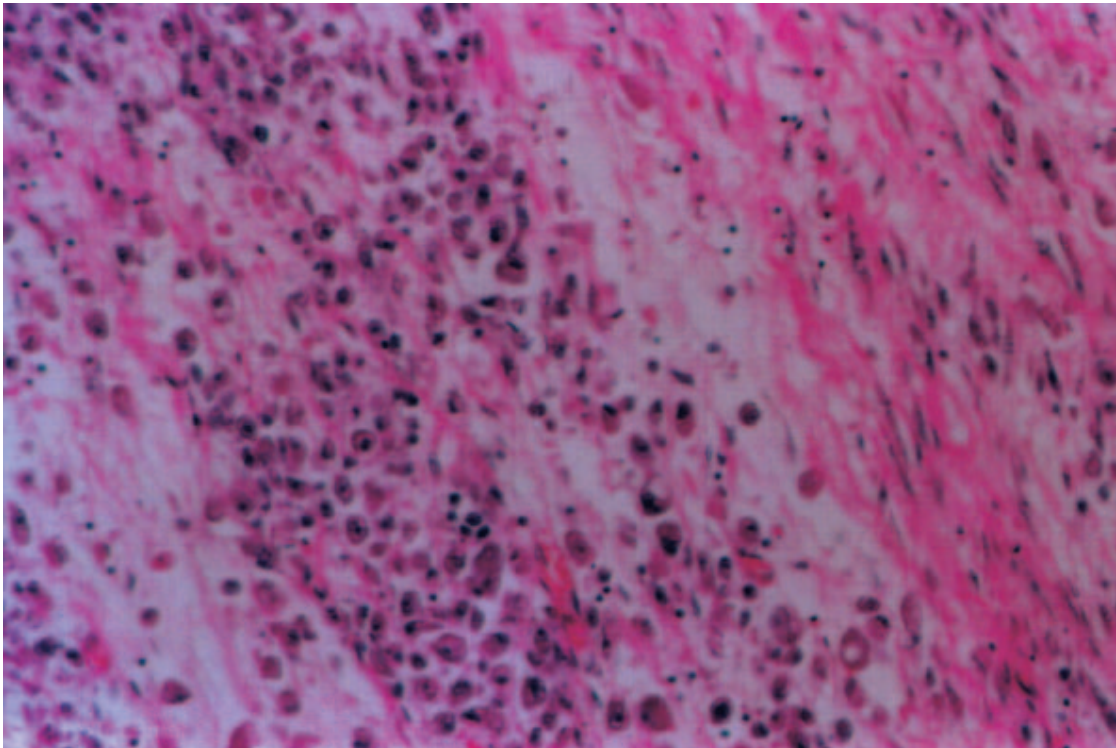


Fig. 4. The tumor composed of round cells. HE.

of Internal Medicine. At admission to the Ward the patient was cachectic and confused. Transthoracic echocardiography and CT showed a tumorous immobile mass located

in the interatrial septum and mediastinum narrowing the left inferior pulmonary vein (Figs. 1 and 2). Mild pericardial effusion and decreased ejection fraction were also found.

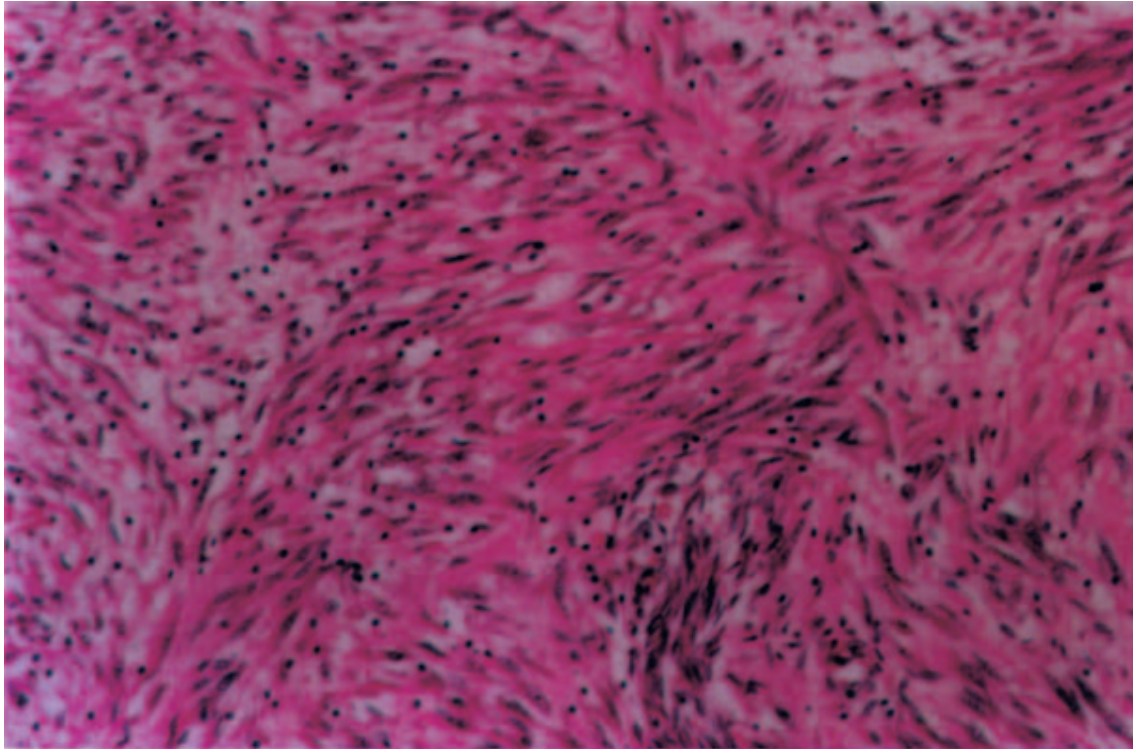


Fig. 5. Spindle cell fibrosarcoma-like pattern of the liposarcoma. HE.

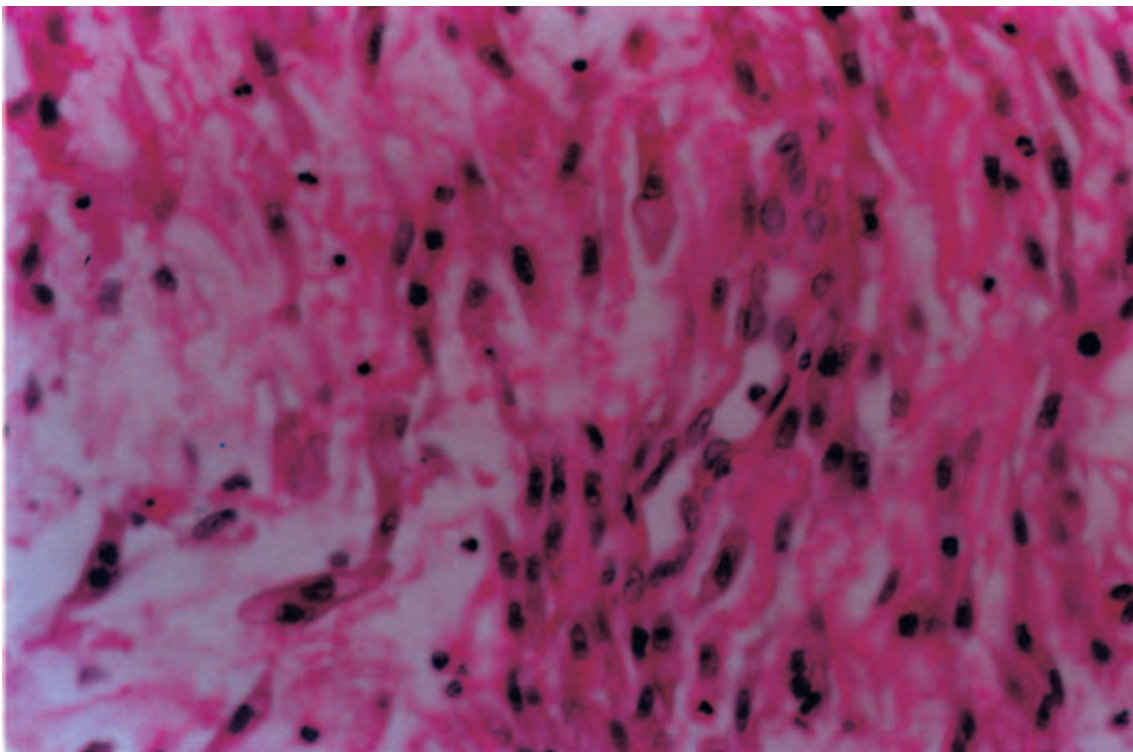


Fig. 6. Empty vacuoles in the cytoplasm of elongated neoplastic cells. HE.

Three days after the admission the patient died and resuscitation efforts were ineffective. The course of the disease was very rapid, ca 3 months from the first symptoms.

The autopsy was performed at the Department of Pathomorphology (No 122554). The pleural cavities contained abundant cloudy, yellowish fluid. Pleura of both lungs was

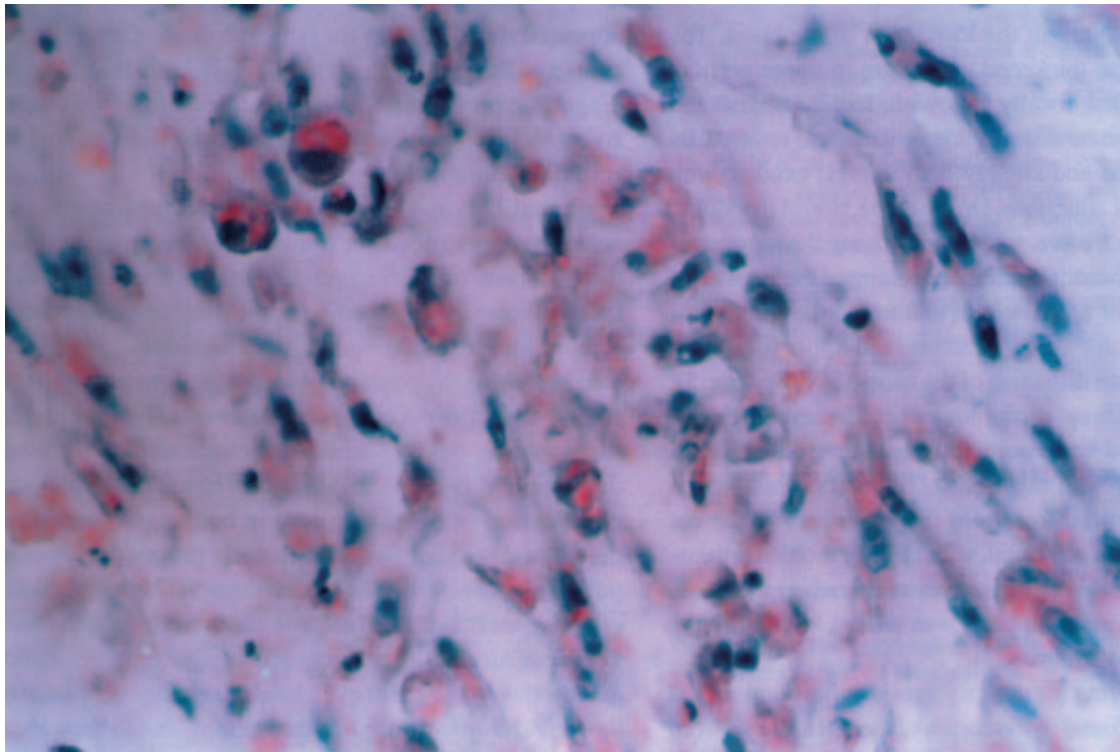


Fig. 7. Fat droplets in the cytoplasm of liposarcoma cells. Sudan.

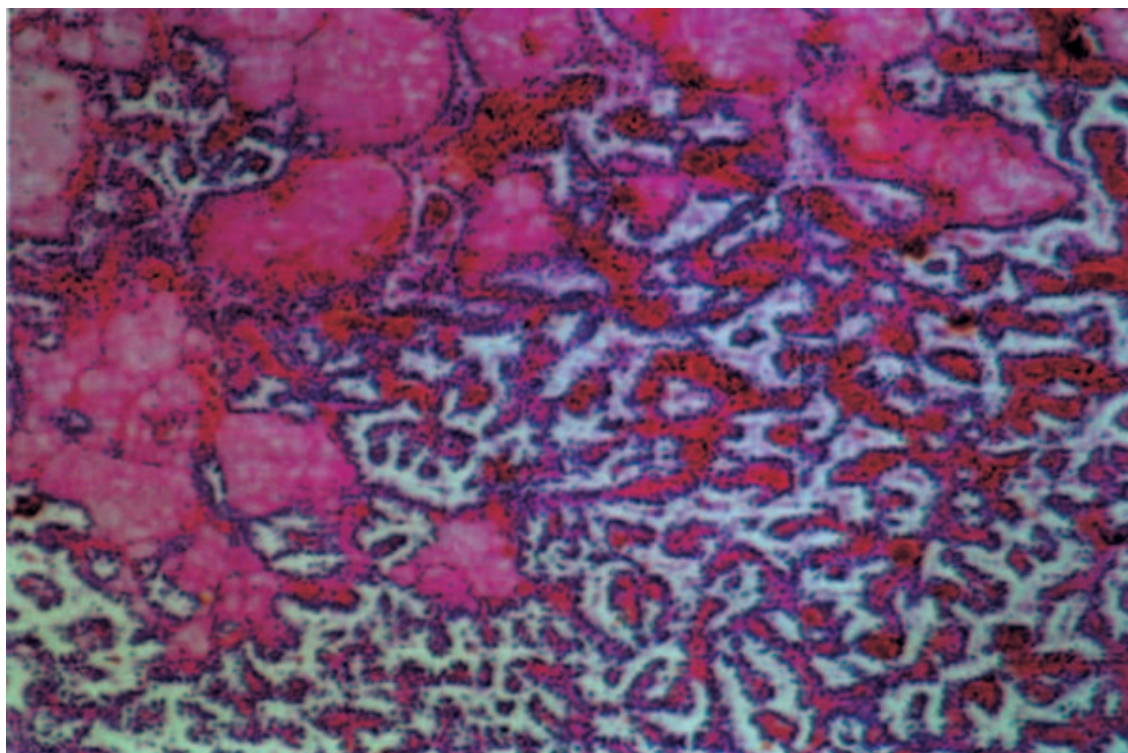


Fig. 8. Classic picture of papillary renal carcinoma - the tumor of the left kidney. HE.

thickened up to 3 - 4mm, especially in lower parts of the lungs, grayish-yellowish in color and covered by fibrinous exudate. Adhesions joined parietal and visceral pleura sur-

faces of lungs as well as the pleura with the pericardial sac. Dispersed nodular infiltrates up to 6mm in thickness, yellowish on the cut section were present within the right

parietal pleura, mainly in the lower part of the chest. The pericardial sac was thickened up to 2cm in upper portion, nodular and mottled, focally yellowish, focally grayish and red with rough visceral and parietal surfaces, covered by abundant fibrinous and slightly hemorrhagic exudate with formation of weak adhesions. On the cut section of it nodular, whitish foci were visible, focally infiltrating the subepicardial layer of cardiac muscle. After opening of the right heart the irregular tumor up to 8cm in diameter was visible protruding into the lumen of right atrium and upper part of the right ventricle, with distortion of the tricuspid valve (Fig. 3). The irregular infiltrate of the lower anterior mediastinum, pericardial sac and lower portions of both lung pleura layers was in continuity with this tumor. The mediastinal lymph nodes appeared to be free from the infiltrate. On the cut section the infiltrate was solid, with slightly mottled appearance - focally white-gray, in another areas rather yellowish, focally myxomatous. The heart was dilated, especially the right ventricle and atrium.

Moreover during the autopsy a solid yellow tumor 2cm in diameter was found in the cortex of the left kidney. From the remaining findings congestion of internal organs as well as of the lungs ("wet shock lung") was noticeable.

Histopathological examination of multiple specimens obtained from the right heart tumor and infiltrate of the mediastinum, pericardial sac and both pleuras revealed malignant neoplasm composed from round and spindle cells with small areas resembling fibrosarcoma (Figs. 4 and 5). The infiltrate was diffuse, without formation of any organoid structures. In the relatively abundant pink cytoplasm of the round neoplastic cells small vacuoles were focally visible (Fig. 6). Differential diagnosis included mainly two neoplasm - liposarcoma and malignant mesothelioma, due to diffuse involvement of serosal surfaces. Immunohistochemically the cells were cytokeratin- and EMA-negative and showed focal positivity for S-100 protein. Intracytoplasmic vacuoles were mucicarmine-negative but orange staining of them was observed in Sudan method, what pointed to the presence of fat droplets (Fig. 7). Final diagnosis of the neoplasm was - liposarcoma, dedifferentiated because of the presence of fibrosarcoma-like areas.

The tumor of the left kidney presented microscopically characteristic pattern of papillary renal carcinoma (Fig. 8). No metastases of it were found.

Discussion

The presented case is noteworthy as primary liposarcoma of the heart is a very rare neoplasm, only single cases were reported [4, 8, 9, 11]. Clinical symptoms and radiological signs of primary heart tumor are not characteristic [1, 3], thus its diagnosis may be difficult in a patient at early stage of the disease. In our case the suspicion of the heart tumor

appeared shortly before the patient's death. Certainly, the course of the neoplasm was relatively rapid, but the first diagnostic procedures did not reveal any tumorous mass. Even so effective image procedure as CT may fail to reveal developing liposarcoma, as its density may be close to normal fatty tissue, and without evident distortion of normal organ structure it is difficult to interpret properly the CT scans of mediastinum and the heart, usually rich in fatty tissue. A lipomatous hypertrophy of the interatrial septum and arrhythmogenic right ventricular dysplasia also belong to fat-containing cardiac lesions and must be considered in differential diagnosis of CT pictures [2]. Liposarcoma belongs to tumors characterized by weak dispersion of cells, so the diagnosis based on cytological examination of neoplastic cells in body cavity effusions may be delayed. However, even early proper diagnosis of primary sarcoma of the heart does not allow usually effective treatment. Only single cases were reported with 1 - 3-year survival after combined therapy - surgery/radio-/chemotherapy [5]. But even benign primary neoplasm of the heart frequently are life-threatening due to their location and the malignant ones are additionally usually aggressive in their behavior, especially when they are characterized by dismal histology as in our case. Most primary sarcomas and pericardial mesotheliomas are diagnosed just during the autopsy [13].

In our case the correct pathological diagnosis was relatively easy as multiple pieces of the tumor tissue were available, encompassing besides the spindle cell areas also round cell neoplasm with intracytoplasmic vacuoles. The differential diagnosis must include malignant mesothelioma because of the location, picture of diffuse infiltration and the biphasic histological pattern. Lack of cytokeratin and EMA expression in neoplastic cells excluded this cancer. Positive Sudan staining appeared to be the proof for the nature of the neoplasm. Because of rarity of primary cardiac tumors the metastatic nature always must be excluded. Though metastases to the heart are more frequent from carcinomas, single cases of metastatic liposarcoma were reported [7, 14]. In our case detailed autopsy examination did not reveal any sarcoma in soft tissues.

A coincidence of primary liposarcoma and renal papillary carcinoma seems to be interesting and an accidental finding. We are unable to find any explanation for the synchronous development of heart liposarcoma and renal carcinoma. The genetic background may involve the same chromosome - trisomy 16 may occur in sporadic papillary renal carcinoma and t(12;16) in myxoid and round cell type of liposarcoma [10]. However the detailed genetics of both tumors is not known and there are no proofs for the involvement of the same region of chromosome 16. So far common risk factors also are unknown.

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