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Radiologic-Pathologic Correlations

Mucin-producing adenocarcinoma arising in an atrial myxoma

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ABSTRACT

We describe the unique autopsy findings of a patient who died of a metastasizing giant right atrial adenocarcinoma containing few areas of typical myxoma. That no mucin-producing extracardiac tumor was detected pointed to the atrial adenocarcinoma as being the primary. We hypothesize that the adenocarcinoma may have developed from coexistent bland glandular structures within the myxoma.

1. Introduction

Cardiac myxomas are mesenchymal neoplasms and represent with about 40% the most frequent benign primary tumors of the heart [1].

Rarely, glandular formations expressing cytokeratins (CKs) are found in cardiac myxomas. The glands have been interpreted as arising from sequestered endodermal elements during ontogenesis in which cardiac structures and the foregut are in close contact [2–7]. We present the autopsy findings of a patient with remnants of a cardiac myxoma with glandular structures entrapped in an adenocarcinoma of the right atrium.

2. Case report

A 61-year-old woman with a history of right-sided heart failure was admitted to hospital because of sudden onset of fever, dyspnea resistant to diuretics, and progressive backache.

On clinical examination, the neck veins were extended, a massive edema of the legs was noticed, and the liver was enlarged, findings that were suggestive of right-sided heart failure.

The radiologic and echocardiographic examinations revealed a large mass in the right atrium.

On the second day after admission, the patient had a pathologic fracture of the left femur diaphysis.

She died of acute right-sided heart failure 5 days after admission. An autopsy was performed.

3. Pathology

3.1. Autopsy examination: macroscopy

The heart weighed 1780 g and occupied nearly the whole thoracic space (Fig. 1). The grotesquely distended right atrium contained an intracavitary solid tumor mass measuring 11 × 11 × 16 cm that protruded through the enlarged tricuspid valve of 17-cm circumference into the right ventricle. The tumor was attached to the septum atriale in the region of the rim of the fossa ovalis and broadly infiltrated the posterolateral atrial wall.

The smooth-surfaced and lobulated tumor was partially covered with a friable reddish thrombotic material.

The gelatinous cut surface showed extensive calcifications, yellow necrotic, as well as gray-white fibrotic areas with spotty hemorrhage. The right ventricle was slightly dilated; the left ventricle was of normal size.

The lungs contained multiple myxoid nodules up to 5 mm in diameter concentrated at the lung bases.

The left adrenal gland contained a whitish nodule measuring 0.7 cm. The left kidney displayed a parenchymal white-yellow nodule measuring 3 cm.

At the site of the pathologic fracture, the corticalis of the femur was thinned and eroded.

The fifth thoracic vertebra showed whitish nodules in the spongiosa.

In the remaining organs, no pathologic findings could be detected. Especially the gastrointestinal organs including the pancreas, the gallbladder and the bile ducts were without pathologic findings.

Furthermore, there was a history of appendectomy in childhood.

The genital tract displayed atrophic changes due to old age; no neoplastic infiltration could be identified.

The breasts were lipomatous without any suspicious indurations or nodules.

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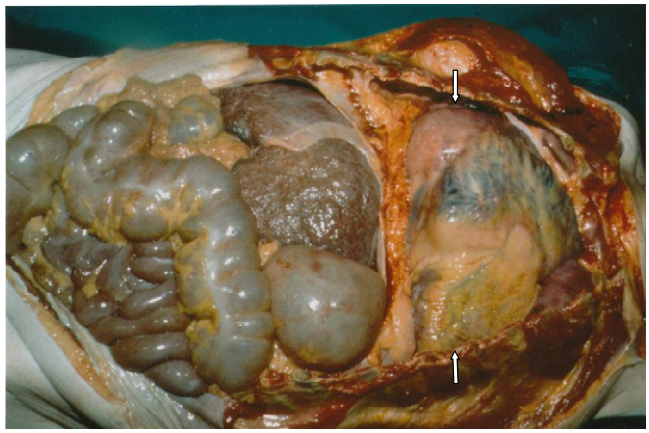

Fig. 1. In situ view of the thoracoabdominal organs with the massive cardiomegaly (arrows) occupying the whole thoracic space.

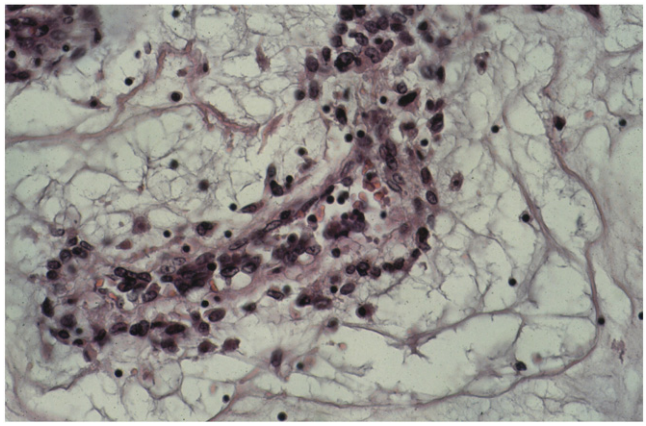

Fig. 3. Typical myxoma with in concentric strands distributed lepidic cells surrounding a central small vascular space (hematoxylin and eosin, ×400).

3.2. Autopsy examination: microscopy and immunohistochemistry

Histologically, the atrial tumor mainly consisted of a mucinous adenocarcinoma displaying tubular and also larger cystic glandular structures and focally irregular epithelial strands that invaded the posterolateral atrial wall (Fig. 2).

On the base of the tumor, adjacent to the atrial septum and at the tumor's surface, areas of a classic myxoma were seen. They encompassed abundant myxoid stroma, with embedded polygonal cells with eosinophilic cytoplasm and round nuclei. These cells were mostly arranged in concentric strands surrounding a central small vascular space or singly in small nests (Fig. 3). Within the myxoma, foci of metaplastic bone with hemopoietic tissue were detected. In addition to the malignant glandular formations of the adenocarcinoma, bland glandular structures with a single layer of cuboidal and cylindrial cells within the myxoma were identified. Therefore, malignant and benign glands could be detected side by side in the small transitional zone between myxoma and adenocarcinoma in the one and only visual field (Fig. 4).

The glandular structures, both benign and malignant, stained strongly with the anti-CK antibody Lu-5 and the carcinoembryonic antigen. The proliferation rate with Ki-67 was less than 10% in the benign glands of the myxoma and about 80% in the malignant glands of the adenocarcinoma and the systemic metastases (Fig. 5).

Immunoreactivity of the adenocarcinoma was diffuse and strongly positive for CK 7 and only focally and weak for CK 20, whereas estrogen and progesterone receptors and thyroid transcription factor 1 were not expressed. The myxoma cells (lepidic cells) stained strongly positive for vimentin and variably for S-100 protein and for neuron-specific enolase. The proliferation rate with Ki-67 was less than 5%.

4. Discussion

A primary mucinous adenocarcinoma of the right atrium was found at autopsy in a 61-year-old woman. Metastases were present in the lungs, adrenal gland, kidney, and bone (Fig. 6).

The existence of a metastatic adenocarcinoma arising from an atrial myxoma was, to the best of our knowledge, so far, not described in the literature. A previous publication from Eckhardt et al [8] described the same case but focusing on radiologic findings.

At admission, plain chest radiography (Fig. 7) revealed a large well-circumscribed intrathoracic mass with coarse calcifications. The heart seemed to be markedly enlarged, but it was not clear whether the tumor was located within the heart. Contrast-enhanced T1-weighted magnetic resonance imaging showed that the tumor was located within the right atrium (Fig. 8). Differential diagnosis of a calcified cardiac mass was made comprising primary cardiac osteogenic sarcoma and cardiac leiomyosarcoma. Other primary cardiac malignancies, such as angiosarcomas, undifferentiated sarcomas, and rhabdomyosarcomas, do not exhibit calcifications. Lung and breast tumors that account for most cardiac metastases usually do not exhibit large calcifications [8,9].

Malignant neoplasms of the heart encompass nearly 25% of cardiac tumors, most of them representing sarcomas [1,10,11]. In contrast to the frequent metastases of carcinomas in the heart, only 2 cases of a

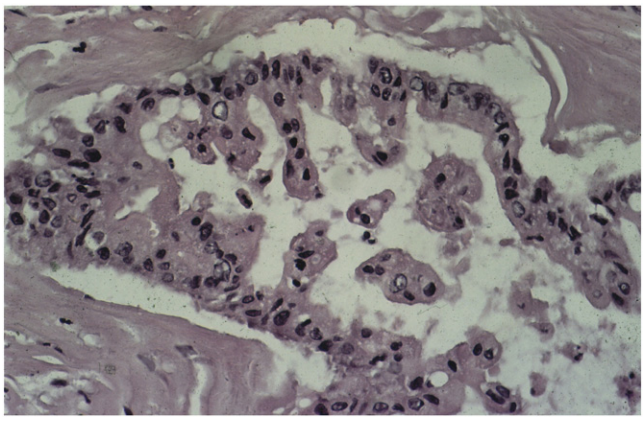

Fig. 2. The mucinous adenocarcinoma shows micropapillary epithelial proliferation with nuclear polymorphism (hematoxylin and eosin, ×400).

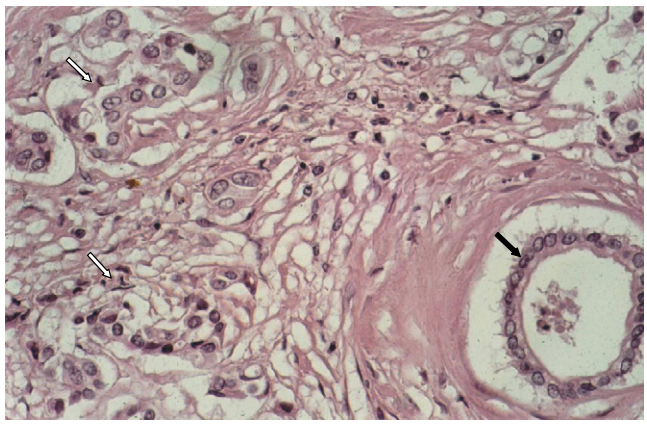

Fig. 4. Tubular and small trabecular structures of the adenocarcinoma (white arrows) in close contact to a benign gland (black arrow). Note the myxoid stroma around the malignant gland and the dense collagenous fibers surrounding the benign gland (hematoxylin and eosin, ×400).