CASE REPORT

Glandular cardiac myxoma

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Abstract: Cardiac myxoma is the most common primary neoplasm of the heart and arises from the endocardium as a polypoid mass, which is commonly seen in the left atrium. Glandular cardiac myxomas are extremely rare cardiac myxomas that contain glandular structures and the pathogenesis of the glandular differentiation is unknown. The present case study reports a case of glandular cardiac myxoma. The tumor was located in the left atrium without invasion of the myocardium. Microscopically, the myxoma contained well-formed mucinous glands in a background of classic cardiac myxoma. Immunohistochemically, the mucinous glandular cells were strongly positive for cytokeratin 7 (CK7) and partially positive for calretinin, but negative for CD34, CK20, D2-40, WT-1, and TTF-1. In conclusion, glandular cardiac myxoma is very rare and it is important to recognize this entity correctly so that it is not misinterpreted as secondary adenocarcinoma.

Keywords: cardiac myxoma; glandular differentiation


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Received: 02nd November 2015; Accepted: 05th February 2016; Published Online: 08th June 2016

Introduction

Cardiac myxomas are the most frequent primary heart tumors in adults. Most cardiac myxomas arise from the endocardium as a polypoid, which often exist as a pedunculated mass in the left atrium1,2. The majority of the myxomas are attached to the atrial septum around the foramen ovale1,2. Clinically, the symptoms can range from being nonspecific and systemic to sudden cardiac death. Cardiac myxomas have a range of appearances including being pedunculated, sessile, polypoid, or globoid. The surface can either be smooth or have multiple finger-like projections.

Most cardiac myxomas are soft with a gelatinous appearance. Microscopically, cardiac cardiac myxomas contain abundant myxoid matrix that is composed of acid mucopolysaccharides, myxoma cells (polygonal cells or lepidic cells) with eosinophilic cytoplasm, indistinct cell borders, and round nuclei. Myxoma cells often form ribbons, nests, and gland-like structures1,2. Hemorrhage, hemosiderin-laden macrophages, lymphoplasmacytic infiltrate, and small vessels are also frequently seen. Calcification or ossification is not uncommon1,2. The myxoma cells are considered to be potential mesenchymal cells that persist as embryonal residues during septation of the heart3,4. They are also thought to arise from cardiomyocyte progenitor cells, subendothelial vasofromative reserve cells, or primitive cells that reside in the fossa ovalis and the surrounding endocardium or endocardial sensory nerve5-8. Occasionally, glandular epithelium may be present in cardiac myxomas (so-called glandular cardiac myxoma), which may represent rests of entrapped embryonic foregut9-10. Glandular cardiac myxomas are extremely rare as there had only been about 50 cases reported in the literature11. The present case study presents a case of glandular cardiac myxoma.

Case report

The patient was a 54-year-old male presented with short-
tness of breath and an echocardiography showing the presence of a mass in the left atrium. The patient had no history of malignancy and Carney complex. During the open-heart surgery, the mass was located in the atrial septum of the left atrium, close to the foramen ovale. The mass was an irregular polypoid, showing brown soft tissue mass mottled with hemorrhage and measured 5.0 × 5.0 × 3.5 cm. The surface was gelatinous. One aspect of it showed fibrous tissue (0.8 × 0.6 × 0.5 cm), which was the stalk of the tumor. Sectioning of the lesion revealed solid, variegated surface with hemorrhage.

Microscopically, the lesion consisted of stellate cells with eosinophilic cytoplasm, indistinct cell borders, and indistinct nucleoli (Figures 1 and 2). There was abundant myxoid ground substance in the stroma. The stellate cells formed ribbons. Hemorrhage, lymphoplasmacytic infiltrates, hemosiderin-laden macrophages, and vascular formation were also present. One area contained morphologically benign, well-formed glands (Figures 1 and 2), partially lined by columnar cells with well-developed apical mucin. The glandular cells showed no cytological atypia. No mitosis or necrosis was identified. These features were consistent with a glandular cardiac myxoma. On immunohistochemistry, the glandular cells were strongly positive for CK7 (Figure 3), focally positive for calretinin (Figure 4), but negative for CK20, D2-40, WT-1, and TTF-1 (images not shown). The special stain mucicarmine highlighted the intracellular mucin (Figure 5).

**Discussion**

Myxomas are the most common primary cardiac tumor...
and a majority of cardiac myxomas are located in the left atrium even though some are occasionally found in the right atrium or ventricles. Most cardiac myxomas are benign and sporadic; however, rare familial cases of malignant and metastatic cardiac myxomas have also been reported\textsuperscript{[4,12,13]}. There was no history of Carney complex in our case and the patient had no history of malignancy.

Previous studies have shown that the myxoma cells have some similarities with mesothelial cells. The expressions of both epithelial and vascular antigens reflect the multi-potential nature of myxoma cells\textsuperscript{[1,2]}. The incidence of glandular cardiac myxomas is very low and its histogenesis is unclear. Totipotent cardiomyogenic precursor cells and embryonal residues in the tumor have both been suggested as the origin of the glands\textsuperscript{[10,13]}. Our case showed well-differentiated mucinous glandular structures. The glandular cells were immunohistochemically positive for CK7 and calretinin indicating that they were probably of entrapped foregut rest origin. The differential diagnosis of glandular cardiac myxoma includes metastatic adenocarcinoma. Our case did not show cytological atypia, mitosis, or necrosis that is usually seen in adenocarcinomas. Furthermore, there was no myocardial invasion in our case.

In conclusion, glandular cardiac myxoma is extremely rare and it is important to correctly recognize this entity in order to prevent its misinterpretation as adenocarcinoma. Currently, there are relatively few reports of malignant glandular cardiac myxoma. Nonetheless, patients with glandular cardiac myxomas should be followed up periodically.

**Conflict of interest**

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

**References**