Cardiac Myxoma with Glandular Component

Glandüler Komponentli Kardiyak Miksoma

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Abstract
Glandular component is a very rare finding in myxomas which are one of the most common primary benign cardiac tumors. This rate was found ranging 1% to 5% in various series. They were found more common in middle aged women. Histogenesis of myxomas with glandular component is not known clearly. Cardiac myxomas with glandular component could show recurrence and malignity so they had to be followed. We presented a myxoma case with glandular component which was located in left atrium in 38 years old woman in this report.

Keywords: **Myxoma.**

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Özet
Glandüler komponent, primer benign kardiak tümörlerin en yaygınlarından biri olan miksomalarda oldukça nadirdir. Değişik serilerde oran %1–5 arasında bulunmuştur. Orta yaşlı kadınlarda sık görülmektedir. Glandüler kompenentli miksomaların histogenezi açık değildir. Glandüler komponentli miksomalar rekürrens ve maliniteye dönüşüm gösterebilmeleri nedeniyle yakından takip ve kontrol edilmelidirler. Biz bu yazıda 38 yaşında kadın olgu sol atriumda yerleşmiş glandüler komponentli miksoma vakasını sunduk.

Anahtar kelimeler: **Miksoma.**

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Introduction

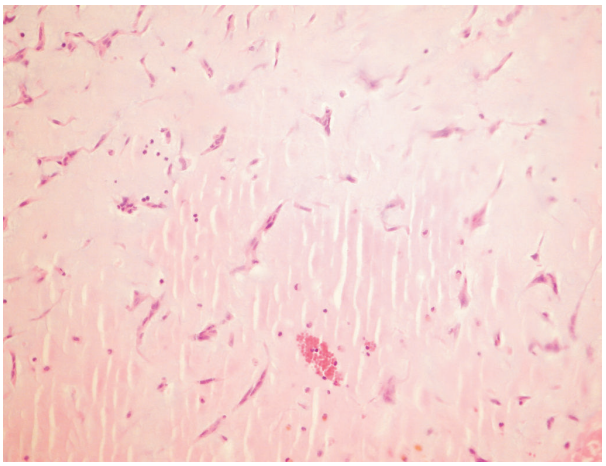
Cardiac myxomas are most common primary cardiac tumor in adults (1). They constitute approximately 50% of primary cardiac tumors. They were found more common in middle aged women. Glandular cardiac myxomas are subgroup of myxomas and histogenesis of myxomas is not well known (2-4). The case is being reported due to its rarity in cardiac myxoma as well as its glandular differentiation, which must be recognized as a spectrum of histomorphologic diversity.

Case Presentation

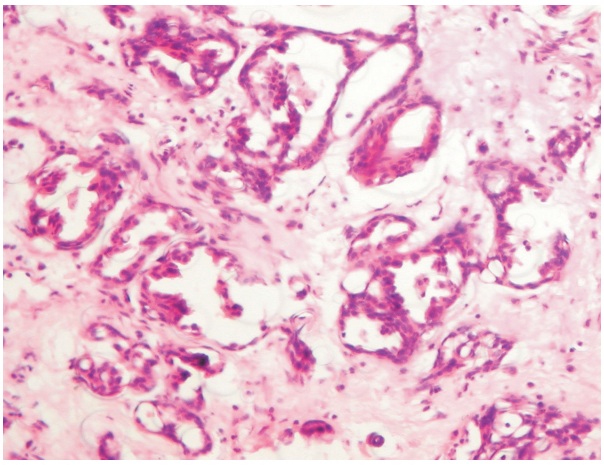
A 38 years old **female** was shifted from a local hospital to our emergency room with 30 minutes duration of sudden, severe crushing left sided chest pain radiating to back and chest. Physical examination and electrocardiography (ECG with or without effort) findings were normal. Coronary arteries were normal. A mass with a size of 6.8x3.9cm, adjacent to septal wall of left atrium, was excised by opening left atrium at open surgery conditions. Patient was discharged after 10 days from operation without any significant post-operative complication. Follow up examination was normal at the day of 10^{th}.

Pathological Findings. Macroscopic examination showed a loose lobulated tumor with incomplete capsular appearance in left atrium. It was 4.5x3.5x1.5 in size. The tumor was fragile and the color was yellow and dark brown. The cut surface was semitransparent, gelatinous and hemorrhagic at some fields. In microscopic examination, myxoma cells that appear round, stellate and spindle-like at some areas were observed in myxoid, fibrinoid stroma. Cells had oval nucleus and eosinophilic cytoplasm (Picture 1). Hemorrhagic areas, hemosiderin laden macrophages, neutrophil, leukocytes and lymphocytes were also observed in myxoid stroma. Glandular arrangement of tumoral cells was only observed in focal fields (Picture 2). In this fields, mucinous containing tubular structures those were laid down with irregular one fold cubic epithelium that was dispatched with columnar epithelium had been observed. Epithelium cells had round nucleus, marked nucleolus and eosinophilic cytoplasm. Nuclear atypia, mitosis and necrosis were not seen.

The glandular structures showed diffuse positivity for epithelial membrane antigen (EMA), B72.3, cytokeratin 7 (CK7), cytokeratin 8 (CK8) (Picture 3). Additionally, the glandular epithelium showed strong positivity with periodic acid-schiff alcian blue (PAS AB) and high iron diamine (HID-AB) staining.



Picture 1. Round polygonal stellate cells are seen surrounded by a highly myxoid stroma.20xHE



Picture 2. Glandular differantiation.20xHE

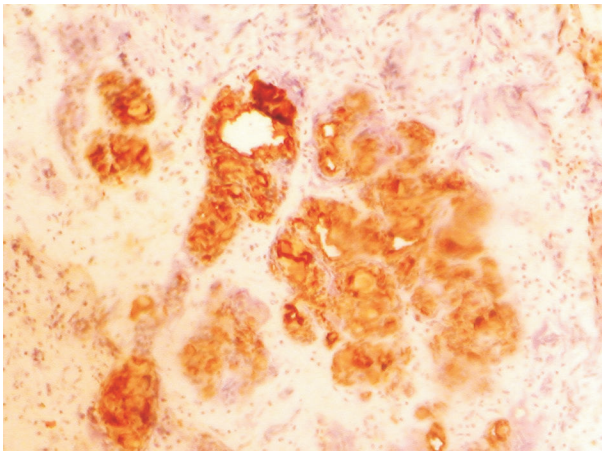


Figure 3. CK8immunohistochemical stain.10xHE.

Discussion

Myxomas are most common primary cardiac tumor. They were accounted nearly half of primary cardiac tumors in adults (1). It was most commonly found in left atrium (%75-%80) and then respectively in right atrium and ventricle. It was more frequently found in middle aged women. Familial cases were reported rarely (3, 4). There was not a family history in our case. Myxomas should have some variable symptoms and findings such as syncope, palpitation, shortness of breath and souffle which were depended on localization, size and whether it had stalk or not. Most of the patients could be diagnosed as intracardiac myxoma by echocardiography before operation (3).

Incidence of cardiac myxomas with glandular component has been found ranging from 1% to 5% in myxamatous stroma (4-7). Histogenesis of myxomas with glandular component is not clear (2, 4). Totipotent cardiomyogenic precursor cells and embriyonal residues in tumor have been suggested as origin of glands (2). There is a well differentiated gland structures that secretes mucinous in the fields. Glandular arrangement show positive staining with pancreatin, EMA and different cytokeratin subtypes (CK8, CK18, CK7, CK20) staining. In glandular cells, acidic mucin stained by PAS AB and sialomucin by HID AB (2, 3, 4, 8, 9, 10) In our case, glandular component showed positive staining with EMA, CK7 and CK8 while it stained towards acidic mucin satain by PAS AB and sialomucin stain by HID AB.

Light microscopic, immunohistochemical and histochemical findings suggest that glandular epithelium should have gastrointestinal or enteric origin (3, 5). Differential diagnosis should be done with metastatic adenocarcinomas that produces mucinous (8). We did not come across with nuclear atypia, mitosis and necrosis which should be seen in metastatic adenocarcinomas in light microscope findings.

Patient did not have any significant post-operative complication. Follow up examination was normal at day 10 after discharge.

The effect on the prognosis by the occurrence of glandular differentatiation has not statistical data because glandular differantiation is rare in cardiac myxoma. However recurrence or metastasis has been reported in literatures (3, 4, 6). Long-term postoperative follow-up and serial echocardiography are advisable especially for young or familial patients. It was reported that metastatic cases are due to embolism that contains tumor fragments. It was reported cardiac myxoma cases with recurrent cerebral metastasis (11).

Consequently, even though rarely, cardiac myxomas with glandular component could show recurrence and malignity so they had to be followed. Glandular component is a very rare finding in myxomas which are one of the most common primary benign cardiac tumors. They had to be followed for recurrence and metastasis.

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