Gastrointestinal stromal tumors (GISTs) are primarily mesenchymal tumors with specific histological features, located in the gastrointestinal tract and the abdomen. They are identified and diagnosed by the expression of the protooncogene protein called CD117 detected using immunohistochemistry. Atrial myxoma is the most common benign primary tumor of the heart. Carney triad is considered to be a specific type of multiple endocrine neoplasia. Three classically related tumors are the GIST, pulmonary chondroma, and extra-adrenal paraganglioma. Carney syndrome is an autosomal dominant syndrome with multiple components, characterized by myxomatous neoplasms (cardiac, endocrine, cutaneous, and neural), pigmented lesions of the skin and mucosa, and multiple endocrine tumors. The patient in this study was a 75-year-old man with no known illness. He had a tumor in the stomach and was operated. The pathology was reported as low-grade GIST; therefore, adjuvant imatinib treatment was not recommended. Further, owing to the shortness of breath in the third month of follow-up, echocardiography was requested. Left atrial mass was detected and operated. His pathology was compatible with atrial myxoma. After both operations, the patient is still being followed up in remission in our clinic. Only one case has been reported worldwide documenting the coexistence of atrial myxoma and GIST, and none has been seen in Turkey. Thus, to the best of our knowledge, this is the second case in the literature and first report from Turkey.

Keywords: Atrial myxoma; gastrointestinal stromal tumors

Gastrointestinal stromal tumors (GISTs) are primarily mesenchymal tumors with specific histological features, located in the gastrointestinal tract and the abdomen. They are identified and diagnosed by the expression of a protooncogene protein called CD117 detected using immunohistochemistry. Atrial myxoma is the most common benign primary tumor of the heart, and atrial myxomas are extremely rare tumors.

Normally, interstitial cells of Cajal regulate gastrointestinal peristalsis in the bowel wall. These cells are found in and around the intestinal myenteric plexus in the adult bowel. GISTs are mesenchymal tumors that originate from precursors of Cajal cells.

GISTs may be present in all parts of the gastrointestinal tract, but most often in the stomach (50%) and the small intestine (25%). In addition, the tumor can rarely be located in the colorectal (10%), omentum/mesentery (7%), and esophagus (5%).

GISTs of 2 cm or less are typically asymptomatic and incidentally detected during surgery, radiology, or endoscopy for any other reason. The most common symptoms are abdominal pain, gastrointestinal bleeding, anemia, abdominal mass, dyspeptic complaints, and dysphagia.

Most of the primary cardiac tumors (80%) are benign, and the most common benign heart tumor is atrial myxoma. Although myxomas may develop from all cardiac cavities, they are sporadic tumors and most commonly localized in the left atrium (75%). These tumors, which can be seen in all ages and more frequently in women, may occur with cerebral or peripheral embolism findings, and patients
may present with clinical findings such as mitral stenosis. In addition, they may cause myocardial infarction with coronary artery embolization and endocarditis.\textsuperscript{7-10}

Carney triad is a specific type of multiple endocrine neoplasia. Three classically related tumors are the GIST, pulmonary chondroma, and extra-adrenal paragangliomas.\textsuperscript{11}

Carney syndrome is an autosomal dominant syndrome with multiple components. It is a group of diseases characterized by myxoma (cardiac, endocrine, cutaneous, and neural); skin pigmentation; endocrine hyperactivity; Sertoli cell tumor, which is the other tumor of the testis; psammomatus melanotic schwannoma; pituitary adenoma; and thyroid tumors.\textsuperscript{12} Carney syndrome is most frequently associated with mutations in the protein kinase A type I-alpha regulatory subunit gene (\textit{PRKAR1A}). The diagnostic criteria for Carney syndrome were defined previously. Major diagnostic criteria are spotty skin pigmentation with typical distribution, myxoma (cutaneous, mucosal, or cardiac), acromegaly because of growth hormone-producing adenoma, thyroid carcinoma, psammomatus melanotic schwannomas, blue nevus, epithelioid blue nevus, breast ductal adenoma, osteochondromyxoma, primary pigmented nodular adrenocortical disease, and primarily large-cell calcifying Sertoli cell tumor of the testis.\textsuperscript{13}

Based on these major clinical findings, three diagnostic criteria have been defined: when two and more major criteria are present, and a pathogenic variant is identified in the \textit{PRKARIA}, or one major criterion is present, and a first-degree relative has Carney syndrome or an inactivating mutation of \textit{PRKARIA}.\textsuperscript{14} Although atrial myxomas may be sporadic, they may also be considered a component of Carney syndrome.

In this study, a patient with GIST and left atrial myxoma is discussed. The patient was diagnosed with low-grade GIST, and left atrial myxoma. This situation neither complied with Carney syndrome nor with Carney triad. The exact genetic association is unknown. This case may represent an incidental finding or may be a variant of a syndrome such as Carney syndrome or triad.

CASE REPORT

Our patient was a 75-year-old man with no known crucial medical and family history. He was admitted because of early satiety and reflux symptoms. In addition, he had complains of dyspnea. The patient underwent endoscopy, and no endoscopically significant pathology was detected. On abdominal ultrasonography, a mass of 4x5 cm was detected on the outer surface of the greater curvature of the stomach. It was confirmed by positron emission tomography-computed tomography (PET-CT) imaging. Subtotal gastrectomy was performed to the patients who had no evidence of distance metastasis in PET-CT. The lesion was reported as GIST in stage T2. Immunohistochemical staining with CD117 and CD34 showed positive results. Ki-67 proliferation index was 2% (Figure 1a, Figure 1b). No lesions were observed on the surgical margins. No tumor rupture was observed macroscopically, and tumor diameter was 4.5x4.5x4 cm. Therefore, it was regarded as low-risk GIST and followed-up. Adjuvant imatinib treatment was not considered.

The patient was still experiencing shortness of breath. Chest X-ray film and electrocardiogram were normal. The transthoracic echocardiography revealed a 2x2 cm moving mass in the left atrium. The mass was confirmed with CT-angiography. The mass was excised because of the suspicion of metastasis on the patient who had known malignancy. The patient was reoperated to remove the cardiac mass approximately four months after the first gastric operation. The pa-

![FIGURE 1a: Gastrointestinal stromal tumor; stomach (HE, x100): Some tumor cells with clear cytoplasm, epithelioid appearance and some spindle palysate were observed.](image-url)
Patient underwent successful resection of the left atrial mass, which was of 1.7x1.6 cm size on gross examination. After cardiac mass excision, the pathology report of the case was as follows: immunohistochemical calretinin (positive), S-100 (positive), PanCK (negative), desmin (negative), and smooth muscle antibody (negative) (Figure 2a, Figure 2b). Thus, the pathologic diagnosis of the lesion was evaluated as the histopathologic findings consistent with myxoma. There was no known history of Carney syndrome in the first-degree relatives of the patient.

The patient was evaluated to have a low-risk GIST at approximately three months with upper abdominal ultrasonography and echocardiography without drugs after the excision of the two masses. During the one-year follow-up, the patient is asymptomatic and has no signs of recurrence. The patient is still being followed up in remission in our clinic.

**DISCUSSION**

GISTs are primarily mesenchymal tumors with specific histological features, located in the gastrointestinal tract and the abdomen. Normally, interstitial cells of Cajal regulate gastrointestinal peristalsis in the bowel wall. These cells are found in and around the intestinal myenteric plexus in the adult bowel. GISTs are mesenchymal tumors that originate from precursors of Cajal cells.

The standard treatment for the early-stage GIST is the primary resectable surgery with negative microscopic margins. After complete resection, adjuvant treatment is recommended for high-risk GIST. In adjuvant therapy, imatinib, a tyrosine kinase inhibitor, is the mainstay.

Atrial myxomas are most commonly observed in the left atrium. According to a recent meta-analysis, 83% of cardiac myxomas are noted in the left atrium and 12.7% in the right atrium. Cardiac myxomas are sporadic and mostly solitary. However, as in Carney syndrome, coexistence with multiple tumors can be observed. Sporadic cardiac myxomas and cardiac myxomas associated with Carney syndrome cannot be differentiated histologically.

Atrial myxoma is a component of Carney syndrome, but in our patient, there were no other features. In addition, there was no known history of Carney syndrome in the first-degree relatives of the patient. The presence of the PRKAR1A mutation was unknown. Therefore, our patient did not meet the di-
agnostic criteria of Carney syndrome. GIST is a component of Carney triad, but again, no other features for Carney triad were noted in our patient. To the best of our knowledge, only one case has been published worldwide.\textsuperscript{30}

In that case, high-grade GIST and atrial myxoma were present together. In this study, a patient with low-grade GIST associated with left atrial myxoma is described, which is the first known case in the literature. It is unclear whether there is a coincidental association or it is a component of syndromes such as Carney syndrome and triad.

In conclusion, the exact genetic relationship between the two tumors is unknown. To better understand this relationship, more cases should be reported in the future.

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\section*{Conflict of Interest}

No conflicts of interest between the authors and/or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

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