CASE REPORT OLGU SUNUMU

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A Primary Right Atrial Leiomyosarcoma in a 59-Year-Old Female Patient

59 Yaşında Kadın Hastada Primer Sağ Atrial Leiomyosarkom

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ABSTRACT Leiomyosarcoma is a mesenchymal tumor originating from smooth muscle cells and constitutes a small proportion of the primary cardiac tumors. The tumor usually tends to settle on the left side of the heart and infiltrates the pulmonary veins, causing chest pain, dyspnea, and heart failure. Since primary cardiac leiomyosarcoma is a rare entity, it is difficult to distinguish clinically from myxoma, which is the most common benign tumor of the heart. Therefore, histopathological examination is necessary. The prognosis is poor due to the high rate of postoperative recurrence and distant metastasis. In this study, a case of primary cardiac leiomyosarcoma originating from the right atrium in a 59-year-old female patient is presented.

Keywords: Leiomyosarcoma; right atrium; myxoma; prognosis; differential diagnosis

ÖZET Leiomyosarkom düz kas hücrelerinden köken alan mezenkimal bir tümördür ve primer kalp tümörlerinin çok az bir kısmını oluşturur. Tümör genellikle kalbin sol tarafına yerleşme eğilimi gösterir ve pulmoner venleri de infiltre ederek göğüs ağrısı, dispne ve kalp yetmezliğine neden olur. Primer kardiyak leiomyosarkom nadir görülen bir antite olduğundan kalbin en sık görülen benign tümörü olan miksomadan klinik olarak ayrımı zordur. Bu nedenle histopatolojik inceleme gereklidir. Postoperatif nüks ve uzak metastaz oranı yüksek olduğundan prognoz kötüdür. Bu çalışmada 59 yaşında kadın hastada sağ atriyumdan köken alan primer kardiyak leiomyosarkom olgusu sunulmuştur.

Anahtar Kelimeler: Leiomyosarkom; sağ atrium; miksoma; prognoz; ayırıcı tanı

Primary cardiac tumors are rare tumors detected between 0.001% and 0.03% in autopsy series. The majority of primary cardiac tumors are benign and are often atrial myxomas, but up to 25% are malignant. Angiosarcomas are the most common primary malignant tumors, followed by rhabdomyosarcoma, mesothelioma, and fibrosarcoma. Among all cardiac sarcomas, leiomyosarcoma is rarely seen (approximately 8% of all cases).[1,2] In this study, we presented a case of primary cardiac leiomyosarcoma originating from the right atrium in a 59-year-old female patient.

CASE REPORT

A 59-year-old female patient with a history of diabetes and hypertension presented with complaints of difficulty in walking and weakness for a week. Abdominal ultrasonography (USG) and thorax computed tomography (CT) revealed bilateral pleural and pericardial effusion, therefore the patient was referred to our institution for further investigation of the effusion etiology. Transthoracic echocardiography revealed a 5x3.5 cm mass originating from the entrance of the pulmonary arteries. No extension to the vena

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cava was observed. Mild to moderate tricuspid regurgitation was observed. Transesophageal echocardiography (TEE) revealed a 7.1x4.6 cm mass in the right atrium consistent with myxoma. The patient was operated and the hard mass originating from the right atrium fossa ovalis was removed and tricuspid annuloplasty was performed.

Macroscopic examination of the 7x5x2.5 cm sized material showed an off-white tumoral mass with gelatinous consistency and well-defined solid areas. When the material was sliced, the cut surface was off-white-yellow in color and soft in consistency. In the microscopic examination, tumoral tissue with spindle-like bundled cells lined in different directions was observed. Atypia level in the tumor ranged from mild to moderate. In parts of increased cellularity and atypia, mitosis was observed 4 in 1 high power field. No necrosis was detected (Figure 1). In addition, scattered mononuclear type inflammatory cell infiltration within the tumor was noted. The tumor did not have a good border with the surrounding endocardial tissues. In the immunohistochemical examination, the tumor was vimentin and desmin positive, and smooth muscle actin (SMA) were focally positive in the limited foci. CD34, CD117 and S100 were negative. Ki67 was 30% positive (Figure 2).

With these findings, the case was reported as Grade 2 leiomyosarcoma.

Informed consent was obtained from the patient who participated in this case.

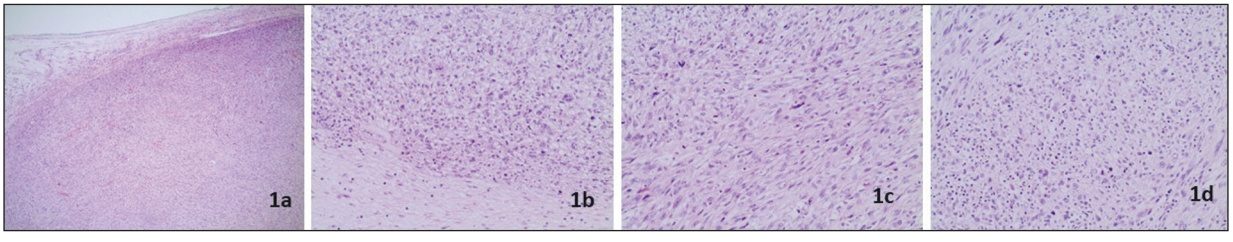

FIGURE 1: Tumoral tissue with the spindle-like cell bundles surrounded by endocardial tissue was observed in different directions (**1a**, H&E, x40). The level of atypia in the tumor ranged from mild to moderate and, atypical mitoses were detected (**1b-1c,** H&E, x200). In addition, scattered mononuclear type inflammatory cell infiltration was observed within the tumor (**1d,** H&E, x200).

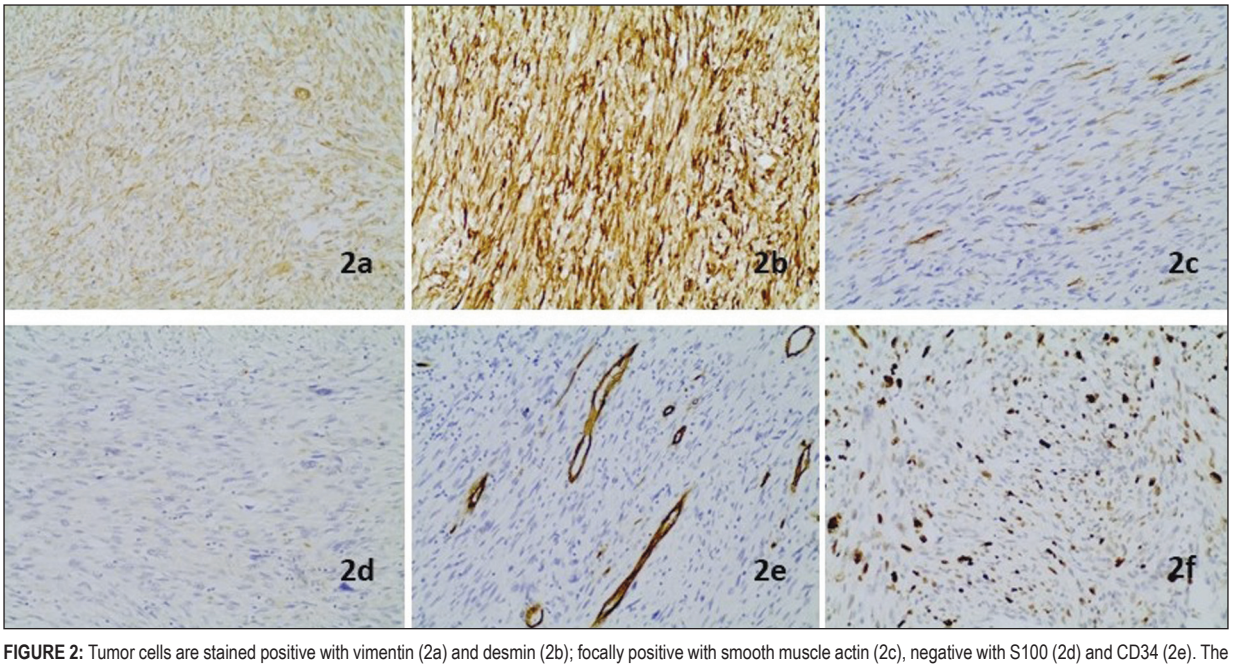

FIGURE 2: Tumor cells are stained positive with vimentin (2a) and desmin (2b); focally positive with smooth muscle actin (2c), negative with S100 (2d) and CD34 (2e). The Ki67 (2f) proliferation index was found to be positive at a rate of approximately 30% (x200).

DISCUSSION

We describe a case of leiomyosarcoma originating from the right atrium fossa ovalis a distinct localization.[1] Fewer than 50 cases of cardiac leiomyosarcomas are reported in the literature.[1-9] Leiomyosarcoma typically occurs in the fourth decade of life, slightly earlier than the mean age of patients with cardiac sarcoma.[3] Leiomyosarcoma is a mesenchymal tumor originating from smooth muscle cells. As seen in other cardiac sarcomas, it tends to be in the left side of the heart and is often located in the left atrium. Thus, infiltrating the pulmonary veins; causing chest pain, dyspnea, pericardial effusion, and eventually heart failure.[1,4] In clinical investigation of the patient we observed pleural and pericardial effusion in the abdominal USG and CT.

Since it is difficult to distinguish between benign atrial myxoma and primary cardiac leiomyosarcoma clinically, the diagnosis should be confirmed by histopathologically and immunohistochemically.[1] Macroscopically, the tumor appears as a gelatinous mass and may be multifocal in up to 30% of cases.[3] In the 24 primary cardiac sarcoma case series of Donsbeck et al., they found that the majority of tumors microscopically consisted of spindle-like cell bundles and a small portion of oval cells. They also detected significant nuclear pleomorphism, peri-tumor lymphoplasmocytic infiltration, and fibrous acellular areas in the stroma.[5] In our case, the mass was grossly gelatinous and consisted of microscopic spindle-like cell bundles. The level of atypia in the tumor ranged from mild to moderate. Mitosis increased in parts of increased cellularity and no necrosis was detected.

Primary immunohistochemistry is needed to perform differential diagnosis of cardiac sarcoma. In immunohistochemical study, it is diagnostic that tumor cells of leiomyosarcoma show positive staining with SMA and desmin.[1] Studies also support this staining prototype.[5,6] Unlike these studies, Kornberg et al. found positive staining with SMA in tumor cells while only some cells were positive with desmin.[7] In our case, positive staining with desmin was observed in the tumor, and positivity with SMA was detected in limited foci, contrary to what is reported in the literature.

Echocardiography (ECHO) is an extremely useful and noninvasive imaging method for screening suspected cardiac tumors and determining tumor differentiation.[7] Many features of the tumor were defined using ECHO, CT, and magnetic resonance imaging (MRI). Among these findings, tumor immobility, presence of pericardial effusion, broad-based tumor and neovascularity are in favor of malignancy.[8] In cases where imaging methods are insufficient, definitive histological diagnosis of the tumor with preoperative transvenous or percutaneous biopsy allows tumor-specific treatment planning.[7] In our case, the TEE report was a mass compatible with myxoma in the foreground in the right atrium, however the differential diagnosis of myxoma from sarcoma could not be made.

Surgery is the primary therapy for leiomyosarcoma. The tumor may not be totally excised due to its localization in the heart.[9] Therefore, complete macroscopic resection can be performed in only 33% of patients who are suitable for surgery.[3] In addition, it has a high rate of local recurrence and distant metastasis after resection due to its rapid growth capacity. Considering the possibility of incomplete tumor excision and metastasis, adjuvant chemotherapy and radiotherapy may be considered. The superiority of chemotherapy and radiotherapy to each other is controversial.

Prognosis is affected by the tumor localization, extension of the tumor to the heart chambers, tumor grade and presence of distant metastases. About half of cardiac sarcomas have metastasized at the time of the diagnosis, more than 30% are multifocal, and the rest are cases with no chance of surgical resection.[3] It was observed that our patient was asymptomatic during the 8-month follow-up period. No recurrence or metastasis was detected in the ECHO and CT follow-ups.

In conclusion, primary cardiac tumors are rare and highly aggressive tumors. There are limited case reports on cardiac leiomyosarcomas. It may not be differentiated preoperatively from atrial myxoma, which is the most common tumor of the heart. For this ECHO, CT, MRI and when necessary, histopathological examination should be used together. Thus, the follow-up and treatment of the patient can be carried out in accordance with the diagnosis.

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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.

Authorship Contributions

Idea/Concept: *Şeyma Öztürk;* ***Design:*** *Şeyma Öztürk;* ***Control/Supervision:*** *Çetin Boran;* ***Data Collection and/or Processing:*** *Şeyma Öztürk, Çetin Boran;* ***Analysis and/or Interpretation:*** *Çetin Boran;* ***Literature Review:*** *Şeyma Öztürk;* ***Writing the Article:*** *Şeyma Öztürk;* ***Critical Review:*** *Çetin Boran;* ***References and Fundings:*** *Çetin Boran;* ***Materials:*** *Şeyma Öztürk, Çetin Boran.*

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