Fever, dyspnea and chest pain with pericardial effusion

Ateş, dispne, göğüs ağrısı ile birlikte perikard sıvısı

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Right answer: 2. Primary cardiac angiosarcoma

MRI showed a heterogeneous cardiac mass accompanying pericardial effusion, 6.5 x 3.5 cm within size. The mass was surrounding the right atrium from posterior, lateral and inferior walls and invading the atrioventricular sulcus and the inferior vena cava wall (Fig. 3A, B). Delayed and heterogeneous enhancement of the mass was also noticed during cardiac MRI. With these findings, the patient underwent cardiac surgery with sternotomy and pericardiotomy. During the operation, a tumoral tissue was observed covering the right atrium and extending to the inferior vena cava (Fig. 4). A small fragment of the tumor was sent for intraoperative cytological and histopathological examination. Frozen section and imprint cytology were performed. Using modified May Grunwald Giemsa (MGG) stain the imprint smears obtained were found to be cellular mainly consisting of irregular, anisocytotic spindle-shaped atypical cells with oval hyperchromatic nuclei (Fig. 5A). After confirmation of the mesenchimal nature of the tumor, partially surgical resection was performed due to locally advanced tumor. Macroscopically, the surgical specimen obtained was a 3x3x2cm soft, red colored mass with irregular lobulated edges. The cut surface of the tumor was red-brown and hemorrhagic with variegated tan-brown solid areas and necrosis. The specimen was fixed in 10% buffered formalin, and processed routinely for paraffin embedding. Four micrometer-thick sections were stained with hematoxylin and eosin (HE). Microscopic examination of the HE stained sections revealed vessels of varying sizes and exhibited sinusoid structures composed of abnormal proliferations of malignant endothelial cells (Fig. 5B, C). Due to the fragmented nature of the resected specimen surgical margins could not be evaluated. Immunohistochemically, the tumor cells were positive for vimentin, Factor VIII, CD34 (Fig. 5D) and negative for desmin, calretinin, S-100, smooth muscle actin, C-kit, confirming the diagnosis of angiosarcoma. Ki-67 proliferation index was 40%. With these morphological findings the diagnosis of angiosarcoma was made. Further investigations for detecting metastases to other sites were negative. The patient was referred to an oncology clinic for further planning of chemotherapy. She underwent chemotherapy with paclitaxel (Dose of 80 mg/m^3 was administered weekly). Despite the persistence of residual cardiac mass with regression, the systemic symptoms disappeared completely. The patient is continuing this regimen and she is still alive at 7 months after the surgery.

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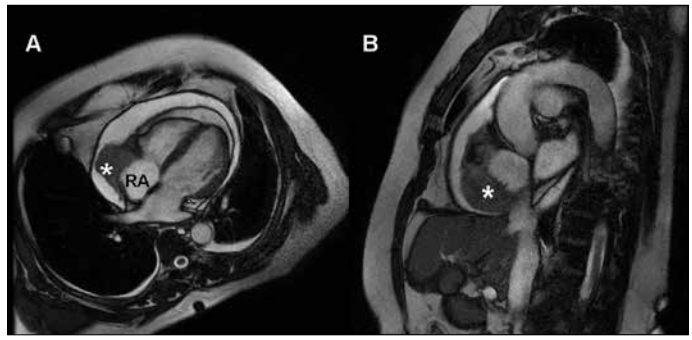

Figure 3. A) Cardiac MRI reveals the mass (asterisk) surrounding the right atrium from posterior, lateral and inferior walls and invading the atrioventricular sulcus in the axial plane. B) In the sagittal plane the tumor (asterisk) invades the inferior vena cava wall and shows extension to the root of aorta superiorly

MRI - Magnetic resonance imaging, RA - right atrium

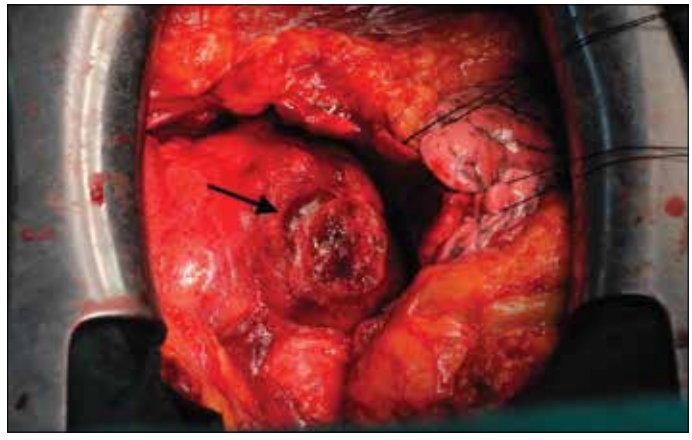

Figure 4. The appearance of the mass (black arrow) during cardiac surgery

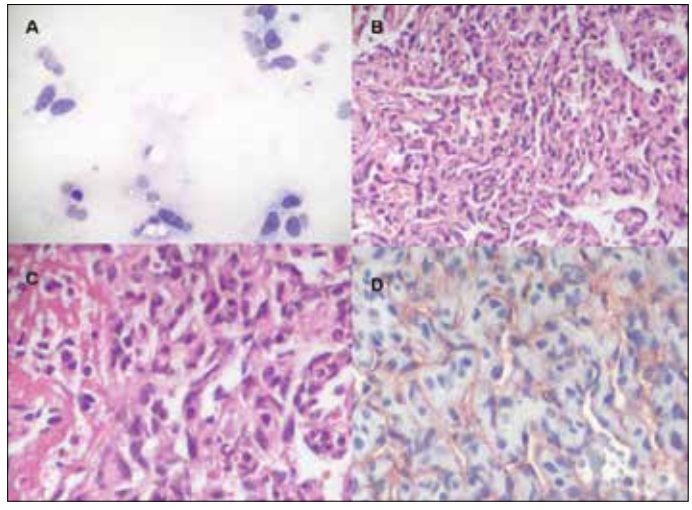

Figure 5. A) The imprint smears show loosely connected atypical mesenchymal cells with oval hyperchromatic nuclei, undefined cytoplasmic borders, and several cytoplasmic projections. (Imprint smear of angiosarcoma. Anisocytotic spindle-shaped tumor cells). B) H&E section shows vasoformative malignant neoplasm with predominantly spindled tumor cells and variable nuclear pleomorphism. C) There is a pleomorphic tumor with necrosis. The tumor shows endothelial differentiation. (HE) D) İmmunohistochemical staining for CD34 shows endothelial differentiation