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Atrial angiosarcoma imaged by F-18 FDG PET/CT

F-18 FDG PET/CT ile görüntülenen atriyal anjiyosarkom

Sarcomas arising primarily in the heart are rare. Angiosarcoma is an extremely rare, most common primary cardiac malignant tumour. We report the F-18 fluorodeoxyglucose positron emission tomography (F-18 FDG PET) / computed tomography (CT) images of a patient with an invasive angiosarcoma arising from the right atrium. A 55-year-old woman with primary high-grade cardiac angiosarcoma underwent tumour resection at another centre was referred to our institution for postoperative F-18 FDG PET/ CT scans to assess the residual/metastatic disease. Preoperative magnetic resonance imaging revealed a 6.5x3.5-cm mass extending along the lateral wall of the right atrium surrounding the inferior vena cava and atria ventricular junction, extending to aortic root as well as into the right pericardium. Concomitant pericardial effusion was noticed. The debulking surgery was performed to remove the mass. Four weeks after surgery, the patient underwent the F-18 FDG PET/CT scans (1hour after the administration of 465 MBq F-18 FDG with the subject fasted for 6 h beforehand). Three-plane images (PET, contrast-enhanced CT and fused PET/CT in axial, sagittal and coronal projections) revealed a residual mass with significantly increased uptake of F-18 FDG (SUVmax of 16.4) in the right atrium measuring 5.0x4.5 cm in size (Fig. 1, Video 1, 2. See corresponding video/movie images at www.anakarder.com). There was no evidence of distant metastatic disease. The patient subsequently underwent chemotherapy regimen.

Still, primary cardiac sarcoma is a rare clinical entity, with an incidence of 0.0001% in collected autopsy series. The majority of patients with cardiac sarcomas presents with unresectable tumours and have a poor prognosis. Prognosis of primary cardiac angiosarcoma is generally poor with usually a short and fatal course: the mean survival for patients with primary cardiac angiosarcoma is 9-12 months following diagnosis. Treatment options for these sarcomas include surgery, chemotherapy, and radiation therapy, alone or in combination. Complete resection of cardiac sarcoma is difficult, in view of the location and extent of involvement. PET with F-18 FDG, an analogue of glucose, provides valuable functional information based on the increased glucose uptake and glycolysis of cancer cells and depicts metabolic abnormalities. F-18 FDG PET/CT acquires PET and CT data in the same imaging

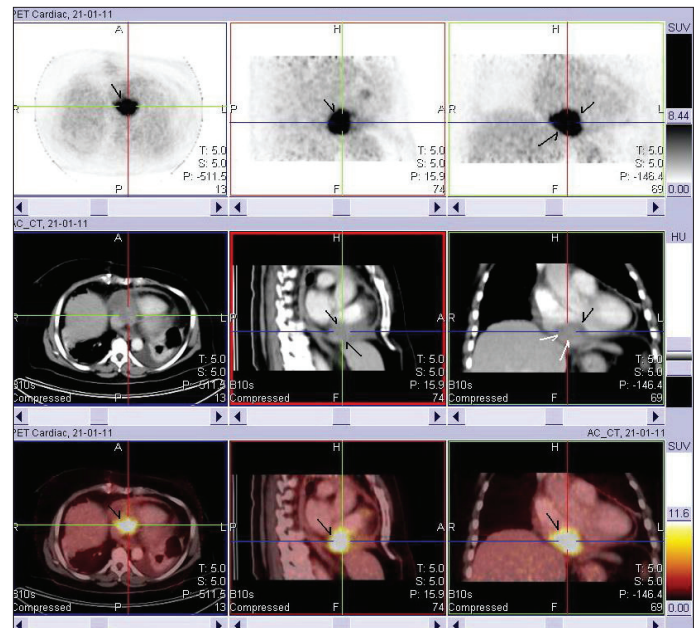


Figure 1. PET (top), contrast-enhanced CT (middle) and fused PET/CT (bottom) images in axial, sagittal and coronal projections of a residual mass in the right atrium

CT - computed tomography, PET - pozitron emission tomography

session and allows accurate anatomical localization of the lesions detected on the PET/CT scan.

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Naxos-Carvajal disease: a rare cause of cardiomyopathy with woolly hair and palmoplantar hyperkeratosis

Naxos-Carvajal hastalığı: Palmoplantar keratozis ve yünsü saç ile karakterli nadir bir kardiyomiyopati nedeni

Naxos-Carvajal disease is a rare autosomal recessive inherited disease characterized by a triad of ventricular dysplasia/dilated cardiomyopathy, woolly hair and palmoplantar hyperkeratosis. The pathological process is characterized by progressive loss of myocardial fibrils