Yasin Türker, Mustafa Kayan[1], Mehmet Munduz[1], Selçuk Yaşar[1], Mehmet Özaydın[2]
Department of Cardiology, Faculty of Medicine, Düzce University, Düzce,
[1]From Departments of Radiology, and [2]Cardiology, Faculty of Medicine, Süleyman Demirel University, Isparta-*Turkey*

Address for Correspondence/Yazışma Adresi: Dr. Yasin Türker
Department of Cardiology, Faculty of Medicine, Duzce University, Düzce-*Turkey*
Phone: +90 380 542 13 90 Fax: +90 380 542 13 87
E-mail: dryasinturker@hotmail.com

Available Online Date / Çevrimiçi Yayın Tarihi: 18.05.2011

doi:10.5152/akd.2011.098

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

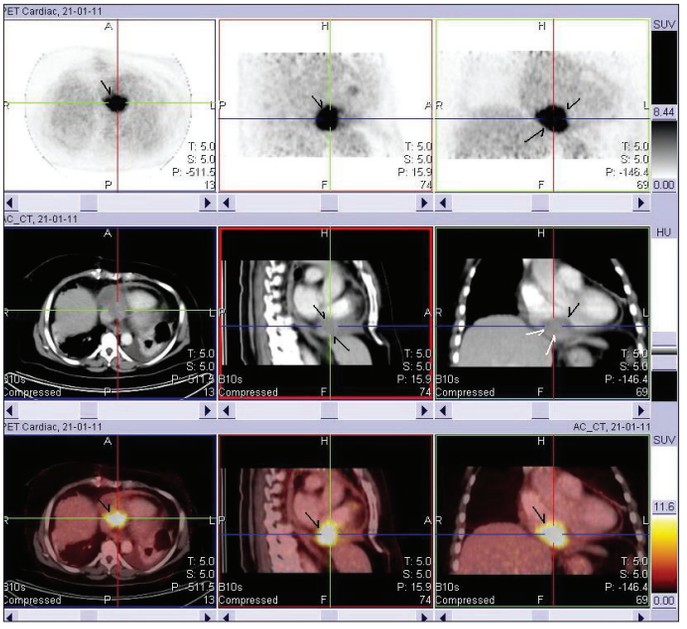

Figure 1. PET (top), contrast-enhanced CT (middle) and fused PET/CT (bottom) images in axial, sagittal and coronal projections of a residual mass in the right atrium
CT - computed tomography, PET - pozitron emission tomography

İlknur Ak, Öznur Dilek Çiftçi, Zeki Üstünel[1], Muammer Cumhur Sivrikoz*
From Departments of Nuclear Medicine and *Thoracic Surgery, Faculty of Medicine, Eskişehir Osmangazi University, Eskişehir
[1]Clinic of Oncology, Eskişehir Ümit Hospital, Eskişehir-*Turkey*

Address for Correspondence/Yazışma Adresi: Dr. İlknur Ak
Department of Nuclear Medicine, Faculty of Medicine, Osmangazi University 26480 Eskişehir-*Turkey*
Phone: +90 222 239 29 79 Fax: +90 222 229 11 50
E-mail: ilknur_ak@yahoo.com

Available Online Date / Çevrimiçi Yayın Tarihi: 18.05.2011

doi:10.5152/akd.2011.099

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

Anadolu Kardiyol Derg
2011; 11: E15-E18

and replacement with fibro-fatty tissue. We present a 3-year-old case of Naxos-Carvajal disease who is to our knowledge the youngest patient in literature.

Physical examination revealed diffuse palmoplantar hyperkeratosis and curly hair, which was present from the birth. On physical examination, blood pressure was 84/40 mmHg, heart rate 112/min and gallop rhythm. Chest X-ray revealed distinct cardiomegaly with pulmonary congestion. The baseline electrocardiograms showed sinus tachycardia with decreased QRS amplitudes. Echocardiographic examination revealed dilatation of the left and right heart and global hypokinesia with a left ventricular ejection fraction of 20%. The patient expired from cardio-pulmonary arrest after 4 days.

A postmortem examination was performed. The heart was heavy (196 gr) from twice the normal and the both ventricles were dilated (Fig. 1). External surface of the heart was yellowish brown. Fibro-fatty replacement was observed especially on both ventricular regions (Fig. 2). The histopathological alteration lied in perpendicular from ventricles to the left atrium. Cardiac myofibrils showed destruction with the apoptosis and degenerative changes. Confluent compact hyperkeratosis and slight irregular acanthosis were observed at the skin of the palmoplantar region (Fig. 3).

Naxos-Carvajal disease should be kept in mind, in cases of a patient presenting with undetermined dilated cardiomyopathy from Mediterranean, Arabic and Ecuadorian regions.

Ragıp Ortaç, Vedide Tavlı*, Gülden Diniz, Murat Muhtar Yılmazer*, Savaş Demirpençe*
From Clinics of Pathology and *Cardiology, Dr. Behçet Uz Children Hospital, İzmir-*Turkey*

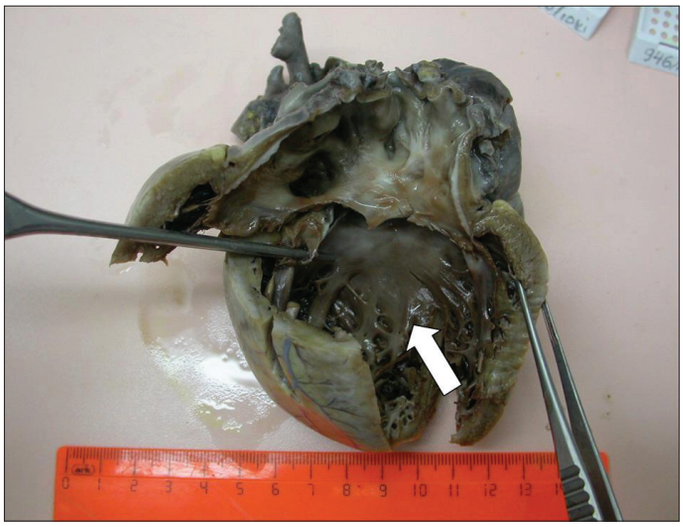

Figure 1. Pathology view of ventricular dilatation

Address for Correspondence/Yazışma Adresi: Dr. Ragıp Ortaç
1420 Sok No:78 D:1 Alsancak, İzmir-*Turkey*
Phone: +90 232 463 89 80 Fax: +90 232 463 89 80
E-mail: rortac@gmail.com

Available Online Date / Çevrimiçi Yayın Tarihi: 18.05.2011

doi:10.5152/akd.2011.100

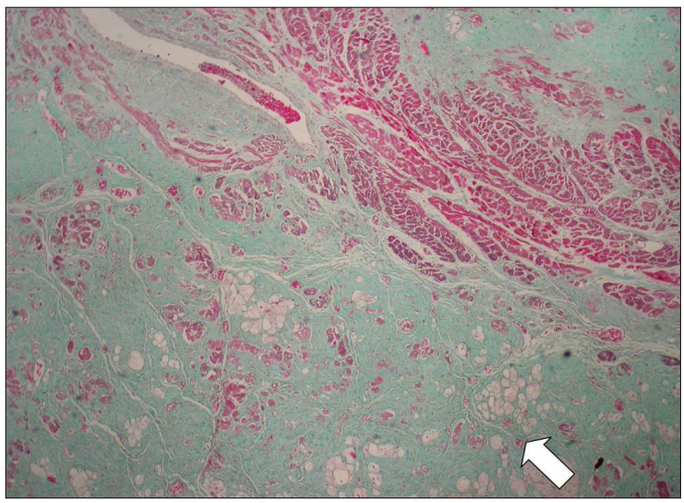

Figure 2. Histopathologic view of fibrofatty replacement of the myocardium (x40, Gomori's Trichrome)

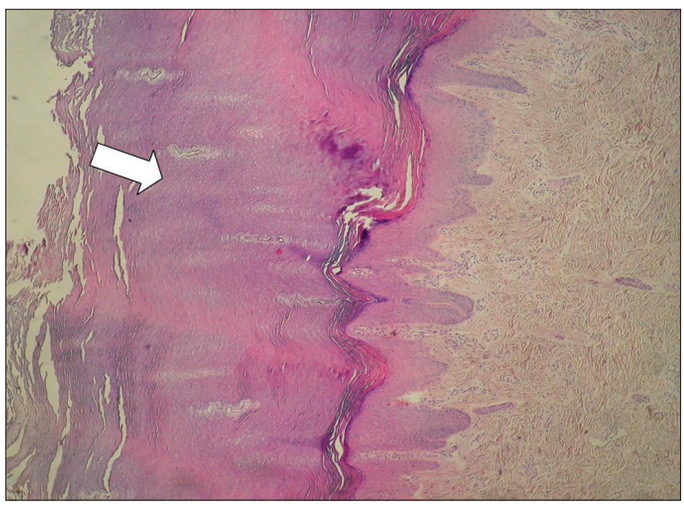

Figure 3. Histopathologic view of hyperkeratosis of the skin sample from plantar area (x40 HE)