Atrial angiosarcoma imaged by F-18 FDG PET/CT

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 atrioventricular 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 (1 hour 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 (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.

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

Naxos-Carvajal hastalığı: Palmoplantar keratoz 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
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.

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Figure 1. Pathology view of ventricular dilatation

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)