Tangier Disease With Continuous Massive and Longitudinal Diffuse Calcification in the Coronary Arteries
Demonstration by the Sagittal Images of Intravascular Ultrasonography

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A 48-year-old man was first referred to our clinic in January 1989 because of marked hypocholesterolemia with very low HDL cholesterol, anemia, and hyperbilirubinemia. He had large tonsils, corneal opacities, hepatosplenomegaly, and thrombocytopenia. Serum levels of total cholesterol, triglycerides, and HDL cholesterol were 0.72, 2.6, and 0.16 mmol/L (28, 232, and 6 mg/dL), respectively. Concentrations of apolipoproteins (apo) A-I and A-II were 1.3 and 0.9 μmol/L (3.9 and 1.5 mg/dL), respectively. His daughter’s serum levels of total cholesterol, HDL cholesterol, and apo A-I were 3.3 mmol/L, 0.64 mmol/L, and 34.7 μmol/L (128, 24, and 104 mg/dL), respectively. He was diagnosed with Tangier disease by clinical manifestations, analysis of lipoproteins, and 2D electrophoresis, which con-

Figure 1. Puncture sample of bone marrow stained with Sudan III and analyzed by light microscopy shows foam cells with accumulated lipid droplets (arrow). Bar=10 μm.

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firmed the increase of preproapo A-I. Interestingly, xanthoma in the Achilles’ tendons, which had rarely been reported in patients with Tangier disease, was observed in the patient, and the thickness was 9 mm (mean of bilateral determinations; control, 6±2 mm). Subsequently, the diagnosis of homozygous Tangier disease was also established by biopsy of the patient’s bone marrow, showing the presence of foam cells (Figure 1).

Our patient began having exertional chest pain in January 1997. Myocardial perfusion images with 201 Tl revealed a defect in the inferoposterior wall with an incomplete redistribution; thus, a coronary angiogram was performed in July 1997. It revealed massive and longitudinal diffuse calcifications in the 3 coronary arteries that could be seen only on the scout radiograph (Figure 2A and 2C). We also found severe atherosclerosis in all 3 vessels, including the left main trunk (LMT); 90% stenosis in the mid portion of the right coronary artery (RCA) (segment 2), 75% stenosis in the LMT (segment 5), 75% stenosis in the proximal portion of the left anterior descending coronary artery (LAD) (segment 6), 75% stenosis in the mid portion of the LAD (segment 7), 90% stenosis in the just proximal portion of the left circumflex artery (LCx) (segment 11), and 90% stenosis in the mid portion of the LCx (segment 13), respectively (Figures 2B, 2D, and 3).

We performed intravascular ultrasonography (IVUS) simultaneously with the coronary angiogram, and it demonstrated notably that the atherosclerotic plaque with intimal thickening and its superficial calcification protruded toward the center of the lumen in cross-sectional IVUS images (Figure 3, I and II). Conversely, only a very thin plaque was observed on the opposite side of the protrusion (Figure 3, I and II). Consequently, the lumen there, especially in II, appeared crescent-shaped (Figure 3, red trace). We then obtained different sagittal sections of 3D-reconstructed IVUS images to analyze the coronary arteries more precisely. It was also noteworthy that massive and longitudinal diffuse calcifications extended continuously from distal to the second diagonal branch to the LMT (Figure 3, bottom right, white arrows).

Figure 2. Coronary angiogram. A and B, Left anterior oblique projections of right coronary artery. C and D, Right anterior oblique caudal projections of left coronary artery. Arrows indicate longitudinal and massive diffuse calcifications of coronary arteries in scout films (A and C). In RCA, 90% stenosis was observed in mid portion (B). Significant stenosis is also shown in proximal and mid portions of LAD and in proximal and mid portions of left circumflex artery (D). Moreover, 75% stenosis is seen in left main trunk (D).
Figure 3. Coronary angiogram (left anterior oblique cranial projections) and IVUS images of LAD. Black arrows indicate longitudinal and massive diffuse calcifications in scout films (top left). Left circumflex artery and first diagonal branch (D1) are shown in 4 o’clock position in cross-sectional IVUS images (top right, I and II). Both cross-sectional IVUS images, especially II, demonstrate atherosclerotic plaque with intimal thickening and its superficial protruded calcification in 2 o’clock position. Only a very thin plaque is seen on opposite side. Lumen there, especially in II, appears crescent-shaped (red trace). Differential sagittal sections of IVUS images 3D-reconstructed by NetraIVUS (Scimage Inc) (bottom right) demonstrated massive and longitudinal diffuse calcifications extending continuously from distal to second diagonal branch (D2) to left main trunk (bottom right, white arrows).
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