Coronary Artery Features of Immunoglobulin-G4-Related Coronary Periarteritis With Multi-Modality Visualization

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Immunoglobulin-G4-related disease is a rare systemic fibroinflammatory condition with multiple organ involvement. A 55-year-old woman was diagnosed with IgG4-related coronary periarteritis (IgG4-RCP) based on elevated serum IgG4 concentration (2,010 mg/dL), lymphocytes and plasma cell infiltration on mediastinal tumor biopsy; increased 18F-fluorodeoxyglucose uptake in the mediastinal tumor (Figure A) and in soft tumors surrounding the coronary arteries (Figure B, C) on positron emission tomography computed tomography (CT); and 20–40 mm soft tumors around the left ascending and circumflex arteries (yellow and red arrows) with calcified lesions on CT angiography (Figure D). Coronary angiography showed severe stenosis with calcified and dilated lumen lesions (Figure E). Optical coherence tomography (OCT) and intravascular ultrasound (IVUS) showed a layered signal pattern with microchannels (red arrowheads, Figure F-2, 3) in the coronary periarteritis and coarse isoechoic lesions without a normal 3-layered tunica (yellow arrowheads, Figure G-2, 3). Despite mild atherosclerotic changes, as evidenced by low-grade yellow plaque on coronary angioscopy (Figure H), there were calcified stenotic lesions (Figure F-I, G-1), indicative of long exposure to the inflammatory processes of periarteritis. After steroid therapy, periartrial soft-tissue thickening decreased to 10–25 mm, the coronary stenosis had regressed mildly, but the intimal surface/internal periarteritis remained unchanged (Figure I-M). Therefore, steroid therapy suppressed the coronary periarteritis externally, leading to mild regression of the coronary stenosis, but its effect might have been decreased for coronary calcified stenotic lesions due to the long-term inflammation status. This case may provide clinical insight into the mechanism of the coronary atherosclerotic process and emphasizes the importance of early therapeutic intervention in IgG4-RCP.

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