1085 Cardiac amyloidosis followed by chronic Ischemic heart disease

A 76 year-old man presented with dyspnea. He had a hypertension and well controlled diabetes (Hemoglobin A1C : 5.9). His resting electrocardiogram (ECG) showed sinus rhythm, low voltage in limb leads and frequent ventricular premature beats. Transthoracic echocardiogram showed mild left ventricular hypertrophy (1.13 cm) and right coronary artery territory hypokinesis with ejection fraction 50-55%, restrictive pattern of left ventricle diastolic function.

Previously, he had coronary angiography 2 times. At first time, there was triple vessel disease with total occlusion in the mid segments of the left anterior descending (LAD) artery, 90% distal stenosis in the left circumflex (LCX) artery and 90% proximal stenosis in the right coronary artery with chronic total occlusion of the distal segments. Percutaneous coronary intervention was performed on the mid segments of the LAD artery. 12 years after the first procedure, significant in-stent restenosis in distal edge of the LAD stent was found and second stent was deployed in that segment a 8 month ago.

He has had a pain and limitation of motion of tongue and poor diet. Several mucosal biopsy were performed to identify the reason at other hospital. But, there was no accurate diagnosis of the lesion of tongue so far. After admission, he also complained a pain and limitation of motion of tongue. After examined by otolaryngology department, a tongue biopsy stained with Congo Red yielded a diagnosis of amyloidosis.

Imaging study was performed for evaluating a whether cardiac involvement of amyloidosis. PET CT showed heterogeneous hypermetabolism in heart, but can’t rule out physiologic uptake or amyloidosis involvement. Cardiac magnetic resonance imaging showed delayed gadolinium enhancement of basal, mid septal wall and mid inferolaeral wall that positive finding of amyloidosis.

In most patients, angina pectoris, acute myocardial infarction, and other manifestations of ischemic heart disease are related to atherosclerosis of the major, extramural coronary arteries. However, clinically significant myocardial ischemia and infarction can result from several conditions in the absence of coronary atherosclerosis. Nevertheless, symptomatic ischemic heart disease resulting from obstructive intramural coronary amyloidosis is not a well-documented or recognized entity. Cardiac amyloidosis may also present as an ischemic syndrome with angina pectoris or myocardial infarction.

Although He had no pathologic evidence of severely obstructive amyloidosis of the small intramural coronary arteries with associated myocardial ischemic changes, the association between amyloidosis and ischemic heart disease should be considered because the sameness of involved myocardial territory evaluated by cardiac MRI and coronary angiography.

The diagnosis of obstructive intramural coronary amyloidosis should be considered when obstructive epicardial coronary atherosclerosis is excluded and biopsy or laboratory evidence of systemic amyloidosis is obtained. Appropriate treatment of the cardiac ischemic syndrome should be considered within the context of management of the underlying systemic amyloidosis.