A Case of ST-Elevated Myocardial Infarction Resulting From Obstructive Intramural Coronary Amyloidosis

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SUMMARY

A 49-year-old man presenting with ST-elevated myocardial infarction was brought to our emergency department with AL amyloidosis. Baseline coronary angiography showed no significant stenosis of the epicardial coronary arteries, however, coronary artery angiography in response to acetylcholine and coronary flow reserve in response to papaverine were abnormal, which suggested impairment of vascular endothelial function. Myocardial biopsy revealed amyloid deposition exclusively in intramural coronary arteries. Early amyloidosis without myocardial involvement can produce acute coronary syndrome through the combination of spastic epicardial coronary arteries and obstruction of the intramural coronary arteries. In the management of certain patients with acute coronary syndrome, the possibility of cardiac amyloidosis should be taken into consideration. (Int Heart J 2010; 51: 134-136)

Key words: Cardiac amyloidosis, MRI, Endomyocardial biopsy, Coronary flow reserve, Vascular endothelial function

AL amyloidosis is a multisystem disorder. Cardiac manifestations usually depend on the pattern and amount of amyloid deposition in the heart.¹⁾ Clinical signs consist mainly of right-sided heart failure and restrictive diastolic ventricular failure. However, some patients develop angina due to amyloid deposits in the coronary arteries.²⁾ Early amyloidosis without myocardial involvement can produce acute coronary syndrome by the combination of spastic epicardial coronary arteries and intramural coronary artery obstruction. The present findings indicate that it is particularly important to diagnose amyloidosis in acute coronary syndrome, because the prognosis of patients with intracoronary amyloid is reported to be poor.³⁾

CASE REPORT

A 49-year-old Japanese man was admitted to our hospital complaining of nausea and back pain which had begun 2 days earlier in June 2008. He had a 15-year history of AL amyloidosis. An ECG showed ST segment elevation in II, III, aVF, and inverted T waves in V4-V6. The QRS voltage was low in the limb leads. Subsequently, the serum concentrations of troponin T and creatine kinase rose above the normal upper limits. Echocardiography revealed severe hypokinesis and scar formation of the inferior and posterior wall, and hypokinesis of the anteroseptal wall. However, the left ventricular wall was not thick and left ventricular diastolic performance parameters (E/A, and DcT) were within

normal limits. The baseline coronary angiography (CAG) demonstrated no organic stenosis in the epicardial coronary arteries. However, LVG showed severe hypokinesis of the apex, and inferior and posterior walls. Moreover, intracoronary injections of acetylcholine (ACh) induced chest pain and severe coronary vasospasm which was accompanied by ECG changes of ST elevation in leads V4-6, and II, III, and aVF, indicating vasospastic angina pectoris. The coronary flow reserve in response to papaverine was also abnormal. The percentage change in coronary flow reserve (CFR) was attenuated. Average peak velocity (APV) changed from 21 cm/s (baseline) to 46 cm/s after intracoronary (IC) papaverine administration. CFR in response to papaverine was 2.2 (Figure 1). Late gadolinium-enhanced cardiac MRI (CMRI) showed that late gadolinium enhancement was widespread (anteroseptal, lateral, and inferior walls of the left ventricle) and greatest in the subendocardium, particularly in the inferior wall. This cardiac MRI result suggested two possibilities; multivessel coronary artery spasm, and/or obstructive intramural coronary amyloidosis (Figure 2). Cardiac biopsy revealed amyloid deposition not in the myocardial interstitium or myocardium, but in the small intramural coronary arteries (Figure 3). One year later, ECG showed abnormal Q waves in leads II, III, and aVF, and UCG showed inferior hypokinesis, findings that were compatible with inferior old myocardial infarction.

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Figure 1. Coronary flow pattern. Baseline wave (A) After papaverine IC (B). APV (average peak velocity) changed from 21 cm/s to 46 cm/s. CFR in response to papaverine was 2.2.

Figure 2. Late gadolinium-enhanced cardiac MRI. Late gadolinium enhancement is seen widespread throughout the subendocardium, causing thinning, particularly in the inferior wall.

Figure 3. Histological findings of the heart. Paraffin sections of left ventricle attained with Congo-red (A), polarizing microscope findings (B). Only perivascular amyloid deposition was seen in the left ventricular wall, which was positive for Congo-red staining, and shown as green using polarizing microscope filters.

DISCUSSION

The clinical presentation of cardiac amyloidosis is usually dominated by diastolic ventricular failure and right-sided heart failure, because of amyloid deposition in myocardial interstitium.¹⁾ In our case, amyloid deposition was recognized only in the intramural coronary arteries. This presentation of amyloidosis is rare, and amyloid deposition in intramural coronary arteries has been described only in a few individual case reports.^{2,4)} In one of these case reports, obstructive intramural coronary artery amyloid deposition was reported to cause angina, not transmural infarction.²⁾ A case of ST elevation MI (STEMI) due to intramural coronary amyloidosis has not been previously reported.

The question is whether the myocardial ischemic events in this case were due to coronary vasospasm or to intramural coronary disease, or both. A differential diagnosis between coronary vasospasm and intramural coronary disease is difficult. Several cases of obstructive intramural coronary artery amyloid deposition which presented low coronary flow reserve and impaired vascular endothelial

cells have been previously described.³⁾ Almost all patients with obstructive intramural coronary artery amyloid deposition seem to have impaired coronary flow reserve^{3,4)} In our patient, we speculate that right coronary artery spasm based on the obstructive intramural coronary amyloidosis occurred.

In conclusion, cardiac amyloidosis can present as coronary artery spasm or STEMI and is associated with coronary flow reserve abnormalities despite normal baseline coronary angiograms. It is important to consider the possibility of amyloidosis in such cases of ischemic heart disease because the prognosis and therapeutic strategy of patients with intramural coronary amyloidosis are different from ordinary ischemic heart disease.^{4,5)} In our case, chemotherapeutic regimens (melphalan and prednisone) for treating AL amyloidosis were started, and they were proven to be effective. Among patients with AL amyloidosis, median survival has been reported to be 1.1 years in those with any sign of cardiac involvement.⁶⁾ However, after 1.4 years, this patient still shows no evidence of deterioration.

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