Isolated Congenital Atresia of the Left Main Coronary Artery and Atherosclerosis

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Successful surgical revascularization in an adult with isolated congenital atresia of the left main coronary artery with coexistent two-vessel atherosclerosis is described. This extremely rare anomaly must be differentiated from single right coronary artery and acquired occlusion of the left main coronary artery. Symptomatic myocardial ischemia in patients surviving to adulthood is an indication for surgical revascularization in them.


Isolated atresia of the left main coronary artery (LMCA) in an adult is an extremely rare lesion. In a review of 126,595 patients undergoing coronary arteriography over a 29-year period, Yamanaka and Hobbs [1] described 1,461 cases of anomalies of origin and distribution of coronary arteries but did not mention a single case of LMCA atresia. Susceptibility to atherosclerosis in the presence of some form of coronary artery anomaly has remained a point of debate. Coronary Artery Surgery Study data indicated clinically significant atherosclerotic stenosis in only 18 of 73 patients with a major coronary artery anomaly in a study population of 24,995 patients [2].

We report operation on an adult with LMCA atresia with coexistent two-vessel atherosclerosis.

A 56-year-old man presented with Canadian Cardiovascular Society class 3 angina 7 months after sustaining an inferior wall myocardial infarction in July 1991. Anginal symptoms had started 5 months before the myocardial infarction. Smoking, obesity, diabetes mellitus, and family history of ischemic heart disease were the risk factors in his clinical history profile. He was being treated with oral antihyperglycemics, nitrates, calcium-channel antagonists, and aspirin. Physical examination was unremarkable except for a 2/6 systolic murmur at the apex. The chest roentgenogram and rest electrocardiogram were within normal limits. Cardiac catheterization in November 1991 had shown normal left ventricular function, normal chamber pressures, and no evidence of left-to-right shunt. The left coronary ostium could not be selectively cannulated, and the left coronary system was not directly visualized. The right coronary artery (RCA) was large and dominant but did not show extensive collaterals to the left system. The pulmonary artery was not opacified on RCA injection. A large conus artery arose from a separate ostium in the right aortic sinus anterior and superior to the RCA, coursed anteriorly subepicardially across the right ventricular outflow tract, anastomosed with the left anterior descending (LAD) artery at its upper third, and filled the entire left system. Both LAD and the circumflex artery were opacified up to their origin on selective injection of the conus artery.

Clinically significant atherosclerotic irregularity of the proximal LAD, 80% stenosis at the origin of the circumflex artery, 90% narrowing at the origin of its first marginal branch, and 70% obstruction at the origin of the first diagonal branch of LAD arising proximal to the natural anastomosis of the conus artery and the LAD were seen (Fig 1). In February 1992, saphenous vein bypass grafting to the first diagonal and the first obtuse marginal arteries was done. At operation, LMCA atresia was confirmed by a fibrous band at the location of LMCA and a small blind dimple at the left aortic sinus. The patient had an uneventful recovery.

Comment

Surgical management of congenital atresia of LMCA has been reported in only 11 patients in eight reports in the literature. Only 4 of them were adults [3–5]. Associated coronary atherosclerosis or congenital cardiac defects were not seen in these adults. In our experience during the last 20 years, this is the fourth patient with congenital atresia of LMCA among more than 13,000 patients undergoing open heart operations including more than 4,500 having coronary artery bypass grafting. The reported case is our only patient with atretic LMCA and associated coronary atherosclerosis.

Atresia of the LMCA is differentiated from single RCA in terms of blood flow pattern. In single RCA the pattern is centrifugal as the blood flows from the aorta toward the periphery through progressively smaller vessels. In contrast, in LMCA atresia the flow is centripetal as the circulation is usually from RCA collaterals to the left system vessels of increasingly larger size, and the flow is reversed in the left system. In the presence of a large conus artery, as seen in our patient, this reversal of flow is partial.

The collateral channels seen in atresia of LMCA include the circle of Vieuussens, the anastomoses between anterior interventricular branches of the RCA and the LAD, and the final ramifications of the posterior descending branch.
of the RCA anastomosing with the terminal branches of the LAD. In the reported patient, the separate conus artery constituted the circle of Vieussens to perfuse the entire left system through its anastomosis with the LAD. The conus artery is considered a normal variant rather than an anomaly as it is reported in 50% of hearts as arising from a separate ostium in the right aortic sinus. Fortwin and Roberts [6] described in their autopsy report an almost similar lesion with extensive calcific atherosclerosis of the entire coronary tree. However, in their case the conus artery arose from the proximal RCA and joined the left coronary artery at the point of its bifurcation.

Clinical and angiographic differentiation of the atresia may not be possible from acquired occlusion of the LMCA, which may be caused by atherosclerosis, polyarteritis nodosa, Takayasu arteritis, or embolism. Only the surgical observation of the presence of the left coronary ostium and the size and external appearance of the LMCA rules out congenital atresia.

Survival to adulthood depends on the “adequacy” of collateral circulation and severity of coexistent congenital heart lesions, if any. Even in the absence of acquired atherosclerosis or vasospasm, myocardial ischemia becomes inevitably evident in all adult patients from the fifth decade onward. Possible early left ventricular hypertrophy and consequent increase in myocardial oxygen demand contribute to the development of ischemic features. Atherosclerosis, as seen in our patient and others, runs a similar course as in other populations with coronary artery disease. The presence of a “natural” arterial bypass graft to the LAD in the form of the conus artery prompted us to revascularize the diagonal and marginal arteries only.

References