Essential Thrombocythemia Associated with Angina Pectoris with Unusual Coronary Artery Findings*

Hiroshi Yoshida, M.D.; Tsuneo Hoshino, M.D.; Takahumi Ishida, M.D.; Tadahiko Shiomura, M.D.; and Tsuneo Kaburagi, M.D.

A case of ET associated with angina pectoris is presented. Angiography showed a 3.0-cm long mosaic-like thrombus shadow consisting of small filling defects in the proximal left anterior descending artery. The lesion could not be reduced with warfarin, ticlopidine, trapidil, urokinase or melphalan. Coronary artery bypass grafting was performed successfully.

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Essential thrombocythemia, a myeloproliferative disorder, may result in thrombus formation and acute vascular occlusion. However, coronary artery occlusion in patients with ET rarely has been described. The effect of chemotherapy suppressing bone marrow activity is controversial. We describe a patient with ET who had unusual coronary arteriographic features and who underwent CABG successfully while being treated with melphalan.

CASE REPORT

A 52-year-old man visited our hospital because of angina pectoris and thrombocythemia. He was well until seven months earlier, when chest pain on effort appeared. He was admitted to another hospital. Coronary arteriography showed a 3.0-cm long mosaic-like thrombus shadow consisting of small filling defects in the proximal left anterior descending artery associated with delayed filling of the distal coronary artery. The first diagonal branch was totally occluded and filled poorly by collaterals from the circumflex artery. The other vessels were entirely normal (Fig 1 and 2). The platelet count was 1,092,000/cu mm and the remainder of the hemogram was normal. Ticlopidine, 100 mg twice daily; trapidil, 100 mg three times daily; and diltiazem, 30 mg three times daily, were started. After three months of medication, coronary arteriography was repeated. Urokinase, in a dose of 240,000 units, was infused into the left coronary artery, but the lesion did not change. Thallium 201 exercise scintigraphy revealed reversible perfusion of the anteroseptal wall of the left ventricle. He was referred to our hospital. Bone marrow examination revealed increased megakaryocytes and a mildly hypoplastic marrow, confirming the diagnosis of ET. The aggregation of platelets with epinephrine, adenosine diphosphate and collagen was normal. Bleeding time, coagulation time, prothrombin time, partial thromboplastin time and thromboelastogram also were normal. Diltiazem, 30 mg three times daily, long-acting isosorbide dinitrate, 20 mg three times daily; metoprolol, 20 mg twice daily;

*From the Department of Cardiology (Drs. Yoshida, Hoshino, Ishida and Kaburagi) and the Department of Hematology (Dr. Shiomura), Shizuoka General Hospital, Shizuoka, Japan.

Reprint requests: Dr. Yoshida, Department of Cardiology, Shizuoka General Hospital, Shizuoka, Japan 420
FIGURE 3. The left anterior descending artery is occluded and the first diagonal branch opacified rapidly but contains many small filling defects similar to the previous lesion of the left anterior descending artery.

warfarin sulfate, 3 mg daily, and melphalan, 2 mg daily, were administered. Three months after the start of melphalan treatment, the platelet count was 400,000/cu mm and a third cardiac catheterization was performed. Coronary arteriography revealed total occlusion of the left anterior descending artery with good filling by collaterals from the right coronary artery, and the recanalized first diagonal branch contained small filling defects similar to the lesions previously seen in the left anterior descending artery (Fig 3). Left ventriculography showed normal contractions. Transient ischemia, coincident with anginal pain, was noted in the anteroseptal wall on thallium 201 stress scintigraphy, similar to that seen in the previous thallium 201 stress scintigraphy. We performed CABG using the left IMA. There were no thrombohemorrhagic complications. He has been free from anginal pain since surgery.

DISCUSSION

Essential thrombocythemia is a myeloproliferative disorder characterized by isolated overproduction of platelets and an increased risk of thrombohemorrhagic complications. The most common symptom of ET is excessive bleeding; thromboembolism is less frequent. One of the potentially fatal but rare thromboembolic complications is ischemic heart disease. Decreased platelet aggregation, particularly with adenosine diphosphate and epinephrine, is often observed. The aim of therapy for ET is suppression of the excessive bone marrow activity, which can be achieved by whole-body irradiation or chemotherapy with small doses of busulfan or melphalan. However, a direct relationship between platelet count or mass and risk of complications has not been conclusively demonstrated, and no randomized prospective data exist to prove that routine suppression of the platelet count prolongs survival or reduces the incidence of symptoms. So the indications for treatment of ET are controversial. However, ET associated with potentially fatal complications should be treated as soon as the diagnosis is made.

Reports of ET associated with ischemic heart disease have been made, and there are two types of coronary arteriographic findings. In one, there are normal coronary arteries without atherosclerosis and in the other, there is coronary artery stenosis in young men. In the former type, thrombus formation could cause myocardial infarction, so chemotherapy and treatment with an antiplatelet agent might be effective in preventing an ischemic event. In the latter type, significant stenosis often is located in the left anterior descending artery. Medical therapy alone cannot reduce the stenosis, and interventional treatment is needed. Pick et al described a patient with a stenotic lesion treated with CABC because of the length of his lesion and the high platelet count with its proclivity toward excessive bleeding and thrombosis. Hanger et al performed PTCA successfully in a patient being treated with busulfan, aspirin and diltiazem. There is no reason for ET to affect the left anterior descending artery most often. In our patient, the coronary artery lesion had more complex morphology and was obviously different from that of previously reported cases. In ours, a 3.0-cm long mosaic-like thrombus shadow made up of small filling defects was observed in the proximal left anterior descending artery. There had been no improvement with urokinase, ticlopidine, trapidil, warfarin sulfate and melphalan treatment, although the platelet count had decreased. The lesion was thought to be neither a so-called thrombus formed at the time of infarction of a related artery nor the atherosclerosis usually observed in ischemic coronary artery disease, but perhaps a collection of small thrombi consisting of a random aggregation of platelet clumps. In this patient, PTCA was not indicated because of the complex long lesion with thrombi. Since the platelet count and platelet aggregation were well controlled, CABC was performed safely.

REFERENCES