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Clinical Case Report

Coronary artery bypass graft in a patient with Fabry's disease

Hiroaki Osada [a,*], Naoki Kanemitsu [a], Masahisa Kyogoku [b]

[a] Department of Cardiovascular Surgery, Mitsubishi Kyoto Hospital, 1 Katsuragoshomachi, Nishikyo-ku, Kyoto, 615-8087, Japan
[b] Department of Pathology, Mitsubishi Kyoto Hospital, 1 Katsuragoshomachi, Nishikyo-ku, Kyoto, 615-8087, Japan

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ABSTRACT

Fabry's disease is a lysosomal storage disease characterized by intracellular accumulation of ceramide trihexoside resulting from alpha-galactosidase A deficiency. While the heart is often involved, coronary artery disease and its management in Fabry's disease patients are extremely rare clinical entities. We report a case of a 72-year-old man with left main disease in Fabry's disease with special consideration of the arterial wall pathology.

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Fabry's disease (FD) is a lysosomal storage disease characterized by intracellular accumulation of ceramide trihexoside resulting from alpha-galactosidase A deficiency. While the heart, especially cardiomyocytes, are often involved [1,2], coronary artery disease and its management in FD patients are extremely rare clinical entities. We report a case of a 72-year-old man with left main disease (LMD) in FD.

A 72-year-old man was referred to our institution for surgical coronary revascularization for LMD-related effort angina. Angiokeratoma and hypohidrosis were present from childhood, and FD was diagnosed 4 years earlier. At diagnosis, his alpha-galactosidase A level was 0.4 nmol/mg/h (normal range: 49.8–116.4 nmol/mg/h), and he received enzyme replacement therapy every 2 weeks, from the time of diagnosis. For the 7 years prior to presentation, he was also receiving chronic hemodialysis for end-stage renal failure and cardiac resynchronization therapy-pacemaker for heart failure and complete atrioventricular block. He also had chronic hematuria because of prostatic cancer. One year earlier, he had undergone percutaneous coronary intervention for 90% stenosis of the proximal left circumflex artery (LCX), with an everolimus-eluting stent (XIENCE Xpedition®, 3.5×15 mm, Abbott Vascular, Santa Clara, CA, USA).

Transthoracic echocardiography revealed left ventricular ejection fraction of 46% (using the modified Simpson's biplane method), symmetrical left ventricular hypertrophy (interventricular septum thickness 14 mm, posterior wall thickness 13 mm), and hypokinesis of the posterior wall and hypo-akinesis of the apical inferior wall. Coronary angiography demonstrated distal left main trunk stenosis of 90% and proximal LCX intrastent restenosis of 90% (Fig. 1A, B).

Based on the finding that the LCX had a relatively small perfusion area, fear of dislocating the left ventricular cardiac resynchronization therapy-pacemaker lead, and the patient's clinical condition (heart failure and renal failure on hemodialysis, and hemorrhagic prostatic cancer), we planned to perform off-pump coronary artery bypass grafting (CABG) for the left anterior descending artery using the left internal thoracic artery (LITA).

Surgery was performed through a median sternotomy. LITA was harvested (completely skeletonized) with a harmonic scalpel (Ethicon Endo Surgery, Inc., Johnson & Johnson Medical, Somerville, NJ, USA) and was prepared for anastomosis. At the same time, the distal LITA graft was evaluated intraoperatively via rapid diagnosis [frozen section sample with hematoxylin and eosin stain (HE)], which revealed no clear evidence of arterial wall thickening or substance deposition (Fig. 1C, D).

Following the intraoperative diagnosis, we performed CABG with LITA to the left anterior descending artery. Graft flow measurement was performed with a transit time flow meter (VeriQ System, MediStim, Oslo, Norway). The mean LITA graft flow was 13 mL/min, pulsatility index was 2.7, and diastolic filling was 76%.

We added percutaneous coronary intervention for the proximal LCX intrastent restenosis with zotarolimus-eluting stents (Resolute Integrity 3.5×15 mm; Medtronic, Inc., Minneapolis, MN, USA) 1 week following the initial surgery. He was discharged after rehabilitation on postoperative day 10.

Postoperative histological evaluation involved light microscopy of the resected LITA graft with paraffin-embedded elastica van Gieson, Masson's trichrome, periodic acid-Schiff stain, and alcian blue staining.

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* Corresponding author. Tel.: +81-75-381-2111; fax: +81-75-392-7952.
E-mail address: osahiro@kuhp.kyoto-u.ac.jp (H. Osada).

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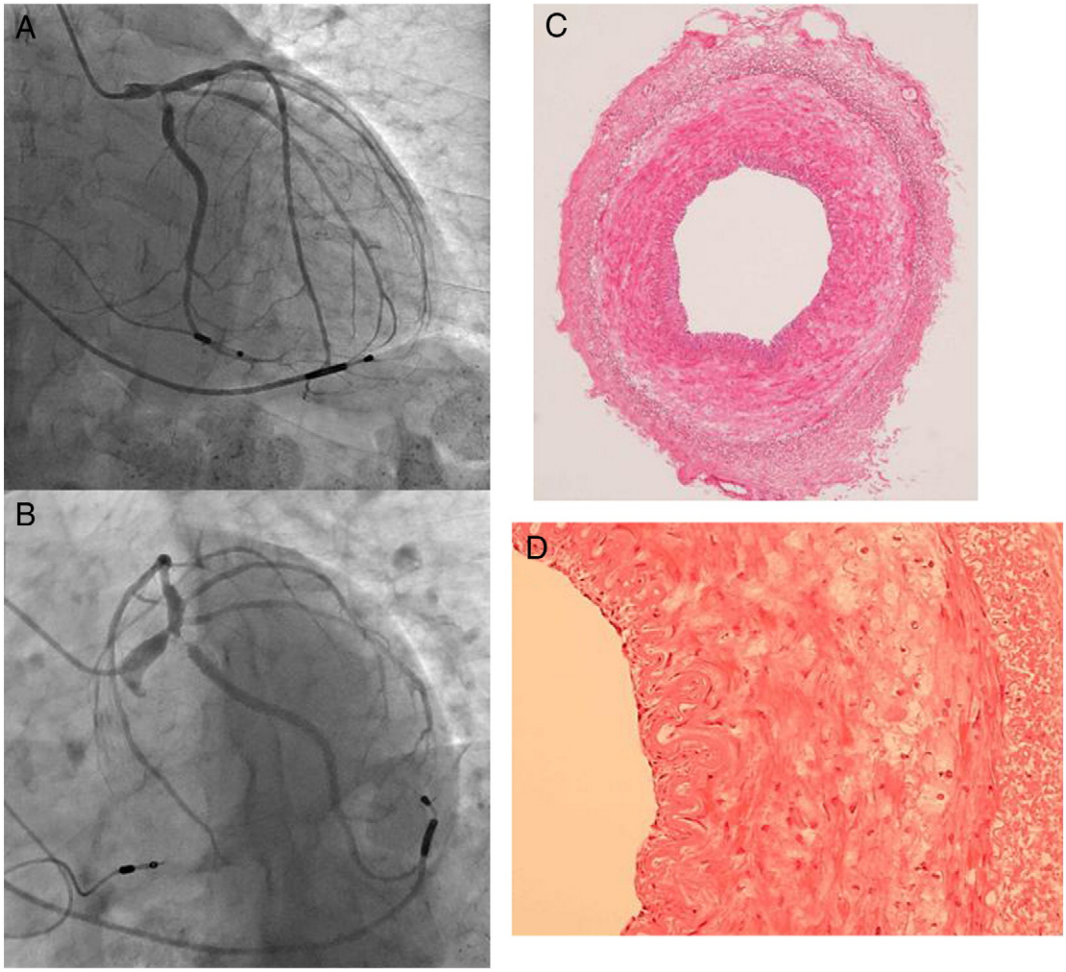

Fig. 1. (A, B) Preoperative coronary angiography showing 90% stenosis of the distal left main trunk and 90% restenosis of the proximal LCX intrastent. (C) Intraoperative frozen section of LITA showing minimal damage. In particular, the intima and internal elastic laminae appeared normal (HE scan). (D) Some vacuoles were present in the outer **portion of the media** (HE×400).

The outer half of the medial smooth muscle layer showed degeneration with large intracellular vacuoles and increased collagen fibers. The controlling peripheral nerve fibers (perineural cells) also contained high numbers of vacuoles, but endothelial cells and internal elastic lamina and the internal layer of smooth muscle cells appeared normal (Figs. 2, 3). Electron microscopy was not performed.

As of the latest follow-up, the patient was alive without increasing angina or heart failure, and the patency of the LITA graft was confirmed 6 months from the initial surgery.

The optimal management and prognosis are not yet clearly understood for coronary artery disease in FD patients. Continuous observational follow-up is necessary.

FD is an X-linked disorder characterized by intracellular ceramide accumulation resulting from alpha-galactosidase A deficiency. The heart is often involved (cardiac variant FD), which induces left ventricular hypertrophy; therefore, FD should be considered in unexplained cardiac hypertrophies [1,2]. Many FD patients complain of angina that results from microcirculation disturbances of the coronary artery [3]. However, the incidence of angina concomitant with abnormal angiographical findings and its optimal management among FD patients are not known. Chimenti et al. [4] reported a case of CABG with LITA graft in which the graft occluded 1 year from the initial surgery. The authors recommended a venous graft and intraoperative diagnosis of the graft. Fisher et al. [5] also reported CABG with LITA graft and showed a patent LITA graft at 19 months but also discussed the possibility of LITA degeneration.

For LMD, surgical coronary intervention is the first-choice therapy in our institution because of expected long-term outcomes [6], and because several reports have been published stating that grafts from the internal thoracic artery have a superior patency rate compared with venous grafts [7], we chose CABG using a LITA graft for the conduit in this case.

Although intraoperative rapid diagnosis revealed no remarkable abnormalities of the LITA graft, postoperative pathology with more analytical staining revealed degeneration in the arterial wall, especially in the outer part of the smooth muscle layers, and clear vacuolar changes in the perineural cells. Fortunately, the intima, internal elastic lamina, and the internal part of the smooth muscle layer were normal. We also confirmed a patent LITA graft 6 months postoperatively. Partial arterial wall degeneration does not necessarily induce early graft occlusion; however, rapid intraoperative diagnosis has several limitations. Careful follow-up is important.

We performed CABG for an elderly patient with FD. The optimal management, especially in graft selection regarding CABG, and prognosis are not yet clearly established for this rare condition. Continuous observational follow-up must be implemented in such patients.

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