induction, an increase in heart rate from 78 to 120 beats/min and an elevation of $P_{ET}CO_2$ from 35 to 60 mmHg were noted. Arterial blood gas measurements at this point showed a severe combined metabolic and respiratory acidemia (pH, 7.006; Pa_{O2}, 291.6 mmHg; Pa_{CO2}, 97.3 mmHg; BE, −11.7 mEq/l). Rectal temperature increased to 39.3°C 30 min after induction, and marked rigidity of the upper and lower limbs was noted. With a likely diagnosis of malignant hyperthermia, the patient was treated with active surface and core cooling and with intravenous administration of 100 mg of dantrolene. The immediate postoperative course was essentially uneventful.

Beginning on the day after the event, the patient complained of severe muscle weakness of the extremities. Muscle weakness did not return to normal even by 1 month after the episode, despite muscle rehabilitation. Hence, manual muscle tests (MMT) of the upper and lower limbs, commonly used for evaluation of muscle strength, were performed. Muscle strength of all muscles in the extremities was reduced to grade 3 or 4 (normal value, 5). Two months after the episode, the distal part of the limbs, such as the muscles of the forearms and those of the crus (those innervated by fibular and tibial nerves), remained unimproved, and a further month was necessary for all the muscles to improve completely. Muscle weakness remained in the areas where severe muscle rigidity had occurred at the time of the episode (table 1). A hand dynamometer test showed marked improvement of grasping power from 20 kg on the right and 19.5 kg on the left at 1 month to 44 kg and 42.5 kg, respectively, at 2 months.

Discussion

Pathohistologic studies have revealed various degrees of skeletal muscle destruction in muscle specimens obtained from patients recovering from MH.[2] The degree of muscle weakness resulting from such muscle destruction may vary, but from the present case, it is suggested that at least 2 or 3 months is required for complete recovery from muscle destruction.

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Fatal Acute Myocardial Infarction during General Anesthesia in a 7-yr-old Boy Associated with Total Intramural Coronary Arteries

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INTRAMURAL coronary artery, mural coronary artery, tunneled coronary artery, myocardial coronary bridging, and myocardial loop are all terms used to describe an anatomic variation in which an epicardial coronary artery, especially the left anterior descending (LAD) artery, becomes surrounded by myocardial fibers for some distance and depth but returns to the epicardial surface distally. We describe the case of a child in whom all epicardial coronary arteries had an intramyocardial course, probably causing fatal myocardial infarction during general anesthesia.

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Key words: acute myocardial infarction, intramural coronary artery, mural coronary artery, tunneled coronary artery, myocardial coronary bridging, sevoflurane.

Case Report

A 7-yr-old boy with shortening of the right Achilles tendon from right hemiparesis as a result of congenital cerebral palsy was scheduled for a lengthening of the Achilles tendon. He had been born at 35 weeks of gestation and had weighed 1780 g. A ventricular septal defect (VSD) was diagnosed immediately after birth. He was treated with digoxin, and at age 3 yr, right ventricular angiography and catheterization revealed little shunt flow through the VSD, a mean pulmonary pressure of 20 mmHg, a cardiac index of 6.45 $l \cdot min^{-1} \cdot m^{-2}$, and a left ventricular ejection fraction

(LVEF) of 69% that documented normal cardiac function. At age 5 yr, echocardiography showed the VSD to be 1 mm in diameter with no shunt flow and an LVEF of 64%. Any other dysmorphologic features were not noticed.

At the time of surgery, he weighed 18 kg, was 114 cm in height, and had no symptoms with daily exertion. However, he could not perform heavy exercise because of the right hemiparesis and the handicap of his leg. Preoperative laboratory examination, chest radiograph, and electrocardiogram (ECG) at rest were within normal limits. Auscultation documented Levine III/IV systolic murmur at apex and normal vesicular sounds at lung fields. Systolic blood pressures and pulse rates were 80–90 mmHg and 70–100 beats/min, respectively. He received atropine sulfate, 0.4 mg, mixed with diazepam, 8 mg orally, 1 h before entering the operating room. General anesthesia was induced with 5% sevoflurane in a mixture of 33% oxygen and 67% nitrous oxide; several minutes later he was tracheally intubated, and anesthesia was maintained with nitrous oxide, oxygen, and sevoflurane. Systolic blood pressures were maintained over 80 mmHg, whereas heart rates gradually increased and approached 120 beats/min. Twenty-five minutes after intubation and just after putting the tourniquet on the right thigh, while the heart rate was 120 beat/min, the systolic blood pressure suddenly decreased to 65 mmHg, and the oxygen saturation (Sp_{O2}) decreased to 97%. The sevoflurane was discontinued, the systolic blood pressure increased to 95 mmHg, surgery began, and sevoflurane was reintroduced. Within 10 min after starting the operation, the systolic blood pressures and heart rates were 90–100 mmHg and 100–110 beats/min, respectively, and the ECG appeared normal. Subsequently, the blood pressure and Sp_{O2} suddenly could not be measured, and bradycardia to 55 beats/min occurred at the same time, when on the surgical field only exposure around the Achilles tendon was advanced. The anesthesia and surgery were discontinued, and atropine sulfate was given immediately. Although heart rate was not changed, systolic blood pressure indicated around 90 mmHg. About 5 min after this episode, the ECG suddenly revealed ST segment elevation, and isosorbide dinitrate and diltiazem hydrochloride were immediately administered, however, ventricular tachycardia appeared soon. External cardiac massage was started, and lidocaine and epinephrine were administered. Cardiac massage maintained systolic blood pressure over 60 mmHg, but without cardiac massage, blood pressure was not measured. Transesophageal echocardiography revealed severe hypokinesis of posteroinferior wall, which suggested acute myocardial infarction. Systemic administration of tissue plasminogen activator (tisokinase, 3,600,000 U) and temporary transvenous cardiac pacing were subsequently performed, but there was no beneficial effect. Despite aggressive cardiopulmonary resuscitation, ventricular tachycardia was altered to ventricular fibrillation, and the heart gradually became totally akinetic. The patient died 5 h after starting resuscitation.

Postmortem examination was performed only on the heart. The heart weighed 116.5 g (heart weight of average boy aged 6–9 yr is 102.7 ± 6.25 g, mean ± SD), the epicardial coronary arteries were not seen, and only coronary veins were observed on the surface. Multiple petechial hemorrhage were found on the right and left ventricles (fig. 1A, 1B). The right and left coronary arteries arose normally from the right and left sinus of Valsalva of aorta and were totally surrounded by myocardial fibers. The right coronary artery (RCA) was narrow and short, and the distal lumen was unclear. The circumflex branch (CX) was also short, and the lumen was unclear. The anterior descending branch was relatively well developed and ran through an intramyocardial course at about 4 mm depth (fig. 1C). The left ventricular free wall and septum were about 10-mm and 7-mm thick, respectively. The membranous part of the interventricular septum showed very small VSD (about 1 mm). Microscopically, transmural coagulation necrosis with hemorrhage existed in part of the right ventricle, anterior wall, ventricular septum, and posteroinferior wall that supported widespread acute myocardial infarction pathologically. However, no other pathologic findings such as any vasculitis, aneurysm, thrombosis, embolus, myocarditis, myopathy, degeneration, or fibrosis were shown.

Discussion

Both main coronary arteries and the main branches commonly run through the fat tissue between myocardium and epicardium. Rarely, a segmental intramyocardial course of an epicardial coronary artery, known as *intramural coronary artery,* has been identified and associated with ischemic heart disease or unexpected sudden death,[1–3] and is clinically documented by systolic narrowing or a "milking effect" on coronary angiography[4–7] with compression of the artery by contraction of overbridging ventricular muscle. All previous reports[1–14] concerning intramural coronary arteries describe the LAD and rarely the RCA, which becomes surrounded by myocardial fibers for some distance and depth but returns to the epicardial surface distally. In the case of severe systolic narrowing in which the patient often complains of chest discomfort accompanying exertion or emotional stress, supraarterial myotomy or coronary bypass is performed and results in a good prognosis.[8–10] Morales *et al.*[11] reported that the intramural LAD is associated with myocardial fibrosis, presumed to be ischemic in origin. Their results were derived from an examination of 39 cases of intramural LAD that were separated into two groups, in which the presence or absence of fibrosis was assessed in sections from the anterior septum and adjoining left and right ventricles, including myocardium adjacent to the intramural LAD. In those with hearts with fibrosis, the intramural LAD was deeper than in those that lacked fibrosis. Angelini *et al.*[4] commented that a typical intramural coronary artery that is considered clinically significant is 2–4 mm in depth and 10–30 mm in length. However, Virmani *et al.*[12] questioned the occurrence of myocardial fibrosis as a cause of cardiac ischemia and emphasized that the cardiac ischemia may be responsible for a degree of systolic narrowing on the angiography. In one study,[7] ST depression on ECG and lactate production were demonstrated during cardiac pacing in patients with severe systolic narrowing of intramural LAD. These patients also had impaired myocardial perfusion and left ventricular function.[13] However, the mechanism of ischemia associated with an intramural coronary

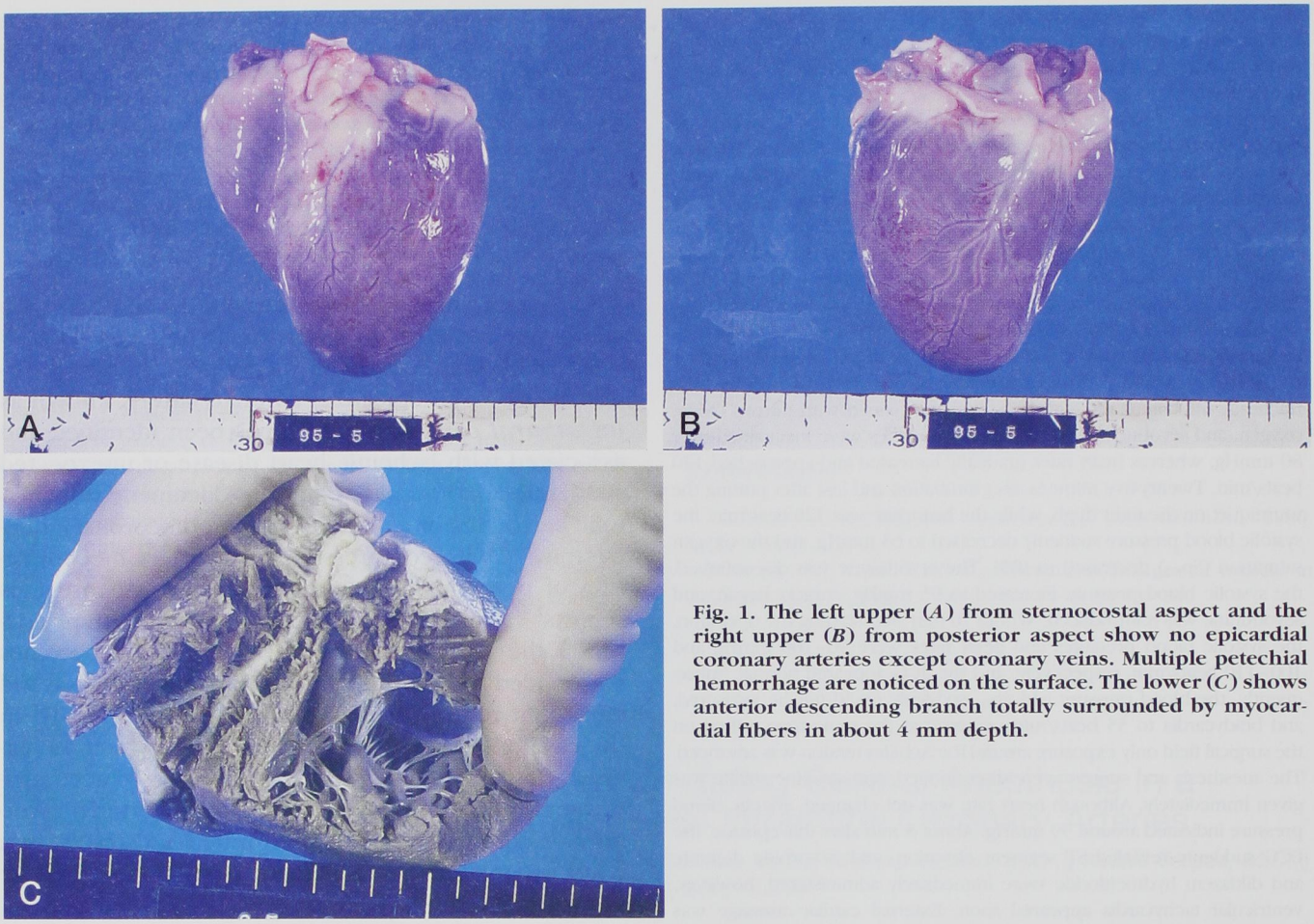

Fig. 1. The left upper (*A*) from sternocostal aspect and the right upper (*B*) from posterior aspect show no epicardial coronary arteries except coronary veins. Multiple petechial hemorrhage are noticed on the surface. The lower (*C*) shows anterior descending branch totally surrounded by myocardial fibers in about 4 mm depth.

artery is difficult to explain because compression is seen in systole and coronary blood flow occurs predominantly during diastole. When the heart rate is increased in patients with an intramural coronary artery, the diastolic filling time may be shortened, and the potential for myocardial ischemia may increase. Hill *et al.*[14] directly measured the coronary flow through an intramural LAD during cardiac operation and documented a severe depression of coronary flow as heart rate increased. Nitroglycerin as a coronary vasodilator has been reported to aggravate a degree of systolic narrowing on coronary angiography.[6] This finding seems to conflict with ordinary management for cardiac ischemia. On the other hand, fetal coronary arteries have an intramyocardial course. Rossi *et al.*[7] then suggests that the intramural coronary artery is a congenital malformation caused by the disorder of exteriorization. However, it is unknown when the coronary arteries are exteriorized and why clinical symptoms from the intramural coronary arteries only occur in adults.

Our patient revealed all the coronary arteries to be surrounded by myocardial fibers for the full length, accompanied by poor development of RCA and CX. The regions of myocardial infarction were right ventricle, posteroinferior wall, anterior wall, and ventricular septum that documented transmural coagulation necrosis pathologically. According to the clinical episode, severe hypokinesis of posteroinferior wall first documented by echocardiography is consistent with poorly developed and intramural RCA and CX. Sevoflurane is also a vasodilator. Therefore, the use of sevoflurane, the increase of heart rate, and the incidental hypotension might have trigger of the fatal decrease of coronary blood flow.

Interestingly, the present patient did not complain of any symptoms with daily life. Left ventricular angiography is potentially able to demonstrate this anomaly, although the patient only underwent right ventricular angiography for the examination of VSD. If intramyocardial coronary arteries are known to be present, care could be altered. Hypotension and tachycardia should

be avoided. Vasodilators such as nitroglycerin (and perhaps volatile anesthetics) may be contraindicated. Although a segmental intramural coronary artery can be managed with supraarterial myotomy or coronary bypass,[8-10] management of total intramural coronary arteries as in the current case has not been described.

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Fatal Pulmonary Embolism during Liver Transplantation

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FATAL pulmonary embolism is a very rare intraoperative complication of orthotopic liver transplantation (OLT), despite the use of antifibrinolytic agents in cirrhotic patients prone to hyperfibrinolysis in the setting of clotting activation and thrombin generation.[1] We report two such fatal complications to which aprotinin may have contributed.

Case Reports

Case 1

The patient was a 38-yr-old man with Laennec's cirrhosis and hepatitis C. Eight months before transplantation he presented with esophageal variceal bleeding and spontaneous bacterial peritonitis. He became progressively encephalopathic and short of breath and was admitted to the hospital. Laboratory studies included hematocrit,

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