tion. The other complications are recurrent diarrhea and opportunistic infections, which include mucocutaneous candidiasis, herpes zoster, pneumocystis carinii pneumonia, and cytomegalovirus disease [3]. The prognosis of the patient with hypogammaglobulinemia, thymoma, and leukopenia is very poor due to such infectious diseases; therefore, immediate diagnosis is important. In our case, although no signs of immunodeficiency were noticed when the thymoma was detected, 6 months after the thymothymectomy, recurrent pneumonia and diarrhea developed, based on which a diagnosis of Good syndrome was made. It should be noted that immunodeficiency can develop after the resection of thymoma in the absence of recurrence. The development of infectious diseases in a patient with thymoma or after the resection of thymoma mandates early and comprehensive immunologic investigation. It should include the evaluation of the peripheral blood count of B cells, $CD4^+$, and $CD8^+$ T cells by flow cytometry, and the quantitative analysis of serum immunoglobulin subclasses to diagnose and treat the disorder at an early stage [3].

The pathogenesis of this syndrome and the association of thymoma and immunodeficiency remain unclear. Most studies report that hypogammaglobulinemia in Good syndrome did not improve after thymectomy. In fact it was observed to be aggravated in some cases. Thymectomy should be performed in most patients with thymomas to prevent locally invasive growth and metastasis. However, thymectomy should not be expected to lead to the normalization of immune function, given the absence of any reports of resolution of the immunodeficiency in Good syndrome that convincingly demonstrated such a resolution to be related to thymectomy [3]. Therefore, IVIG should be administered to the cases of hypogammaglobulinemia in doses appropriate for the treatment of humoral immunodeficiency. The optimal immunoglobulin G level has been reported to be 200 to 500 mg/dL [4]. However, the treatment of patients with leucopenia or T-cell dysfunction, or both, by IVIG alone is insufficient. Degos and colleagues [5] reported a case of Good syndrome with agranulocytosis in which infectious diseases were controlled by plasmapheresis. In our case, the patient was not only administered IVIG, but also figrastim to maintain a leukocyte count between 2,000 and 6,000/mm^3. He has remained symptom free for more than 1 year since the beginning of this therapy.

In conclusion, it should be taken into consideration that immunodeficiency can develop after the resection of thymoma. The development of infectious diseases in a patient with thymoma or after the resection of thymoma mandates an early and comprehensive immunologic investigation. Regular gamma globulin and figrastim injections may be successful in maintaining a symptom-free status in a patient with Good syndrome who also has leukopenia.

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Delayed Repair of Acute Type A Aortic Dissection in a Patient with Gastrointestinal Bleeding and Pulse Deficit

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FEATURE ARTICLES

Acute type A aortic dissections are considered surgical emergencies because these patients are at risk for life-threatening complications. Patients who present with significant neurologic and other end-organ malperfusion may benefit from a more conservative approach. We present a patient with type A aortic dissection and concomitant mesenteric and limb ischemia.

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Acute type A aortic dissection (AAD) is a catastrophic event that carries a significant perioperative mortality. Reportedly, contemporary operative outcomes after the repair of AAD have a perioperative mortality of approximately 15% [1]. The majority of patients present with chest pain that radiates to the back; however, a variety of other presentations have been documented. Preoperative presentation and signs have a significant impact on operative and postoperative outcomes [2]. The most common treatment for AAD is urgent repair of the ascending aorta with reconstitution of blood flow into the true lumen. In selective cases of AAD, conservative management may be prudent when confronted with significant neurologic or other end-organ compromise.

Twenty percent of patients with AAD will present with a pulse deficit or mesenteric ischemia. Mortality of mesenteric ischemia in the setting of AAD has been reported to be approximately 30% [3]. Likewise, preoperative pulse deficit and limb ischemia have both been shown to

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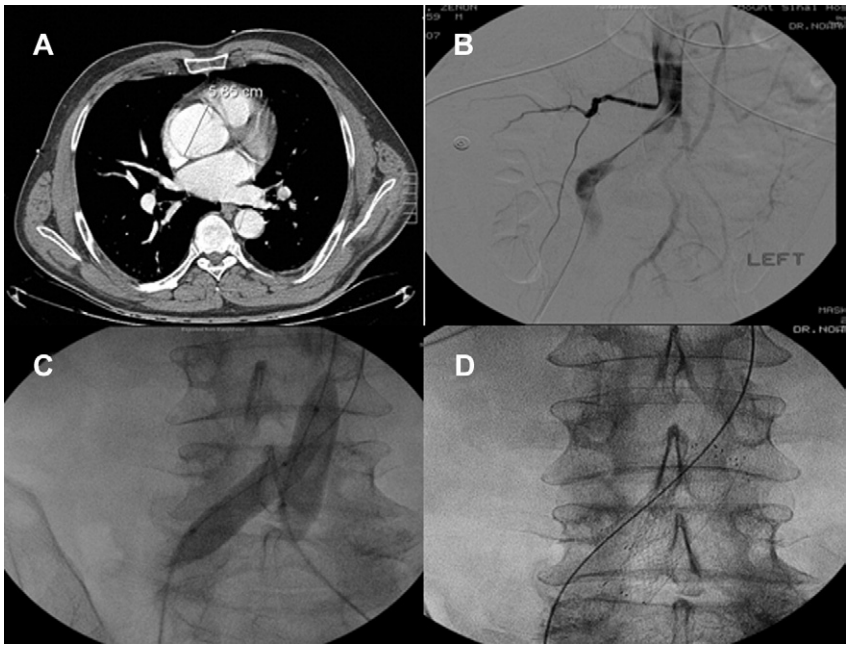

Fig 1. (A) Chest computed tomographic angiogram showing the type A aortic dissection and aortic root aneurysm. (B) Initial aortogram through the true lumen. (C) Bilateral balloon fenestration. (D) Anchored stents fenestrating the distal aorta.

be independent predictors of operative mortality. There may be evidence to suggest better surgical outcomes in this subset of patients if conservatively treated initially or treated with percutaneous interventions [3]. We present a patient with AAD who presented with gastrointestinal bleeding and limb ischemia.

A 42-year-old man was admitted to an emergency room with severe chest pain radiating to his back, along with abdominal pain and right lower extremity pain. A computed tomographic scan of the chest revealed a 5.8-cm aortic root aneurysm and an intimal flap in the ascending aorta, and propagation along the entire aorta (Fig 1A). The false lumen occupied 90% of the aortic diameter. The mesenteric vessels communicated with the true lumen. The dissection involved the innominate artery, both carotids, and the subclavian arteries. Prior to transfer he had two episodes of hematemesis.

At our hospital the hematemesis was persistent and was associated with hematochezia. The patient's abdominal pain was out of proportion to the findings of his physical examination. His laboratory studies showed an elevated lactate level. His right leg was cold, pale and pulseless below the femoral region. In the setting of an active gastrointestinal bleed, we reconsidered surgery. A transthoracic echocardiogram showed no pericardial effusion, a preserved ventricular function, and moderate aortic valve insufficiency.

Vascular surgery was consulted for an endovascular fenestration of the aorta and revascularization of the right lower extremity. The patient was taken to the interventional radiology suite where both femoral arteries were cannulated to access the true and false lumens. The pressure in the false lumen was equal to the radial artery pressure; however, the true lumen pressure tracing was not pulsatile and measured a mean arterial pressure of 25 mm Hg. Retrograde aortogram through the true lumen pigtail catheter revealed sluggish flow into the mesenteric vessels and bilateral renal arteries (Fig 1B). Aortic fenestration from the false lumen to the true lumen was performed in the distal abdominal aorta using two 14 mm $\times$ 4 cm balloon catheters (Fig 1C). The follow-up aortogram through the false lumen catheter did not visualize the mesenteric vessels. The dissection flap was fixed by placing two 14 mm $\times$ 6 cm smart stents that crisscrossed through the membrane and anchored onto the contralateral common iliac artery (Fig 1D). The completion aortogram revealed flow through the fenestration, perfusion of the visceral arteries and right femoral artery with equalization of arterial pressures in both lumens.

The patient's abdominal and right lower extremity symptoms had immediately improved. An upper endoscopy showed superficial ulceration throughout the stomach and duodenum. On his fourth day at our hospital, he became delirious and combative with fevers. A colonoscopy showed patchy areas of necrosis of the ascending colon requiring a right hemicolectomy. Operative findings were consistent with palpable pulses in the mesenteric arteries. When he awakened from the sedation, he was found to have a right hemiplegia. A computed tomographic scan of the head showed an acute infarct in the left occipital lobe and internal capsule. Carotid artery duplex confirmed a partial thrombosis of the left common and internal carotid arteries. The patient was transferred to a rehabilitation center where he made nearly a complete recovery of his neurologic symptoms. His aortic dissection and aortic valve regurgitation were followed periodically with computed tomographic angiograms and echocardiograms.

Eight weeks after his initial presentation, he underwent an aortic root replacement and fenestration of his

aortic arch. During his operation we placed arterial lines into both femoral arteries to assess flow through the fenestration once on cardiopulmonary bypass and after fenestrating the aortic arch. The intimal tear was located above the noncoronary cusp. The aortic tissues were thick, allowing reconstruction to be accomplished without difficulty. His postoperative course was uneventful and he was discharged home.

Comment

Acute type A aortic dissections are considered surgical emergencies because these patients are at risk for life-threatening complications. Deeb and colleagues [3] have surmised that the increased mortality is secondary to a perfusion-reperfusion injury and subsequent increase in tissue permeability leading to organ dysfunction. Although their study indicates an 89% mortality rate for patients with preoperative organ malperfusion other surgical units have a much lower mortality for this subset of patients [1]. Regardless, active gastrointestinal bleeding, and mesenteric and limb ischemia are major reasons to delay immediate repair. Other clinical scenarios to consider delaying aortic repair are hemorrhagic stroke, an unresponsive patient, concomitant and complicated myocardial infarction, and cardiovascular collapse. The absence of pericardial fluid suggested delay might be tolerated. Heparinization, prolonged cardiopulmonary bypass time, and hypothermic circulatory arrest may have exacerbated this patient's clinical predicament. The decision against immediate repair must be counterbalanced with the fact that as much as 15% of these patients will die prior to definitive repair. Definitive surgical repair should be considered earlier if there is an increase in the pericardial effusion and aortic dimensions on subsequent imaging. Once the acute complications have been addressed, the definitive repair can be delayed for at least 4 to 6 weeks when the aortic tissues should have matured and the patient has been given time to convalesce and prepare for the aortic replacement.

Interventional radiology advancements have made a significant impact on the reperfusion of ischemic organs. We elected to pursue an unconventional but recognized treatment for this condition with the intended plan to bridge this patient to a subsequent aortic repair. Despite a favorable outcome in this case, the patient suffered two major complications during the preoperative period. Other options for this patient were conventional ascending aortic repair, open abdominal aortic fenestration, endovascular septum stripping, or stenting of the individual visceral vessels. Immediate surgical repair would have not prevented either complication.

Percutaneous aortic fenestration allowed the patient to clear the ischemia induced-inflammatory mediators, lactate acidosis, and myoglobinuria, and to recuperate from the gastrointestinal bleed. Despite the need for a segmental colonic resection, the aortic fenestration proved successful due to the intraoperative findings of a strong, palpable mesenteric artery pulse and well-perfused surgical margins and viscera overall.

Delayed surgical repair of AAD is a viable option in a subset of patients. The favorable outcome in this case is a product of the patient's youth, overall good health, and accessibility of an experienced vascular interventional radiology team. This form of therapy may prove more favorable on survival in the elderly and moribund patient who could not tolerate a major surgical intervention. However, access to such expertise in catheter-based interventions may not be available at all institutions. At the present time, institutions with interventional radiology or cardiology and vascular surgery should be able to provide these catheter-based interventions. In the absence of these services, patients may benefit from referral to a tertiary-care facility where a skilled, multidisciplinary team may be available.

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FEATURE ARTICLES

Ileofemoral Malperfusion Complicating Type A Dissection: Revascularization Prevents Renal Failure

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We report four cases of lower extremity malperfusion complicating acute type A dissection. Two patients were treated with acute type A dissection repair, followed by axillobifemoral bypass grafting when malperfusion persisted after aortic replacement and required dialysis. Two patients were managed with lower extremity revascularization procedures before acute type A dissection repair and had preserved renal function. Lower extremity revascularization before cardiopulmonary bypass minimizes ischemia and allows for controlled limb reperfusion under hypothermic conditions compared with delayed normothermic reperfusion when performed after acute type A dissection repair. This strategy may increase limb

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