ILEAD—Ischemia of the Lower Extremities Due to Aortic Dissection: The Isolated Presentation

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Summary

Background and hypothesis: Lower extremity symptoms accompanying dissecting hematomas of the aorta are well described. Isolated lower extremity ischemia as the presenting syndrome of aortic dissection is rare and frequently misinterpreted so that the diagnosis of aortic dissection is delayed or missed, often with catastrophic results. This study was undertaken to determine its common characteristics and to reinforce recognition of this life-threatening phenomenon.

Methods: After the first patient in our series presented with isolated lower extremity ischemia due to aortic dissection, a prospective search for similar presentations was undertaken over 2 years. We also conducted a retrospective search for all aortic dissections at our hospital for the past 10 years and a MEDLINE search for all reported aortic dissections in the literature of the past 20 years, as well as for all reports of isolated ischemia of the lower extremities due to aortic dissection.

Results: Three patients with isolated ischemia of the lower extremities due to aortic dissection were found prospectively after the index case. Over 10 years, 40 patients with acute aortic dissection were diagnosed at our hospital. The MEDLINE search revealed 1,751 aortic dissections of which 10% had lower extremity symptoms. Only 10 cases of aortic dissection were reported as lower extremity ischemia with symptoms isolated to one or both lower extremities. Smoking, hypertension, male gender, hypercholesterolemia, and recent history of coronary artery bypass grafting were common predisposing risk factors for this rare presentation.

Conclusion: Symptomatic isolated ischemia of the lower extremities due to aortic dissection is rare and often missed. Awareness of its characteristics, aided by the acronym ILEAD, made it possible to suspect the true origin of this misleading syndrome.

Key words: aortic dissection, lower extremity ischemia, ILEAD, coronary artery bypass grafting

Introduction

Because a dissecting hematoma of the aorta is a first-order emergency, its protean symptoms must be appreciated. Indeed, aortic dissection is frequently missed or misdiagnosed because it can be asymptomatic or imitate other illnesses such as myocardial infarction, stroke, arterial embolus, visceral ischemia, musculoskeletal strain, and acute pericarditis. Because the first patient alerted us, we encountered, within a relatively short time, a consecutive series of four patients with apparent isolated ischemia of the lower extremities, that is, without chest or back pain, due to aortic dissection.

A detailed literature search revealed that lower extremity ischemia as a component of aortic dissection has been described in up to 26% of patients with aortic dissection¹ (Table I), but *isolated* ischemia of the lower extremities due to aortic dissection is rare and is described in only a few case reports, some within small series.^{2,3} Because the isolated presentation lacks the classic symptoms of chest pain, radiating back pain, or other associated symptoms, its presentation is dangerously misleading. We determined its common characteristics from our series and from reported experience. To reinforce cognition of this life-threatening phenomenon, we developed the acronym, ILEAD: "Isolated Ischemia of the Lower Extremities due to Aortic Dissection."

Methods

After the first patient presented with what appeared to be an arterial embolus to his lower extremity, a prospective search was undertaken by alerting our consultation service. Over 2

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TABLE I Aortic dissection presenting as lower extremity ischemia: All cases

Series	Number of aortic dissections	Ischemia of the lower extremities	% ^a
Pacifico	40	4	10
Young	65	17	26
DeBakey	527	37	7
Cambria	325	38	12
Hirst	505	50	10
Schumaker	33	5	15
Spittel	236	8	3
Amer	4	3	75
Shah	3	2	67
Schoon	3	0	0
Magne	7	1	14
White	3	2	67
Total	1751	167	10

^a % = Patients with lower extremity ischemia/patients with aortic dissection for each series.

years, three more patients with aortic dissection presenting with isolated lower extremity ischemia were diagnosed. A 20-year MEDLINE search was conducted for other reports of aortic dissection presenting as lower extremity ischemia. We reviewed another 36 patients with aortic dissection found by a computer search of our medical records. Each of our patients had a history, physical exam, chest x-ray, routine laboratory studies, and a transthoracic echocardiogram which was combined with a transesophageal echocardiogram or a computed tomography (CT) scan.

Results

Of the 40 patients with aortic dissection at our hospital, only 4 had lower extremity ischemic symptoms as their only presenting complaint. The other 36 had chest pain, back pain, abdominal pain, or syncope as their main symptom. In three of the four patients, transthoracic echocardiography (TTE) heightened the suspicion of DeBakey Type I⁴ aortic dissection due to an enlarged aortic root, aortic regurgitation, pericardial effusion, or evidence of an intimal flap. Transesophageal echocardiography (TEE) made the diagnosis in two patients, confirming an intimal flap of the ascending aorta, the dissectional entry site, and true and false lumina. Computed tomography scan with contrast was used in the other two patients when TEE was not available. All four patients were white males with hypertension; one had stopped taking antihypertensive medications on his own 1 month earlier. All had a history of hypercholesterolemia. None had Marfan's syndrome or other connective tissue disorders and none had diabetes. Two had a history of vascular disease and had had repair of an aneurysm of their abdominal aorta. Three patients consumed some alcohol. By definition, none had chest or back pain, but

all had lower extremity symptoms including numbness, weakness, shooting pains, and sensation of coldness. One patient had some periumbilical discomfort.

Three patients had initial diagnoses other than aortic dissection. The one patient who was discharged with a diagnosis of musculoskeletal strain of his lower extremity was admitted 2 days later for syncope; one was taken to the operating room for an arterial embolectomy; one was admitted to the neurology service with a diagnosis of stroke. The fourth patient presented with a sensation of coldness and discomfort in his lower extremities 2 weeks after coronary artery bypass grafting (CABG); he also had some periumbilical discomfort and was originally thought to have a rupture of a previously repaired abdominal aortic aneurysm. One patient had cardiac tamponade which had to be emergently drained by pericardiocentesis. In two patients, the chest x-ray showed a wide mediastinum. One had pulmonary vascular congestion; the other two had enlarged cardiopericardial silhouettes. The electrocardiogram (ECG) in two patients showed questionable ischemic changes; both patients had had CABG 2 weeks prior to presentation. Two patients had asymmetric right and left brachial blood pressures, the right arm about 20 mmHg higher. All patients had equal upper extremity pulses but unequal or absent pulses of the lower extremities. The one patient with absent left lower extremity pulse had a cold and cyanotic leg. Three of the four patients had aortic regurgitation by physical examination and Doppler echocardiography. Laboratory results were nonspecific, with minimal to moderate white blood count elevation in all four patients; none had evidence of infection. Serum creatinine level was upper normal or mildly elevated. All four patients had DeBakey I aortic dissection. Three of the four patients with ILEAD were misdiagnosed on initial presentation, and as a result the treatment of acute DeBakey Type I aortic dissection was delayed. Three were treated by repairing the ascending aorta; the fourth had endarterectomy of the abdominal aorta and repair of the flap of the left iliac artery which restored the flow to the lower extremities. One patient with hypertension at presentation was treated with antihypertensives and a beta blocker; the other patients were either hypotensive and required pressure support or had normotension on presentation.

Morbidity and mortality in our series of ILEAD was high. Three of the four patients died within 2 weeks of admission because of anoxic encephalopathy, gastrointestinal bleeding, and cardiac arrest with pneumonia. The fourth patient survived for 1 year; he died when his aortic valve was replaced for severely progressive aortic insufficiency.

Discussion

These patients with dissecting aortic hematoma presented with ILEAD. Dissection was difficult to diagnose because of this misleading presentation. Only one patient was quickly diagnosed because we were aware of the prospective search. Another patient, admitted for stroke, was diagnosed by TTE that had been ordered by the neurologist to evaluate the heart

for endocardial thrombi; no thrombus was noted, but an enlarged aortic root with a pericardial effusion raised a suspicion of aortic dissection; the diagnosis was made by CT scan showing DeBakey Type I aortic dissection.

Three of our patients had initial symptoms in one lower extremity and the fourth patient in both lower extremities. Hirst *et al.*⁵ reported 10% of patients presenting with aortic dissection to have clinical evidence of lower extremity ischemia, but not whether either extremity was involved more commonly or how many of these patients had isolated lower extremity ischemia. Our investigation of the 40 patients may indicate a male predominance in ILEAD; this is supported by other authors, such as Dutkiewicz and Przybyszewski,⁶ who found male predominance in patients with any ischemia of the lower extremity due to aortic dissection, and earlier authors who conducted large studies of aortic dissection.^{4,5}

Amer *et al.*,² Shah and Clauss,³ and Babu *et al.*⁷ reported black males to have an increased risk for aortic dissection presenting as lower extremity ischemia; our series included only white males. These differences may be explained because this presentation is rare and because the various series may be influenced by local patient populations.

All four patients with ILEAD in our series had a history of hypertension, one of the most important risk factors for aortic dissection.⁵ None had Marfan's syndrome or other connective tissue disorder predisposing to aortic dissection. Other common factors in our patients included smoking, hypercholesterolemia, and a history of recent CABG. Coronary artery bypass grafting may be a risk factor for aortic dissection due to disruption of the aortic intima by arterial cannulation, proximal anastomosis of the venous grafts, and crossclamping of the aorta.^{8,9}

While all patients in our series had DeBakey I aortic dissection, DeBakey Type II has been reported¹⁰ with lower extremity symptoms and signs. The reason that both types can be associated with ILEAD is probably due to the decrease in flow to the lower extremities by occlusion of the true lumen by intimal flaps in the iliac or femoral arteries, and possibly also due to ischemia of the lower extremity muscles caused by insufficient blood flow. ILEAD has mimicked acute arterial emboli, surprising surgeons who find an intimal flap after trying to perform an embolectomy,¹¹ as was the case also in one of our patients.

Other possible explanations for the patients' numbness and weakness of the lower extremity may be malperfusion of the spinal cord due to occlusion of the true lumen, thrombosis of the false lumen causing obstruction to blood flow in the true lumen, or possibly detachment of the branches of the aorta that perfuse the spinal cord. This malperfusion of the spinal cord may result in paraplegia if restoration of circulation is delayed.^{12,13}

The incidence of ILEAD, without other associated presenting symptoms, is difficult to determine from prior reports. Young *et al.*¹ reported that 26% of patients with aortic dissection present with lower extremity symptoms, but that the majority of these had associated chest or back pain as well. In our series, no patient had chest or back pain which could have

alerted the physician to a possible aortic dissection. The single patient who developed periumbilical discomfort had only lower extremity discomfort on arrival at the emergency room.

Because our series and all other series of lower extremity symptoms due to aortic dissection are relatively small, and because very few have truly isolated ischemic syndromes, ILEAD, the data were examined collectively. In our study, isolated lower extremity symptoms occurred in 10% (4/40) of consecutive patients presenting with aortic dissection. This equals the prevalence in the large series of Hirst *et al.*⁵ and approximates the proportion of patients, 10%, with lower extremity ischemia in the collective data summarized in Table I.

We used only reports with series of three or more patients to estimate the proportions of their isolated presentations as lower extremity ischemia. We omitted statistics from studies such as that by Babu *et al.*,⁷ reporting lower extremity ischemia due to aortic dissection without specifying those with isolated presentation, and excluded our patient with periumbilical discomfort from the series of ILEAD to maintain strict criteria of isolated presentation. Consequently, approximately one-third of the small subgroup of patients with aortic dissection presenting with lower extremity ischemia had ILEAD. Table II establishes the rarity of ILEAD. The overall proportion is far from a precise estimate since individual series varied from 2 of 17 (Young) to 3 of 4 (Amer, Pacifico) patients, and none of Schoon's patients. The mainly small denominators preclude a sophisticated statistical approach.

Conclusions

Isolated ischemia of the lower extremities due to ILEAD is rare. It is imperative that patients with ILEAD are diagnosed promptly to permit appropriate medical therapy and surgical repair of the dissecting aorta to restore the flow to the extremity. ILEAD is nearly always misdiagnosed on presentation because typical symptoms and signs of aortic dissection are absent, with frequently devastating consequences due to high morbidity and mortality even after delayed recognition. When a patient presents with nontraumatic acute ischemia of a lower extremity, aortic dissection should be considered in the dif-

TABLE II Aortic dissection: Subgroup with lower extremity ischemia and isolated presentation (ILEAD)

Aortic dissections (series)	Lower extremity ischemia	ILEAD (truly isolated groups)
Pacifico	4	3
Young	17	2
Amer	4	3
Shah	3	2
Schoon	3	0
Magne	7	1
White	3	2
Total	41	13

ferential diagnosis, even in patients without chest or back pain. A history of hypertension, and especially recent CABG, were prominent correlates in our series. It would be detrimental to misdiagnose such patients as having an arterial embolus and prescribe heparin or delay the diagnosis of aortic dissection by attempting embolectomy.

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