



Diagnosis and management of external iliac endofibrosis: A case report

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External iliac artery endofibrosis is an uncommon, nonatherosclerotic disease seen in endurance cyclists. It is poorly identified by providers. These otherwise healthy patients usually present with symptoms of arterial insufficiency, such as thigh or buttock pain, loss of power, or weakness occurring during strenuous exercises. These symptoms subside rapidly with rest. As these patients lack traditional risk factors of peripheral artery disease, their symptoms are often overlooked or are attributed to other etiologies, resulting in mismanagement and delayed treatment. In this case study, we report our experience with the successful management of a 48-year-old male who is a longstanding, avid cyclist. He self-referred to our institution after extensive research of providers familiar with his problem and at the recommendation of other cyclists with similar experiences. The patient underwent a successful left external iliac to common femoral artery endarterectomy and patch angioplasty. Three months after operation, he returned to cycling and, for the most part, has remained without symptoms. (J Vasc Nurs 2018;37:86-90)

The most common presenting symptom to the vascular clinic is lower extremity pain that is often related to peripheral artery disease. The most common presenting symptom of peripheral artery disease is intermittent claudication, which is a reproducible discomfort in the muscles of the calf, thigh, or buttock, which occurs during exercise and is relieved with rest.¹ However, there are nonatherosclerotic etiologies of leg pain that should be considered by practitioners in leg pain and discomfort workup. Aimie and Landry² and Mintz and Weinberg³ conducted a review of the literature on the most common nonatherosclerotic causes of lower extremity claudication. They reported that external iliac endofibrosis is a unique condition among young, healthy cyclists.

External iliac artery endofibrosis (EIAE) was first described in the literature back in the early 1980s primarily in cyclists. EIAE is the narrowing of the iliac artery caused by thickening of the vessel, leading to luminal stenosis, which may reduce blood flow to the lower extremity. The narrowing is due to several factors such as bending of the vessel and cyclist's forward-leaning body position and repetitive hip flexion.^{4,5} Endofibrosis warrants better awareness because it is an uncommon disease that has presented difficulties in diagnosis and management.

Currently, there are no guidelines or standardized pathways for the management of EIAE.⁶

We discuss a case of left EIAE in an otherwise healthy male who has been a longstanding, avid cyclist. We describe his presenting symptoms, diagnosis with noninvasive imaging, management of his problem, and his outcome.

CASE REPORT

A 48-year-old male self-referred to the vascular clinic for evaluation and treatment of his left lower extremity pain. This referral was made after extensive research on his part because of his inability to manage and obtain treatment of his symptoms. He is an otherwise healthy male who has been a longstanding, avid cyclist. For two years, before his visit, he had noticed decreasing power in his left leg while cycling. In addition, he noted left hip, buttock, and thigh discomfort at the end of his rides. There was no history of trauma or injury to his leg. He had a left inguinal hernia repair in the past. History and physical examination were otherwise noncontributory.

Diagnostic evaluation included ankle-brachial index (ABI) with cycling protocol and computed tomography angiogram (CTA) of the abdomen and pelvis that was obtained with cycling provocative maneuver or cycling positions. Bilateral resting ABIs and waveforms were obtained and were normal (Figure 1). Duplex ultrasound of the left lower extremity demonstrated distal iliac artery velocity elevations of 580 cm/second, with evidence of endoluminal echoes and represented intimal thickening (Figure 2). During the cycling protocol, he cycled up to 300 watts and became symptomatic. At this point, the return of ABIs was checked back to normal. On his normal leg, ABIs dropped to 0.8 with exercise, and by 1 minute after exercise, it was 0.9 and back to normal at 2 minutes. On the symptomatic leg, the ABIs dropped to 0.3 with exercise, and at 5 minutes after exercise, the ABI was still 0.5, and at 10 minutes after exercise, it was only back up to 0.7. This was found to be consistent with hemodynamically significant

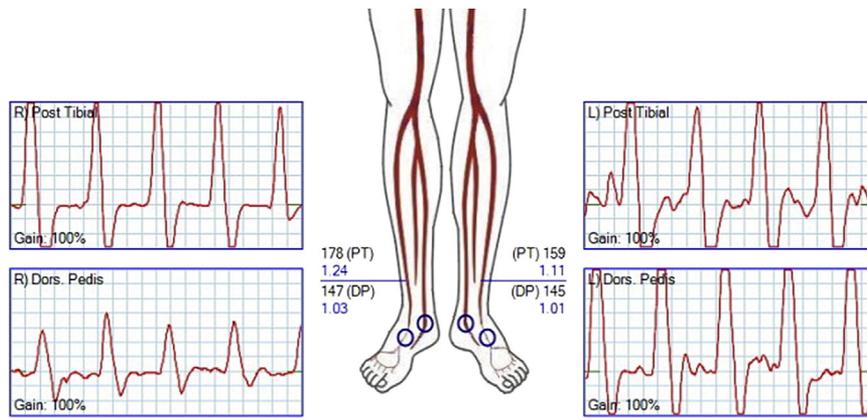
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1.24 Ankle/Brachial Index 1.11

Figure 1. Resting ankle-brachial indices and waveforms. Cycling exercise with ABI protocol includes a 5–6 mins of warmup cycling on a stationary bike. The power is increased by 50 watts every 2 minutes on the bike until the patient becomes symptomatic. This is followed by pressure measurements in the bilateral forearms and legs every minute until pressures return to normal. ABI = ankle-brachial index; PT = posterior tibial; DP = dorsalis pedis.

stenosis on the left leg (Figure 3). CTA of the abdomen and pelvis obtained with cycling maneuvers demonstrated left distal external endofibrosis (Figure 4).

The patient underwent left external iliac to common femoral artery endarterectomy and patch angioplasty. Intraoperative findings demonstrated significant intimal hyperplasia of the external iliac artery and proximal common femoral artery without evidence of atherosclerotic disease. The most significant amount of endofibrosis with intimal hyperplasia was found where the dissection was seen on the computed tomography scan (Figures 4 and 5).

The patient returned to our clinic 3 months after surgery for a follow-up visit having fully recovered from the procedure. He has returned to cycling and, for the most part, has remained without symptoms. His presurgical symptoms have resolved, and he is able to ride without pain. As part of his postoperative evaluation, he completed an ankle-brachial index with cycling protocol, which demonstrated significant improvement from baseline. There was no velocity elevation noted in the left iliac artery, and postexercise ABIs improved from 0.31 to 0.59 (Figures 6 and 7).

DISCUSSION

EIAE is an uncommon finding that usually affects young, healthy athletes or professional male endurance cyclists. This disease occurs in up to 10–20% of professional cyclists.⁷ The incidence of EIAE appears to be increasing because cycling is a booming sport with an increasing number of participants. EIAE can also be observed in other groups of highly trained endurance athletes such as triathletes, long-distance runners, skiers, rowers, and rugby players (Veraldi, Criscenti, Scorsone, Zingaretti, Gnoni, & Mezzetto, 2015). The disease is more prevalent in men because more men practice competitive cycling and endurance exercises than women. Research reported that 87% of the US competitive cyclists are male and 12% are female with age ranges between 35 and 44 years (USA Cycling, 2018). These healthy, young athletes seek treatment from health-care professionals due to leg or hip pain, fatigability, or paresthesia of their



Figure 2. (A) Arterial duplex of left lower extremity. Area of endofibrosis or dissection (green arrow). (B) Limited duplex evaluation of left lower extremity demonstrated distal external iliac artery velocity elevations of 580 cm per second. Left common femoral artery waveforms were turbulent. PT = posterior tibial.

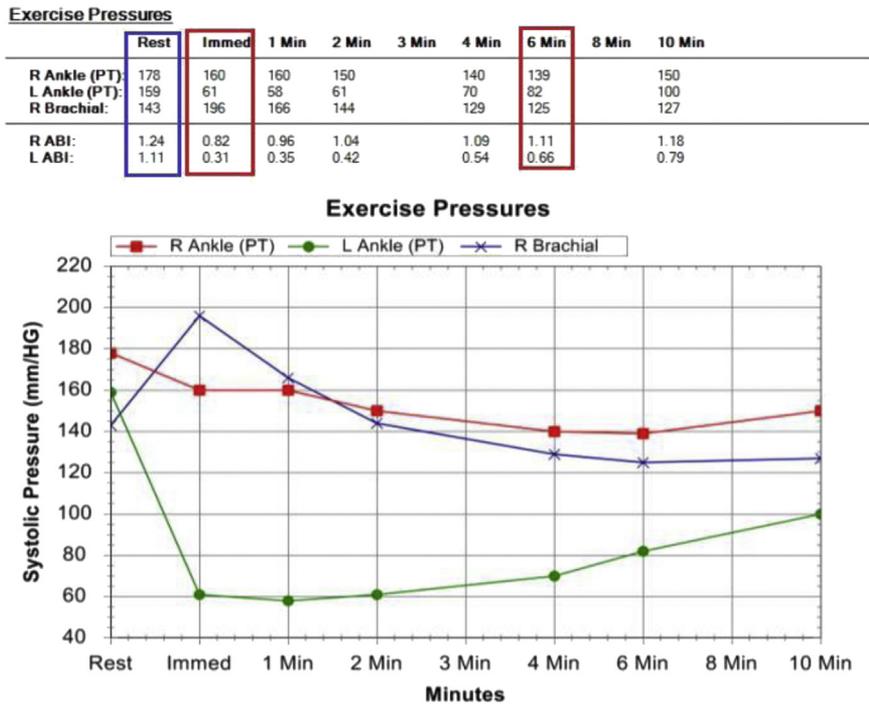


Figure 3. Cycling protocol and exercise pressures. The patient was able to perform nine minutes of cycling on a stationary bike test with increasing resistance every three minutes after five minutes of warmup. The cycling test was stopped due to the severity of the left lower extremity symptoms. There was a mild decrease in right postexercise ABIs with return to baseline after 2 minutes of rest. There was a severe drop in the left lower extremity postexercise ABIs of 0.31, with left ABI recorded as 0.79 after 10 minutes of rest. ABI = ankle-brachial index.

lower extremities that occurs after few minutes of endurance training and resolves quickly with rest.

The pathophysiology of this disease is thought to be related to a repetitive injury of the artery from repeated stretching and compression, leading to the development of endofibrosis. Recurrent injury and mechanical stress of the artery cause an intravascular lesion that resembles arterial wall thickening. The intimal thickening causes a decrease in the arterial lumen, which leads to diminished blood flow in the lower extremity which produces symptoms of exertional claudication.⁸ If left untreated, EIAE can lead to arterial dissection, atheroma, thrombosis, and can eventually predispose the patient to atherosclerotic disease in that area.

The progressive narrowing of the external iliac artery eventually leads to symptoms that include muscle fatigue, cramping, and pain in the affected leg which occur with intense exercise. Most often EIAE is unilateral and usually seen in the left external iliac artery. It can also extend into the common iliac, common femoral, and profunda femoris arteries. Research shows that only 15% of patients have a bilateral disease.⁹ Another reason for left EIAE being more common is because of increasing incidence of lower lumbar degenerative scoliosis as we age. The

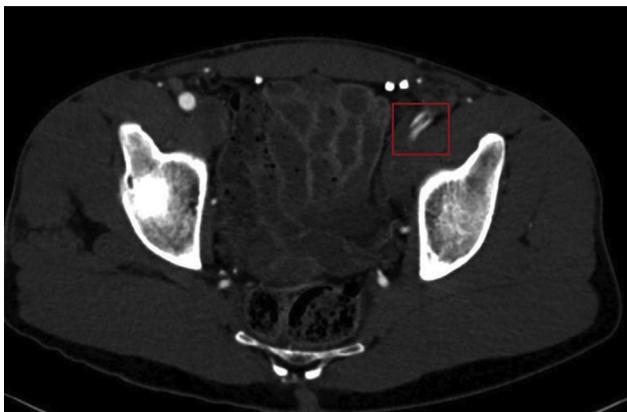


Figure 4. CTA was performed in the cycling position. The cycling position included flat and then tucked in position. The left iliac artery is shown with dissection versus endofibrosis (red box). CTA = computed tomography angiogram.

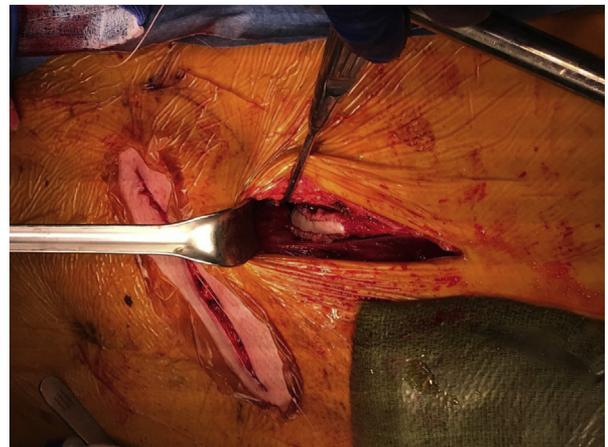


Figure 5. A left inguinal hernia repair mesh was found to be compressing the external iliac artery as it enters the leg, with endostaples prohibiting retroperitoneal approach to the iliac artery. Therefore, a second incision from the common femoral artery was made upward via a vertical incision overlying the common femoral artery.

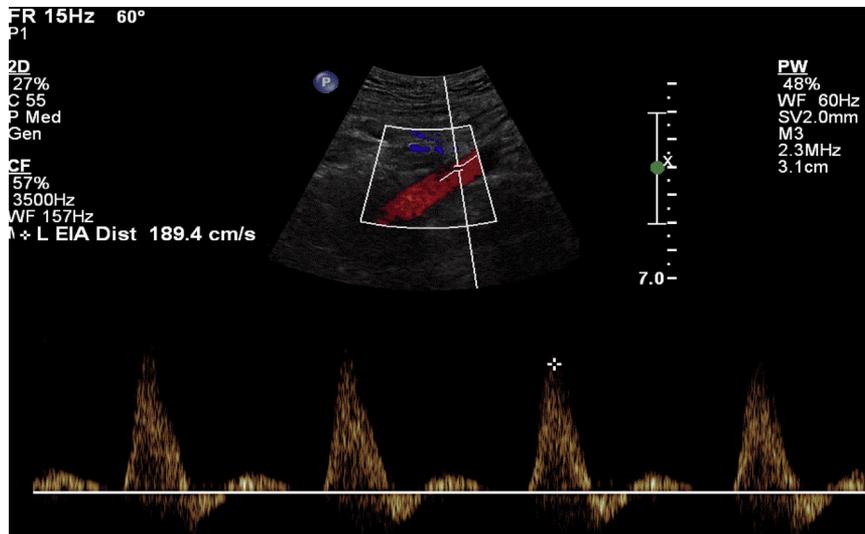


Figure 6. Postoperative duplex ultrasound demonstrated patent left external iliac artery reconstruction without evidence of stenosis or significant velocity elevations. Left external iliac artery waveforms and velocities on 3-month follow-up appointment. Duplex imaging demonstrated widely patent external iliac artery with multiphasic waveforms and normal velocities.

spine degenerates more commonly convex to the left side, and the convex side found to develop psoas hypertrophy, which is associated with EIAE.¹⁰ The external iliac artery is located distally in the anterior portion of the psoas muscle and anterior to the axis of rotation of the hip. This location leads to arterial kinking during hip flexion. The artery can be compressed if there is psoas muscle hypertrophy.

There is a profound delay in diagnosing EIAE. On average, the time to diagnosis averages between 12 and 41 months.⁹ EIAE is challenging to diagnose because the patient's symptoms

are only present after extreme exercise and physical examination. Initial vascular ultrasound testing results are normal at rest, making the provider to think that the blood flow is normal. This delay in diagnosis was evident in our present case study in which diagnosis was delayed for over several years. Noninvasive vascular ultrasound imaging is one of the diagnostic criteria used to assess for EIAE. This is in line with the Delphi Consensus Study in which a consensus was reached that patients should be exercised until they develop symptoms and that pressures should be measured with ultrasound within 1 minute of exercise cessation.⁶

Exercise Pressures

	Rest	Immed	1 Min	2 Min	3 Min	4 Min	6 Min	8 Min	10 Min
R Ankle (PT):	166	171	169	158	149	149	150		
L Ankle (PT):	165	119	140	143	140	141	149		
L Brachial:	128	202	185	161	152	141	130		
R ABI:	1.30	0.85	0.91	0.98	0.98	1.06	1.15		
L ABI:	1.29	0.59	0.76	0.89	0.92	1.00	1.15		

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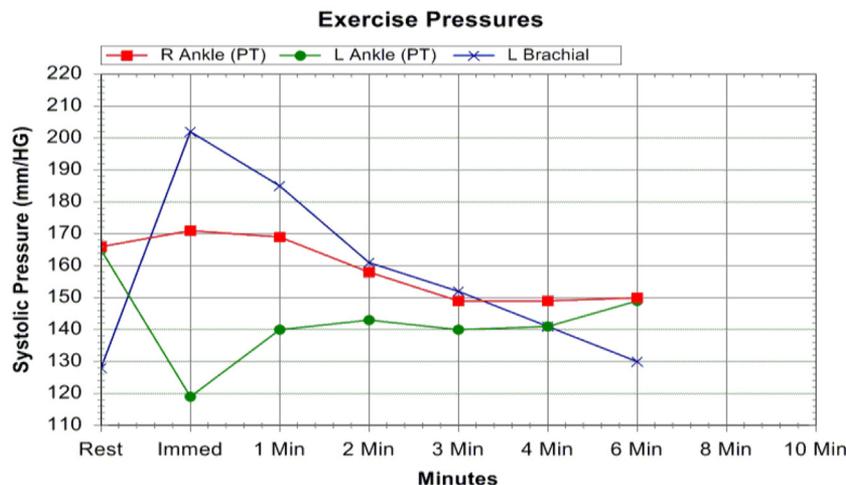


Figure 7. Resting and postexercise (cycling test) pressure measurements on 3-month follow-up appointment. Left postexercise pressures improved compared with those in the initial evaluation. ABI = ankle-brachial index. PT = posterior tibial.

In our case report, the vascular ultrasound laboratory has developed a cycling protocol in which a patient starts with a 6-minute warmup on a bike and gradually increases resistance by 50 watts for every 3-minute interval until symptoms become unbearable. Then the patient is asked to promptly return to the bed where postexercise ABIs are measured by two ultrasound technologists. The first measurement is taken immediately after the exercise, and the next measurement is performed in 1- to 2-minute intervals for the next ten minutes or until ABIs return to baseline. The common iliac, external iliac, and common femoral arteries are scanned with the patient in a supine position and with their affected hip externally rotated. Curvilinear and linear probes may be used to obtain 2D, color and pulse wave doppler images to evaluate for intimal thickening, color flow aliasing, and significant velocity elevations in the affected areas.

The doppler ultrasound was found to be a more discriminatory tool in the diagnosis of iliac endofibrosis.¹¹ Investigators looked at a total of 24 patients who were referred for suspected endofibrosis. Color doppler ultrasound was used with a through B-mode assessment to identify preexercise and postexercise pressures and velocities. Based on the result of the doppler ultrasound, 15 of the 24 patients were diagnosed with endofibrosis and underwent successful surgical management. Peake, D'Abate, Farrah, Morhan, and Hinchliffe⁷ also reported their experience with noninvasive testing in the diagnosis of iliac endofibrosis and found that doppler ultrasound with pressure index was sufficient. They showed that 84% of their case studies had a postexercise ABI drop of > 0.3. computed tomography angiography or magnetic resonance angiography can also provide comprehensive information regarding the location and severity of the arterial stenosis. It is also a great tool to use in presurgical planning.

Peach et al¹² completed a review of the literature to examine the assessment and management of iliac endofibrosis. The surgical management technique available includes patch angioplasty, vessel shortening, bypass, and inguinal ligament release. They also found a lack of consistent methodology to identify the best treatment options for EIAE. In our case study, the patient underwent successful endarterectomy and patch angioplasty. He was symptom free on follow-up. Peake, D'Abate, Farrah, Morgan, and Hinchliffe⁷ also reported successful surgical management with endarterectomy and patch angioplasty over a seven-year period. Similarly, Franco, Rigberg, and Ruehm¹³ described resolution of symptoms with patch angioplasty and resection.

In our institution, long-term functional outcomes of patients with EIAE were evaluated. Wu, Olcott, and Lee¹⁴ evaluated the operative results after surgical repair of symptomatic EIAE in cyclists over a 15-year period. They found that 82% of postsurgical patients were able to return to their prior level of competitive cycling, although 50% of those patients had small recurrence of mild symptoms.

CONCLUSION

EIAE is a disorder characterized by intimal thickening that is most often seen in athletes and cyclists. This diagnosis is uncommon but should be considered in the presentation of claudication in otherwise young, healthy individuals.

Workup should include a thorough history and physical examination, cycling exercise ABIs, and positional CTA. Once a diagnosis of EIAE is established, surgical treatment is recommended.

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