A Rare Case of Leriche Syndrome presenting as Acute Paraplegia in a known case of Mitral Stenosis with Mitral Valve Replacement

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Abstract
Leriche Syndrome is a aortoiliac occlusive disease (AIOD) which is atherosclerosis affecting the distal abdominal aorta, iliac arteries, and femoropopliteal vessels [1]. Leriche Syndrome when symptomatic presents with a classical triad of claudication importance and absence of femoral pulses. In this article we discuss a case of 49-year-old female, a known case of Rheumatic Valvular Heart Disease with mitral stenosis, post mitral valve replacement (prosthetic mitral valve in situ), who presented with complains of sudden onset weakness of both lower limbs associated with tingling and numbness. On examination, femoral and peripheral vessels were not felt. Hereby, this article is about Leriche Syndrome in a patient of Rheumatic Valvular Heart Disease with post mitral valve replacement presenting as acute paraplegia.

Keywords: sudden onset weakness of both lower limbs, claudication pain, absence of femoral and peripheral pulses.

Introduction

Leriche Syndrome, a aortoiliac occlusive disease (AIOD) most commonly caused by atherosclerosis. The extent and localization of atherosclerosis arteries determine the classification of disease. Type I (confined to the distal abdominal aorta and common iliac arteries), Type II (predominately distal abdominal aorta with disease extension into common iliac and external iliac arteries), and Type III (affecting the aortoiliac segment and femoropopliteal vessels) [2]. Risk factors for which can be modifiable or non-modifiable. Modifiable include hypertension, diabetes mellitus, nicotine, hyperlipidemia, hyperglycemia, and homocysteine. Non-modifiable include age, race, and family history [3]. Leriche Syndrome occurs secondary to atherosclerotic induced arterial wall injury leading to formation of a plaque in the arterial lumen. A detailed history is essential in determining the location, severity and duration of symptoms. Due to collateral supply limb threatening ischemia is not universal [5].

Case Report

A 49-year-old female, housewife, right-handed person, from a lower socio-economic background, came with chief complaints of pain in the hip, buttocks and thigh region radiating to both lower limbs, weakness of both lower limbs associated with tingling and numbness. Weakness in both lower limb was acute in onset, in such a way that patient could not able to walk which was associated accompanied by sensory loss below the T10-level. Patient also gave history of urinary and bowel incontinence. No history of trauma. No history of lower back ache.

Past History - The patient is a known case of Rheumatic Valvular Heart Disease with mitral stenosis with prosthetic mitral valve in situ on Tab. Warfarin 1mg OD, Tab. Digoxin 0.25mg OD and Tab. Lasilactone 20/50mg BD.

No history of Diabetes, Hypertension, Tuberculosis or Malignancy. No past surgical history. No past history of vaccination. No significant personal and family history.

Clinical Examination: Afebrile, Pulse was 88/min in the right upper limb, peripheral pulse was not felt in both lower limbs. Peripheries were cold. BP was 110/80mmHg measured in supine position in the right upper limb, SpO2 was 95 % on room air, BSL- 128mg/dL, Ankle brachial index was 0.4.

No pallor, icterus, cyanosis, clubbing, lymphadenopathy, oedema was present. Systemic Examination: On CVS examination,

ECG was suggestive of atrial fibrillation associated with left atrial enlargement.

CNS examination: Higher mental functions examination, Patient was conscious, oriented to time, place, and person. Cranial nerve examination**:** All cranial nerve examinations were normal. On Motor Examination, hypotonia was present in both lower limbs, power in both lower limbs was 0/5, Reflexes were absent in both lower limbs. Bilateral plantar were mute. Motor examination of both upper limbs were normal. All modalities of sensation were lost below T10 level. No signs of meningitis present.

CT AORTOGRAPHY showing the infrarenal aorta about 3.2cm distal to the origin of the right renal artery shows complete hypodense filling defect of size approx. 39 x 14 x 13 mm (CC X AP X TR), extending into the proximal halves of bilateral common iliac arteries -suggestive of thrombus / embolus. No obvious collateral vessels noted. Partial filling defect in the distal halves of bilateral common iliac arteries – suggestive of partial thrombosis. Bilateral external iliac arteries show reduced caliber but normal opacification. Bilateral internal iliac arteries appear normal. Inferior mesenteric artery shows reduced opacification. Visualized bilateral common femoral arteries, profunda femoris and superficial femoral arteries show reduced caliber but normal opacification.

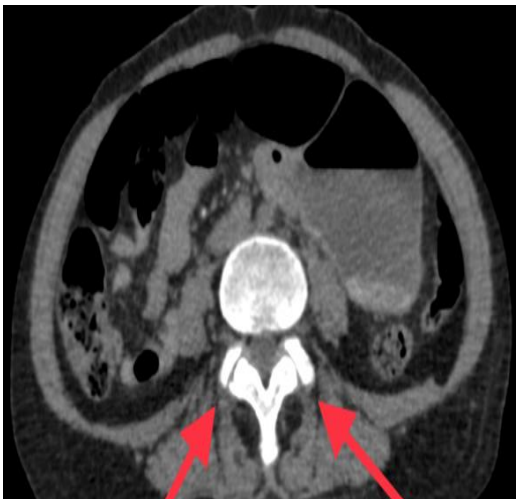
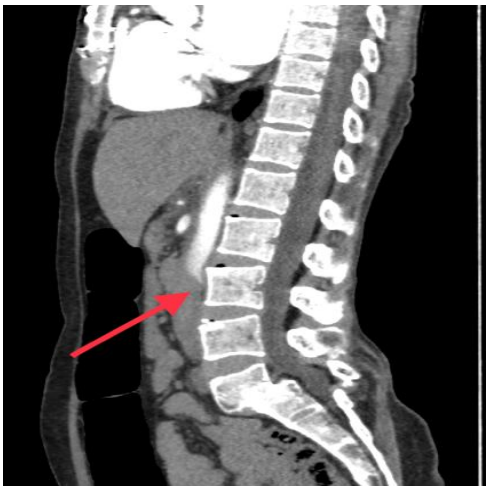


Image 1



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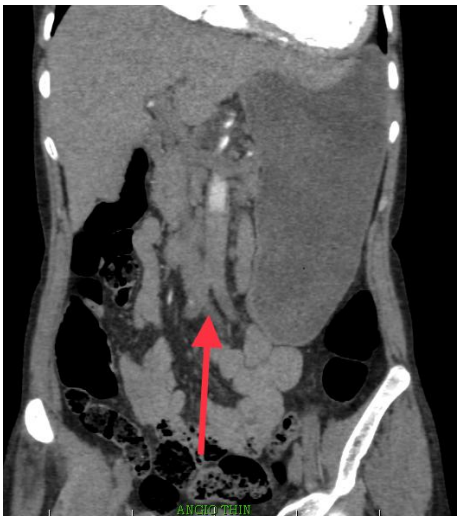

Image 3

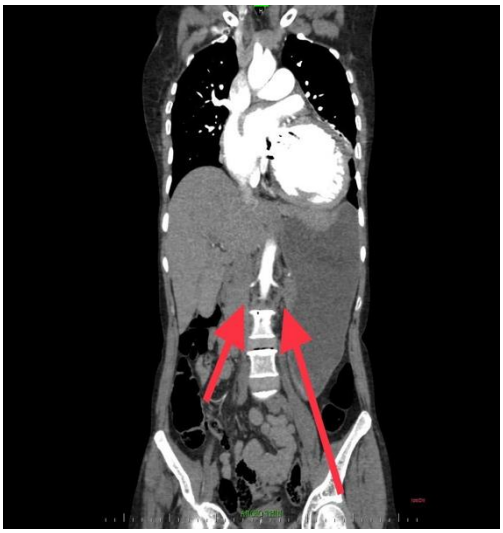

Image 4

Image 1 shows filling defect in **bilateral common iliac arteries** ; **Image 2 and 3** shows filling defect in **infrarenal aorta**; **Image 4** shows filling defect at **bifurcation of bilateral common iliac arteries**

Lab Investigations- CBC, LFT, RFT, Lipid Profile and Serum electrolytes were normal. Serum Homocysteine levels were elevated (>50ng/ml). Anti-cardiolipin antibodies were negative. Protein-C / S deficiency tests, Factor V Leiden mutation tests, and anti-thrombin III level tests were normal. PT was 13 secs and INR was 0.93

2D echo was suggestive of global LV Hypokinesia, LVEF = 20-25%, Dilated Left Atrium and Left ventricle, Prosthetic mitral valve in situ, Right Ventricular Hypertrophy with moderate Tricuspid Regurgitation, and moderate Pulmonary Arterial Hypertension.

Discussion

An aortoiliac occlusive disease is a form of central artery disease involving the blockage of the abdominal aorta as it transitions into the common iliac arteries. It included a triad of claudication of the buttocks and thighs, absent or decreased femoral pulses, and erectile dysfunction (seen in males). This combination is known as Leriche syndrome. However, any number of symptoms may present, depending on the distribution and severity of the disease, such as muscle atrophy, slow wound healing in the legs, and critical limb ischemia [5]. A lipid profile, HbA1c, Lipoprotein A, and homocysteine levels should be evaluated [3]. Peripheral pulses should be checked. An ankle-brachial index (ABI), the first screening test should also be performed to evaluate the perfusion of the lower extremities. An ABI lower than 0.9 is considered abnormal and indicates the patient has PAD significant enough to cause claudication [6]. Coronary artery disease (CAD) is seen in 10% - 71% of patients with Leriche Syndrome, so obtaining an ECG as well to rule out CAD is recommended [4]. Surgical treatment includes thromboendarterectomy (TEA), aortobifemoral bypass (AFB), and percutaneous transluminal angioplasty (PTA) with or without stenting [3] [7] [8]. PTA and stenting are the intervention of choice in patients with multiple comorbidities. Medical management includes smoking cessation, management of diabetes, antiplatelet, and statin therapy, in addition to antihypertensive therapy. Daily exercise for 30 minutes should be suggested. Intolerable claudication pain should be the endpoint for exercise [9]. Cilostazol, a phosphodiesterase III inhibitor, can be used to treat claudication symptoms of the patient and is beneficial in graft patency and prevention of stenosis [10]. Signs of critical limb ischemia include sudden onset of pain in the affected extremity, paresthesia, paleness or coolness, and severely diminished or absent pulses [11]. Without treatment, the prognosis of Leriche Syndrome is poor. In some cases where progression is slow or collaterals develop since the onset of LS, these collaterals may act as a self-compensating mechanism [12] The complication of Leriche Syndrome includes Limb ischemia [13] as well as heart failure, myocardial ischemia/infarction, gangrene, and even death [14].

Conclusion:

The acute form of this syndrome is rare. The neurological disorders in the present patient were explained by spinal cord ischemia (Adamkiewicz's artery). Vascular myelopathies, especially vascular diseases of the aorta, must be considered in acute paraplegia.

Patients with Leriche Syndrome usually have a poorer prognosis as they present very late in the course of the disease and when limited or no interventions are possible. As Leriche Syndrome is so complex, a purposeful interprofessional team should be involved [15] when Leriche Syndrome is suspected. The team includes a physician, cardiologist, radiologist, vascular surgeon, and interventional cardiologist. This approach helps in increasing success outcomes with better management and patient satisfaction. In our case report, the female came at a late stage with already severe occlusion in the infrarenal aorta and its branches, leading to rapid progression and death. It is thus essential that medical professionals, especially cardiologists, vascular surgeon, radiologist and physician, be aware of various causes of acute paraplegia or quadriplegia, and must check for peripheral pulses in all such patients, especially those with suspected Leriche Syndrome.

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Referrences:

1. Pascarella L, Aboul Hosn M. Minimally Invasive Management of Severe Aortoiliac Occlusive Disease. J Laparoendosc Adv Surg Tech A. 2018 May;28(5):562-568. doi: 10.1089/lap.2017.0675. Epub 2018 Jan 18. PMID: 29346011.

2. Wooten C, Hayat M, du Plessis M, Cesmebasi A, Koesterer M, Daly KP, Matusz P, Tubbs RS, Loukas M. Anatomical significance in aortoiliac occlusive disease. Clin Anat. 2014 Nov;27(8):1264-74. doi: 10.1002/ca.22444. Epub 2014 Jul 25. PMID: 25065617.

3. Frederick M, Newman J, Kohlwes J. Leriche syndrome. J Gen Intern Med. 2010 Oct;25(10):1102-4. doi: 10.1007/s11606-010-1412-z. Epub 2010 Jun 22. PMID: 20568019; PMCID: PMC2955474.

4. Diehm C, Schuster A, Allenberg JR, Darius H, Haberl R, Lange S, Pittrow D, von Stritzky B, Tepohl G, Trampisch HJ. High prevalence of peripheral arterial disease and co-morbidity in 6880 primary care patients: cross-sectional study. Atherosclerosis. 2004 Jan;172(1):95-105. doi: 10.1016/s0021-9150(03)00204-1. PMID: 14709362.

5. Ahmed S, Raman SP, Fishman EK. CT angiography and 3D imaging in aortoiliac occlusive disease: collateral pathways in Leriche syndrome. Abdom Radiol (NY). 2017 Sep;42(9):2346-2357. doi: 10.1007/s00261-017-1137-0. PMID: 28401281.

6. de Groote P, Millaire A, Deklunder G, Marache P, Decoulx E, Ducloux G. Comparative diagnostic value of ankle-to-brachial index and transcutaneous oxygen tension at rest and after exercise in patients with intermittent claudication. Angiology. 1995 Feb;46(2):115-22. doi: 10.1177/000331979504600204. PMID: 7702195.

7. Metcalfe MJ, Natarajan R, Selvakumar S. Use of extraperitoneal iliac artery endarterectomy in the endovascular era. Vascular. 2008 Nov-Dec;16(6):310-5. doi: 10.2310/6670.2008.00080. PMID: 19344587.

8. Krankenberg H, Schlüter M, Schwencke C, Walter D, Pascotto A, Sandstede J, Tübler T. Endovascular reconstruction of the aortic bifurcation in patients with Leriche syndrome. Clin Res Cardiol. 2009 Oct;98(10):657-64. doi: 10.1007/s00392-009-0052-y. Epub 2009 Aug 14. PMID: 19685001.

9. Fakhry F, Spronk S, van der Laan L, Wever JJ, Teijink JA, Hoffmann WH, Smits TM, van Brussel JP, Stultiens GN, Derom A, den Hoed PT, Ho GH, van Dijk LC, Verhofstad N, Orsini M, van Petersen A, Woltman K, Hulst I, van Sambeek MR, Rizopoulos D, Rouwet EV, Hunink MG. Endovascular Revascularization and Supervised Exercise for Peripheral Artery

Disease and Intermittent Claudication: A Randomized Clinical Trial. JAMA. 2015 Nov 10;314(18):1936-44.

10. Tara S, Kurobe H, de Dios Ruiz Rosado J, et al. Cilostazol, Not Aspirin, Prevents Stenosis of Bioresorbable Vascular Grafts in a Venous Model. Arteriosclerosis, Thrombosis, and Vascular Biology. 2015 Sep;35(9):2003-2010. DOI: 10.1161/atvbaha.115.306027. PMID: 26183618; PMCID: PMC4548543.

11. Yoon DH, Cho H, Seol SJ, Kim T. Right calf claudication revealing leriche syndrome presenting as right sciatic neuropathy. Ann Rehabil Med. 2014 Feb;38(1):132-7. doi: 10.5535/arm.2014.38.1.132. Epub 2014 Feb 25. PMID: 24639938; PMCID: PMC3953357.

12. Morotti A, Busso M, Cinardo P, Bonomo K, Angelino V, Cardinale L, Veltri A, Guerrasio A. When collateral vessels matter: asymptomatic Leriche syndrome. Clin Case Rep. 2015 Nov;3(11):960-1. doi: 10.1002/ccr3.390. Epub 2015 Sep 10. PMID: 26576282; PMCID: PMC4641484.

13. Pillai J, Monareng T, Rangaka TB, Yazicioglu C, Jayakrishnan R, Veller MG. Aorto-internal iliac artery endovascular reconstruction for critical limb ischaemia: a case report. S Afr J Surg. 2015 Oct 8;53(1):26-7. doi: 10.7196/sajs.2458. PMID: 26449602.

14. JOHNSON JK. Ascending thrombosis of abdominal aorta as fatal complication of Leriche's syndrome. AMA Arch Surg. 1954 Nov;69(5):663-8. doi: 10.1001/archsurg.1954.01270050067013. PMID: 13206556.

15. Rubimbura V, Rostain L, Duval AM, Akakpo S, Boukantar M, Boiron P, Mouillet G, Gallet R, Belarbi A, Le Corvoisier P, Dubois-Randé JL, Teiger E. Outcomes and safety of same-day discharge after percutaneous coronary intervention: A 10-year single-center study. Catheter Cardiovasc Interv. 2019 Jul 01;94(1):105-111.