Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2016; 4(6):376-378 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2016.v04i06.004

A case of fibro muscular dysplasia with abdominal aortic aneurysm

Danish E[1], Vishnu H[2], Unnikrishnan P[3], Sharath K Krishnan[4], T.M. Anoop[5]
[1]Assistant professor, Department of Medicine, Calicut Medical College, Kozhikode, Kerala, India
[2, 3]Senior Resident, Department of Medical Oncology, Regional cancer center, Thiruvananthapuram, Kerala, India
[4]Senior resident in Surgical Oncology, Regional cancer center, Thiruvananthapuram, Kerala, India
[5]Assistant professor, Department of Medical Oncology, Regional Cancer Centre, Thiruvananthapuram, Kerala, India

***Corresponding author**
Dr. Anoop. T.M
Email: dranooptm@yahoo.co.in

Abstract: Fibro muscular dysplasia is a non-atherosclerotic, non-inflammatory disease that most commonly affects the renal and internal carotid arteries. Fibro muscular dysplasia as a cause of abdominal aortic aneurysm is rarely reported. Here we report a case of fibro muscular dysplasia associated with abdominal aortic aneurysm in a 35 year old lady, who presented with vague abdominal discomfort and claudication pain of both lower limbs.
Keywords: Fibro muscular dysplasia, aneurysm, non-atherosclerotic.

INTRODUCTION

Fibro muscular dysplasia (FMD) is a group of non-atherosclerotic, non-inflammatory arterial diseases that most commonly involve the renal and carotid arteries [1]. Most common presentations are renovascular hypertension and stroke. Fibro muscular dysplasia as a cause of abdominal aortic aneurysm is rather rarely reported [2, 3, 4]. Here we report a case of fibro muscular dysplasia associated with abdominal aortic aneurysm who presented with vague abdominal discomfort and claudication pain of both lower limbs.

CASE REPORT

A 35 year old lady was admitted for evaluation of vague abdominal discomfort and bilateral claudicating pain of both lower limbs. During her second pregnancy, she was detected to have hypertension and was on follow up with 5 mg of Amlodipine 4 years back. On examination, she had blood pressure of 140/90 mm of Hg in the left upper limb. Both dorsalis pedis arterial pulsations was feeble .An abdominal bruit heard was heard. Systemic examination was normal. Laboratory investigations showed hemoglobin of 12 gm/dl, total count of 8500 cells with polymorphs 56 %, lymphocyte 43%, Erythrocyte sedimentation rate of 3mm at first hour and platelet count 1 lakh. Renal function tests showed blood urea of 30 mg per day and serum creatinine 1.3mg/dl .Liver function test and electrolytes were normal .Urine microscopy was normal. Doppler evaluation of lower limbs showed monophasic pattern in both dorsalis pedis. Anti Nuclear Antibody was negative. Fasting lipid profile was normal. Ultrasnography of abdomen revealed an abdominal aortic aneurysm.

A Magnetic Resonance Angiography showed string of beads appearance in distal renal arteries (Fig A), splenic (Fig B) suggestive of fibro muscular dysplasia. An abdominal aortic aneurysm of 3.5cms width and 6cms long extending up to origin of renal arteries was also noted. (Fig A). A temporal artery biopsy was performed. Histopathological changes of intimal thickening, medial fibroplasia and hyperplasia with disruption and dissection of internal ilastic lamina with no inflammatory infiltrate or giant cells characteristic of fibro muscular dysplasia was seen in the biopsy specimen (Fig C).

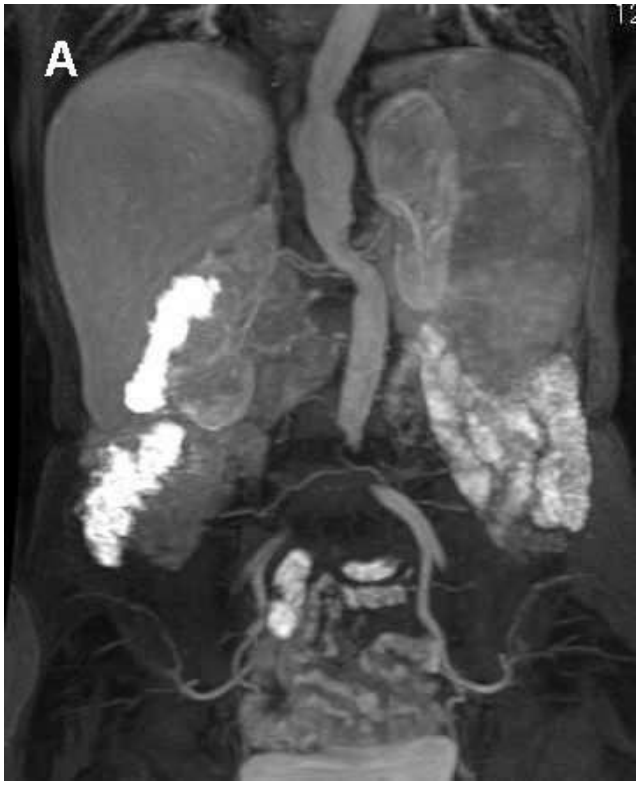

Fig A: Magnetic Resonance Angiography showing abdominal aortic Aneurysm of 3.5cms width and 6cms long is extending up to origin of renal Arteries and characteristic finding of string of beads appearance in distal renal arteries suggestive of fibro muscular dysplasia.

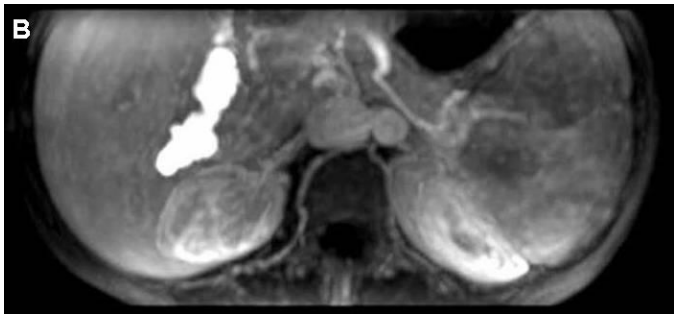

Fig B: Magnetic Resonance Angiography showing string of beads appearance in left splenic arteries

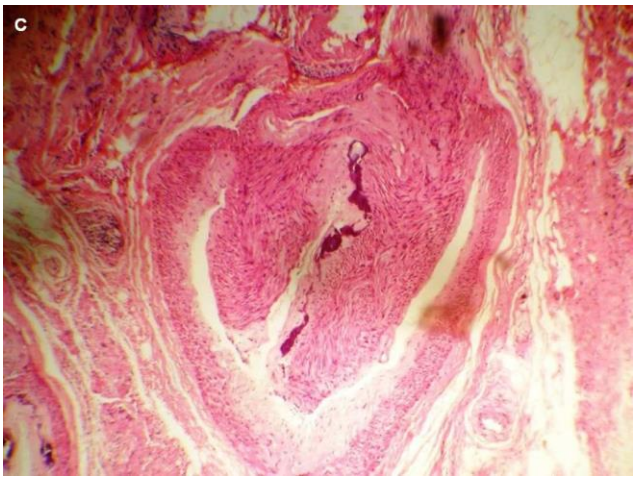

Fig C:Temporal artery biopsy showing changes of intimal thickening, medial fibroplasia and hyperplasia with disruption and dissection of internal ilastic lamina with no inflammatory infiltrate or giant cells characteristic of fibro muscular dysplasia

DISCUSSION

Fibro muscular dysplasia (FMD) is a group of non-atherosclerotic, non-inflammatory arterial diseases that most commonly involve the renal and carotid arteries [1]. Most common presentations are renovascular hypertension and stroke. Fibro muscular dysplasia as a cause of abdominal aortic aneurysm is rather rarely reported [2, 3, 4]. Clinical presentations of patients with FMD are varied. Cervical FMD may presents with Horner's syndrome, dissection, transient ischemic attack, ischemic stroke or subarachnoid hemorrhage. FMD affecting extra-renal and extra-cervical arteries has been reported in celiac, superior and inferior mesenteric, hepatic, splenic, and coronary arteries [5]. Mesenteric artery FMD can present with mesenteric ischemia or rarely mesenteric infarction .Patients with lower limb artery FMD may present with cold legs, intermittent claudication, or evidence of distal embolic disease. Subclavian artery FMD may present with arm weakness, paresthesias, claudication of upper limbs and subclavian steal syndrome [6].

Harrison and McCormack established criteria for the pathological classification of FMD [7]. Medial fibroplasia is most common (60-70%) and they often affecting the distal two thirds of the main artery and its branches, giving an appearance of "strings of beads" on arteriogram .Diagnosis of mesenteric FMD is challenging and it can closely mimic vasculitis and atherosclerosis. Catheter angiography remains the gold standard for the diagnosis of fibro muscular dysplasia. Management depends on the presentation of patients.

Surgery should generally be used only to treat macro aneurysms and in cases with complex FMD that extends to segmental arteries [1]. In this case, patient had predominant abdominal aortic aneurysm with FMD involving distal the renal and splenic arteries which are rarely reported.

REFERENCES

1. Olin JW; Recognizing and managing fibro muscular dysplasia. Cleve Clin J.Med. 2007; 74(4):273-4, 277-82.
2. Matsushita M, Yano T, Ikezawa T, Sakurai T, Nimura Y, Shionoya S; Cardiovasc Surg. Fibro muscular dysplasia as a cause of abdominal aorticaneurysm.1994; 2(5):615-8.
3. Odero A, Bozzani A, Arici V, Agozzino M; Hypoplasia and fibromusculardysplasia of infrarenal abdominal aorta with downstream aneurysm: case report and review of the literature. J Vasc Surg. 2008; 48(6):1589-92.
4. Letsch R, Kantartzis M, Sommer T, Garcia M; Arterial fibromuscular dysplasia. Report of a case with involvement of the aorta and review of theliterature. Thorac Cardiovasc Surg. 1980; 28(3):206-10.
5. Luscher TF, Lie JT, Stanson AW, Houser OW, Hollier LH, Sheps SG; Arterial fibro muscular dysplasia. Mayo Clin Proc 1987, 62: 931-952.
6. Plouin PF**,** Perdu J**,** La Batide-Alanore A, Boutouyrie P, Gimenez-Roqueplo AP, Jeunemaitre X; Fibromuscular dysplasia.Orphanet J Rare Dis. 2007; 7: 2:28.
7. Harrison EG Jr, McCormack LJ; Pathological classification of renal arterial disease in renovascular hypertension. Mayo Clin Proc 1971; 46:161-167.