

# Central Retinal Artery Occlusion in a Young Female with Fibromuscular Dysplasia

Rajan Paul\*, Dai Barr

Aberystwyth Eye Centre, Hywel Dda University Health Board, Abersywyth SY23 2EG United Kingdom

\*Corresponding author: [rajan.paul@wales.nhs.uk](mailto:rajan.paul@wales.nhs.uk)

Received April 13, 2015; Revised April 26, 2015; Accepted April 28, 2015

**Abstract** Fibromuscular dysplasia (FMD) is a rare non-arteriosclerotic, non-inflammatory arterial disease affecting the large arteries e.g renal, carotid, mesenteric and coronary. We describe a case of Central retinal artery occlusion with embolus reorientation in a patient with FMD.

**Keywords:** *Fibromuscular dysplasia, central retinal artery occlusion, embolus reorientation*

**Cite This Article:** Rajan Paul, and Dai Barr, "Central Retinal Artery Occlusion in a Young Female with Fibromuscular Dysplasia." *American Journal of Medical Case Reports*, vol. 3, no. 5 (2015): 148-152. doi: 10.12691/ajmcr-3-5-9.

## 1. Abstract

To report a case of Central Retinal Artery Occlusion in a young female with Fibromuscular Dysplasia with the characteristic "String of beads" sign on magnetic resonance angiography characteristic of medial Fibromuscular dysplasia.

Fibromuscular dysplasia has been associated with coagulation disorders, genetic and hormonal anomalies, stress, and smoking however, the true cause of Fibromuscular dysplasia is not known. Fibromuscular dysplasia is pathologically characterized by circumferential or eccentric collagen deposition, and is classified according to the arterial wall layer that is primarily affected, ie, intimal, medial, or adventitial.

Central Retinal Artery Occlusion in patients with Fibromuscular dysplasia is rare in literature.

## 2. Introduction

Fibromuscular dysplasia (FMD) principally involves the renal and carotid arteries [1] and was first described by Leadbetter and Burkland, 1938 [2].

It is a rare disease and may present as stenosis, aneurysm, dissection, or occlusion while other cases may be clinically asymptomatic, being accidentally discovered while imaging for some other problem or on detection of an asymptomatic bruit [3,4].

In our case it was the Central retinal artery occlusion that drew our attention to the presence of FMD in this young female.

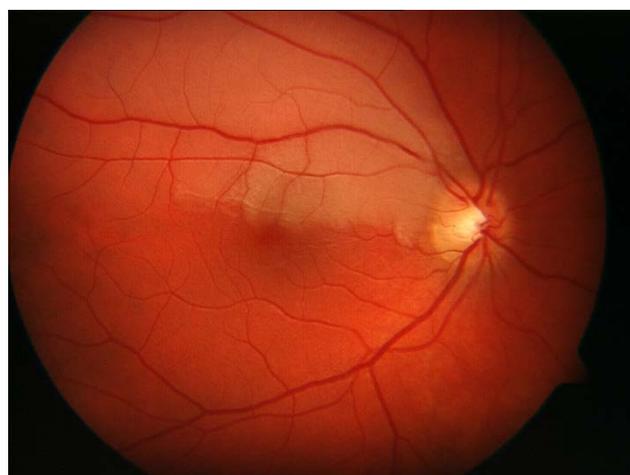
## 3. Case Report

### 3.1. History

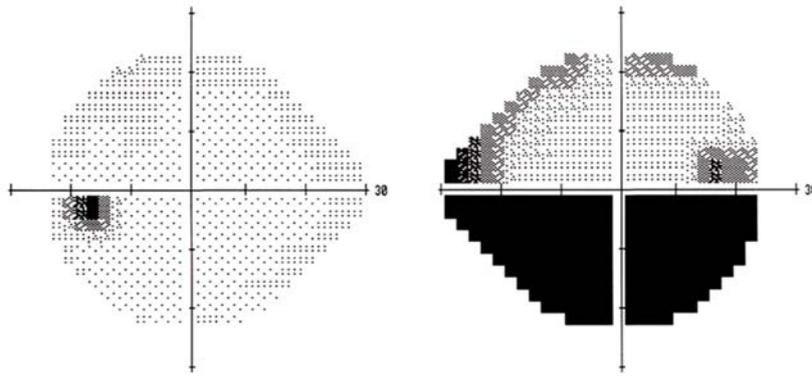
A 22 year old female presented in our clinic with complaints of a Right lower field defect on waking up a week back. There was no recovery, no pain, redness or any other symptoms. The left eye was asymptomatic. There were no neurological symptoms. There was no significant past ocular history. She suffered from classical migraines during her teens. She was not on any treatment including oral contraceptive pills.

### 3.2. On Examination

Her best corrected visual acuity (BCVA) was Right 6/60 and Left 6/5. She had relative afferent pupillary defect (RAPD) Grade IV. The colour vision with Ishihara's Test Plate was Right 8/12 and Left 10/12. Her ocular motility was full. There was no proptosis or abnormal synkinetic movements. Corneal sensations were bilaterally present and equal, there was no cranial neuropathy. The anterior segments were unremarkable. Intraocular pressures were 18 mmHg in each eye.



**Figure 1.** Right fundus: Upper temporal retina appears whitish due to infarction and edema of inner retinal layers



**Figure 2.** Right Inferior altitudinal defect



**Figure 3.** Right FFA: Early filling of vessels which were expected to be occluded



**Figure 4.** Right FFA Contd

There was no orbitocranial bruits, no lymphadenopathy. The oropharynx was normal, the carotids were pulsatile, non tender and there was no radio-popliteal delay. The fundi were examined under mydriasis. The upper temporal retina appeared pale and ischemic due to nerve fibre layer edema in the right eye (Figure 1). This corresponds to the inferior altitudinal Right field defect (Figure 2).

However, the Fundus Fluorescein angiography results were surprising. The vessels which were expected to be occluded, filled in first. Further, the break up of the dye

appearance time in the early, mid and late venous phases was the earliest in the vessels involving the affected area. (Figure 3-Figure 8). This patient had a RIGHT Incomplete CRAO which could either be embolic or non embolic.

Looking at the clinical picture in our case the findings were attributed to embolus reorientation which occurred while the artery possibly underwent a spasm (Figure 9). It is known in FMD there is involvement of vasa vasorum and the arteries could undergo spasm [1].



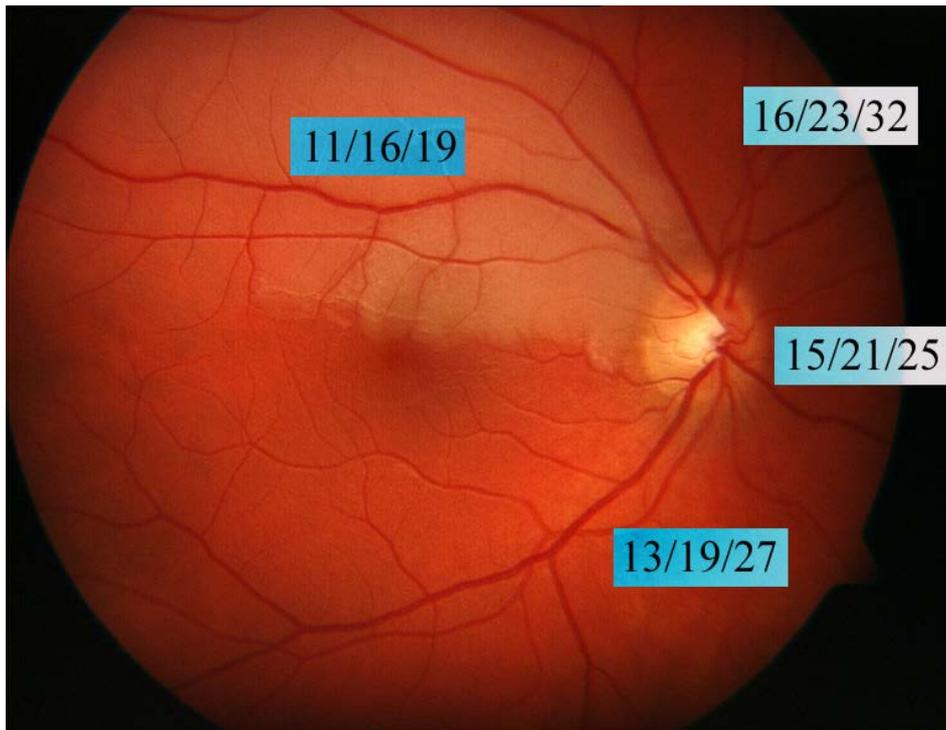
Figure 5. Right FFA Contd



Figure 6. Right FFA Contd



**Figure 7.** Right FFA Contd



**Figure 8.** Comparison of the dye appearance on FFA: Early/ Mid / Late venous

Magnetic resonance angiography of the carotids showed the typical “String of beads sign”, bilaterally.

Other investigations including Transthoracic and Transoesophageal Echocardiogram were normal.

#### 4. Discussion

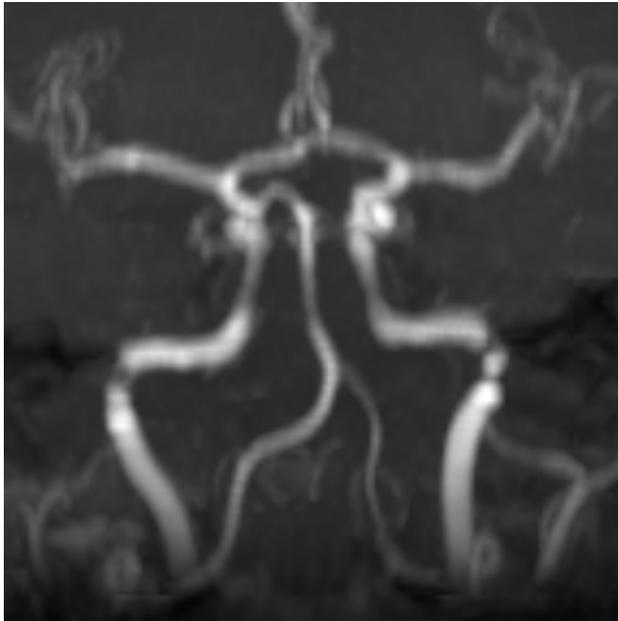
FMD is a nonatherosclerotic, noninflammatory vascular disease that primarily affects women in the age group 20 to 60 years, however men and older individuals may also be affected [4].

Commonly known to occur in the renal and carotid arteries, it has been found to involve almost every artery in the body. There are, however, data to suggest that FMD is much more common than previously thought. It is estimated that the incidence in the general population is 2% to 3% [5].

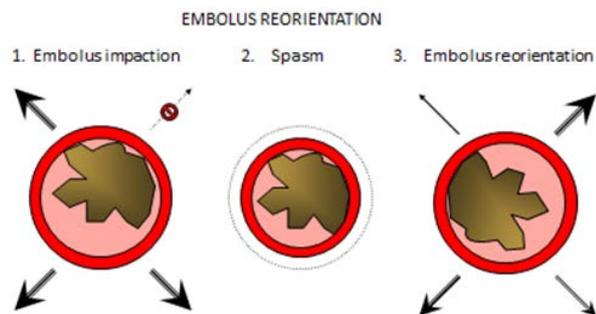
The etiology is multifactorial including vessel wall ischemia, smoking, estrogens, environmental as well as genetic factors. The female to male ratio is 9:1 [6,7].

Histologically FMD may involve the tunica intima, tunica media, and adventitia. Involvement of the media is further classified into medial fibroplasia,

perimedial fibroplasia, and medial hyperplasia. The commonest of these is Medial fibroplasia which occurs in 70% to 95% of cases. [8] It shows the classic “string-of-beads” appearance on angiography. Adventitial (periarterial) fibroplasia (<5 %) is the least common characterized by replacement of adventitial fibrous tissue by dense collagen [9].



**Figure 9.** Magnetic Resonance Angiography showing bilateral ‘String of Beads’ sign



**Figure 10.** Embolus reorientation

Embolus impaction produces retinal artery occlusion which causes a secondary phenomenon of obstruction of vaso vasorum to arteriole and that medial ischemia of the arteriole results in spasm. The spasm physically alters the orientation of embolus producing the re-establishment of flow (evident on FFA) but too late for re-establishment of visual function.

The authors searched PubMed without date or language restriction in Sept 2014, using the following combinations of terms: Central retinal artery occlusion AND Fibromuscular dysplasia.

Arterial complications include stenosis, dilatation, thrombosis and dissection. Stenosis and dilatation can lead to occlusion and aneurysm respectively. In turn each can be an embolic source.

CRAO as a complication of FMD is very rare. Only four cases of CRAO in FMD, (and one case of cilioretinal artery occlusion) have been reported in the literature [10,11,12,13,14] however none of them had embolus reorientation.

## References

- [1] Fievez ML. Fibromuscular dysplasia of arteries: a spastic phenomenon? *Med Hypotheses*. 1984 Mar; 13(3):341-9.
- [2] Leadbetter WF, Burkland CE. Hypertension in unilateral renal disease. *J Urol* 1938; 39: 611-26.
- [3] Slovit, D.P. and Olin, J.W. Fibromuscular dysplasia. *N Engl J Med*. 2004; 350: 1862-1871.
- [4] Pascual A, Bush HS, Copley JB. Renal fibromuscular dysplasia in elderly persons. *Am J Kidney Dis* 2005; 45: e63-6.
- [5] Olin JW, Froehlich J, Gu X, et al. The United States Registry for Fibromuscular Dysplasia: Results in the First 447 Patients. *Circulation*. 2012; 125: 3182-3190.
- [6] Stanley JC, Fry WJ. Renovascular hypertension secondary to arterial fibrodysplasia in adults: criteria for operation and results of surgical therapy. *Arch Surg* 1975; 110: 922-8.
- [7] Olin JW, Pierce M. Contemporary management of fibromuscular dysplasia *Curr Opin Cardiol*. 2008 Nov; 23(6): 527-36.
- [8] Kolluri R, Ansel G. Fibromuscular dysplasia of bilateral brachial arteries--a case report and literature review. *Angiology* 2004; 55(6): 685-9.
- [9] Persu A, Touzé E, Mousseaux E, et al. Diagnosis and management of fibromuscular dysplasia: an expert consensus. *Eur J Clin Invest* 2012; 42: 338-47.
- [10] Altun A, Altun G, Olcaysu OO et al. Central retinal artery occlusion in association with fibromuscular dysplasia. *Clin Ophthalmology* 2013; 7: 2253-5.
- [11] Matonti F, Prost Magnin O, Galland F et al. Internal carotid artery dissection on arterial fibromuscular dysplasia causing a central retinal artery occlusion: a case report. [Article in French]. *J Fr Ophtalmol*. 2006 Sep; 29(7):e15.
- [12] Sawada T, Harino S, Ikeda T. Central retinal artery occlusion in a patient with fibromuscular dysplasia. *Retina* 2004 Jun; 24(3): 461-4.
- [13] Choi JH, Jung J, Park KP et al. Intracranial fibromuscular dysplasia presenting as various ocular manifestations. 2014 Feb 15; 337(1-2): 232-4.
- [14] Warrasak S, Tapaneya-Olarn W, Euswas A et al. Fibromuscular dysplasia: a rare cause of cilioretinal artery occlusion in childhood. *Ophthalmology*. 2000 Apr; 107(4):737-41.