

Fibromuscular Dysplasia: One unit's experience

Dr Aine De Bhailis¹, Dr Constantina Chrysochou¹, Ms Emma Flanagan¹, Dr Edward Lake², Dr Amy Hudson¹, Prof Philip A Kalra¹

¹Salford Royal Foundation Trust, Salford, United Kingdom, ²Manchester Royal Infirmary, Manchester, United Kingdom

Introduction

Fibromuscular dysplasia (FMD) is a non-atherosclerotic, non-inflammatory disease of medium arteries that has been described in every vascular bed with a wide variety of manifestations such as beading, stenosis, occlusion, aneurysm or dissection.

The United States Registry and European Registry for FMD have added to our knowledge of this rare disease, however questions remain. The RaDaR (National registry of rare kidney diseases) FMD in the unit aims to recruit UK based patients to a national registry. The vision is that this database will align with the European registry.

Our unit has the only dedicated clinic in the UK which aims to effectively diagnose, screen and treat those with suspected FMD. Here we describe the experience of a dedicated FMD clinic the first of its type in the United Kingdom.

Methods

Screening of patient records from January 2010 to October 2019 was undertaken for diagnosis of fibromuscular dysplasia, coronary artery dissection, middle aortic syndrome along with the acronyms "SCAD" and "FMD".

The diagnosis had to be confirmed in the vascular bed by angiography. FMD was diagnosed as non-atherosclerotic arterial stenosis affecting the trunk or branch of medium sized vessels, in the absence of aortic wall thickening, inflammation and known syndromic arterial disease

The diagnosis of SCAD was made based on coronary angiographic characterised by a tear in the arterial wall that is nontraumatic and non-iatrogenic with no secondary aetiology of the dissection

Clinical data collected included:

- Clinical presentations leading to diagnosis (i)Renal such as accelerated phase hypertension or poorly controlled hypertension (ii) Cerebral/Neck such as migraine, cerebrovascular events or tinnitus and (iii) Cardiac such as acute coronary syndrome
- Sex, age, positive family history, smoking history and number of anti-hypertensive agents
- Secondary prevention agents
- Intervention
- Formal screening

Results

From January 2010 to September 2019 36 patients were identified using the pre-specified search terms. 9 patients were excluded leaving 27 patients.

The median age was 50 with a 2:1 ratio for females to males.

Most patients with FMD (17/20) presented with a renal manifestation i.e. hypertension.

Renal artery FMD was confirmed by CTA/MRA in 20 patients. 60% had extra-renal screening. 11 patients had cerebral screening (8 CTA and 6 MRAs). 7 patients had aortic imaging (4 CTAs, 4 MRAs). 50% had head to toe screening as recommended by international guidelines.

50% of those with an FMD diagnosis were on a statin and 45% were on an anti-platelet agent.

In relation to those with an initial diagnosis of SCAD (5 patients), 3 patients had screening in terms of CTA/MRA and 1 patient was found to have cerebral vessel manifestations consistent with FMD.

Conclusion

A dedicated clinic for FMD allows for prompt diagnosis and appropriate screening for possible involvement of other vascular beds. It also allows appropriate treatment with secondary prevention agents such as aspirin and statins to those who are most likely to benefit. Although in its infancy there is great potential for this bespoke clinic to continue to expand to become a national centre of excellence.