

A case of Coronary Fibromuscular Dysplasia

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Introduction: Fibromuscular dysplasia (FMD) is a rare non-atherosclerotic non-inflammatory vasculopathy often involving medium-sized vessels. It is well-known to present with non-atherosclerotic stenosis of renal arteries and hence hypertension. Involvement of other arteries, including the carotid arteries, illiac arteries and visceral arteries have also been reported. Fatality due to involvement of heart vasculature has also been described in both literature and local coroner cases.

Recently, there was a case involving major coronary artery leading to coronary occlusion and death.

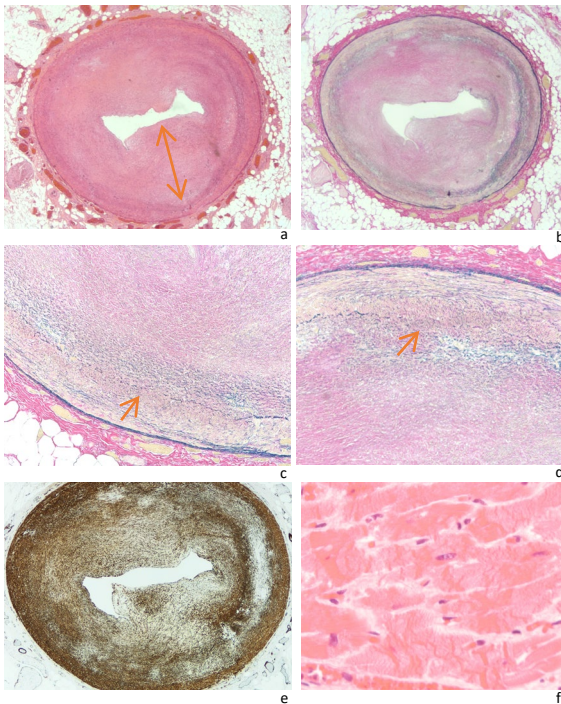
Case:

The case was a 37-year-old male with good past health and unremarkable family history who was found collapsed at home. No prior complaint of discomfort was noted. Autopsy showed significant non-atheromatous occlusion of the anterior descending branch of left coronary artery. Heart size and configuration were unremarkable. No recent or old myocardial infarcts were noted grossly. Other organs were normal. Toxicology examination was unremarkable.

Histology:

Staining with Hematoxylin and Eosin (H&E) stain, Periodic acid–Schiff (PAS) stain, Elastin van Gieson (EvG) stain and Immunohistochemical (IHC) staining for actin were performed.

H&E and PAS staining both showed significant occlusion of the artery without atheromatous or hyaline changes. No sign of inflammation. EvG staining showed pronounced intimal proliferation with collagen and disruption of the internal elastic lamina. IHC staining with actin showed muscle infiltration in the intimal layer as well.



(a)= H&E (b-d) = EvG (e): Actin; (f) H&E of the myocardium of the corresponding territory
 (a) Marked intimal thickening; absence of atheromatous plaque or cholesterol cleft
 (b-d) Disruption of the internal elastic lamina;
 Red = collagen; Blue = elastic fiber; Yellow = muscle & others
 (e) Patchy muscular infiltration in the intima, interspersed by abundant collagen as identified in the EvG
 (f) = contraction band necrosis of the myocardium of the corresponding coronary territory, consistent with early myocardial ischaemia.

Overall findings were compatible with being intimal type of fibromuscular dysplasia.

Other sudden death cases in the Public Mortuary:

Materials and methods:

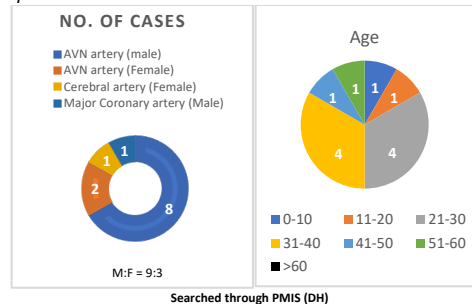
Patient Data: Local cases with cause of death given as fibromuscular dysplasia (FMD) were collected from the Public Mortuary Information System (PMIS) of the Department of Health (DH) from 2005 to 2022.

Results:

Cases: A total of twelve cases of FMD were found in the PMIS. 10 of them listed FMD as the direct cause of death (I), with 8 of them involving the atrioventricular nodal artery (AVN artery). 1 case was intracerebral haemorrhage (ICH) involving intracerebral artery. The other case is the present case of the author, involving the major coronary artery.

Concerning the two cases listing FMD as a significant condition contributing to the death (II), both of them involved the AVN artery. One ascribed the direct cause of death (I) as “Adverse effects of methamphetamine” with “Gastroenteritis” being another significant condition (II). Another case listed “Acute on chronic renal failure” due to “Adverse effects of Ketamine” as the direct cause of death (I).

Demographics:



Clinical symptoms:

Preceding symptoms	No. of cases (%)
Chest pain	2 (16.7%)
Shortness of breath	1 (8.3%)
Vomiting/Diarrhoea/Abdominal discomfort	2 (16.7%)
Headache	1 (ICH case) (8.3%)
Fever / flu-like symptoms	3 (25%)
No known prior complaint	4 (33.3%)
Circumstances at the time of collapse	
While exertion	1 (8.3%)
At rest/in bed	5 (41.7%)
Not known / not witnessed	6 (50%)
Other causes of death	
Anomalous origin of LCA (listed as II)	1 (8.3%)
Gastroenteritis (listed as II)	1 (8.3%)
Drugs (Meth, Ketamine) (listed as I)	2 (16.7%)

Conclusions:

This is the first reported fatal case due to FMD of major coronary artery in our centre. Both clinicians and pathologists should be aware of the possibility of non-atherosclerotic coronary occlusion and the potential fatal outcome.

References:

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