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Acute Coronary Syndrome with Av Malformation – A Rare Presentation

KEYWORDS: AV malformation, coronary artery disease, coronary angiogram

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ABSTRACT *Vascular malformations of the heart are very. Rare Only a few type arteriovenous of malformations were reported. Cardiac Arterio venous malformation (AVM) , which are intramural conglomerates of malformed arteries and veins fed by one of the coronary arteries have been reported incidentally. Its incidence is about 0.002% of the general population.*

We report a case of 62 yr old male, smoker who presented to the emergency department with tightness of chest and shortness of breath since 10days .He was a known hypertensive and on regular medication since 6yrs .His echo showed no regional wall motion abnormality with degenerative aortic valve disease and sever concentric left ventricular hypertrophy .Then coronary angiogram was done which showed Coronary artery disease(CAD), two vessel disease with multiple AVMs and normal left ventricular function. He was advised for coil embolization and revascularization. Coil embolization was done and patient is under follow up.

INTRODUCTION

Arterio venous malformation (AVM) is an abnormal connection between arteries and veins, and are termed arterio-arterial or arterio-venous when the malformation is between two arteries or an artery and a vein respectively. These connections can be congenital, occurring in variety of sizes and different locations .The symptoms range from asymptomatic to myocardial necrosis, bacterial endocarditis and heart failure depending on the size of the AVM. We present a case of CAD with multiple AVM.

CASE REPORT

A 62 yr old male smoker presented with complaints of tightness of chest associated with shortness of breath for 10days. He also complained of increase in severity of breathlessness even after walking for a short distance. It was not associated with any pain radiation to the jaw; arm or back .There is no vomiting or sweating. There is no history of similar complaints in the past. He is a known hypertensive and on regular medication since 6yrs .There is no past history diabetes mellitus or family history of CAD. Pt is a chronic smoker since 10yrs, 5-6 cigarettes per day.

Clinical examination showed afebrile PR -70/min, BP – 160/90 mmHg, RR – 20/ min, JVP not raised, CVS examination showed heaving type of apex at 6th ics 1.25cm lateral to midclavicular line. Systolic murmur heard in the mitral area and an ejection systolic murmur in the aortic area. Respiratory examinations revealed bilateral vesicular breath sound and no added sound. Other system examination revealed normal He was advised for ECG and it showed LVH, no ischemic changes.

Echocardiography showed no regional wall motion abnormality with degenerative aortic valve disease, mild calcific AS, severe concentric left ventricular hypertrophy, dilated left atrium, and normal left ventricular systolic function .Troponin-T was negative. Patient diagnosed as chronic stable angina class iii, advised for coronary angiogram.CAG revealed mid LAD 70% stenosis with large AVM arising from septal branch. Large AVM arising from proximal LCX mid RCA 80% stenosis with large AVM arising from conus branch and a AVM arising from PLVB

At this stage the diagnosis was CAD with DVD, multiple AVMs.

Patient was advised for coil embolization and revascularization.

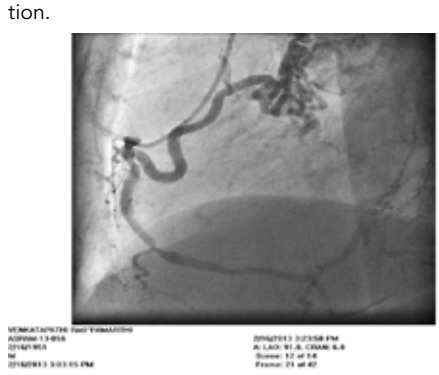

Fig 1 shows AVM from conus with mid RCA 80% stenosis

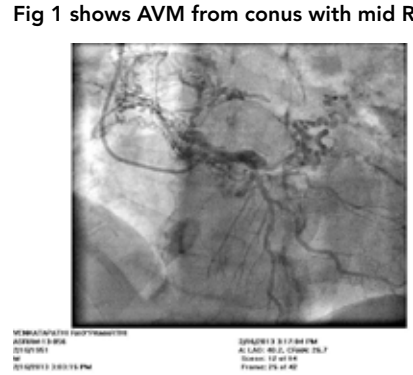

Fig.2 shows AVM from septal & proximal LCX

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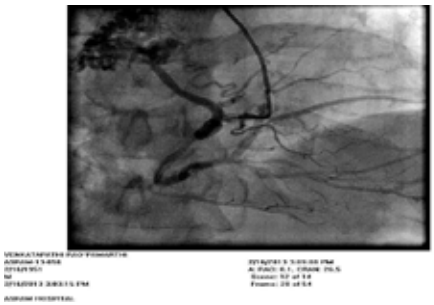

Fig.3 shows AVM from conus and PLVB

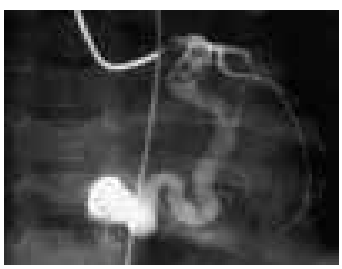

Fig.4 shows after coil embolisation

DISCUSSION

Coronary Arteriovenous Malformation (AVM) consists of abnormal communication between coronary artery and one of the cardiac chambers or vessels adjacent to the heart . Coronary Arteriovenous Malformation is a very rare anomaly present in 1 in 50,000 live births or 0.002% of the general population and is visualized in nearly 0.25% of patients undergoing catheterization Coronary AVM may be congenital or acquired.(1) It constitutes nearly half of all coronary artery anomalies and is the most common cause of hemodynamic ally significant coronary lesions. In 20–45% of the cases, Coronary AVMs are associated with some other congenital heart disease including atrial septal defect, tetralogy of fallot, patent ductus arteriosus, ventricular septal defect or pulmonary atresia(2) It is present as an isolated finding in 55–80% of the cases It was firstly described by Krause in 1865.Most of the patients with coronary AVMs are older than 20 years. Approximately half of all patients with AVM remain asymptomatic and some coronary AVMs might disappear spontaneously during childhood. Symptoms and complications may develop with increasing age, and when surgery is performed in later life mortality and morbidity is increased .Coronary AVM can arise from any of the coronary arteries. In the literature, the most common site reported is right coronary artery or its branches (55%), left anterior descending artery (35%) and circumflex coronary artery been rarely involved Single origin is the most common form of coronary AVM, ranging from 74% to 90% while multiple malformations have been reported in 10.7% to 16% of the cases. Over 90% of the fistulas drain into the venous structures of circulation which includes right ventricle (40%), right atrium (26%), pulmonary artery (17%), coronary sinus(7%), and superior vena cava (1%). Less frequently i.e. about 3–5 % of the AVMs drain into the left sided cardiac chambers With increasing age, drainage into the main pulmonary artery is a relatively common occurrence(3) .

Coronary AVMs cause shunting of blood and clinical presentation depends on the size of the malformation and thus degree of shunting . Coronary artery dilatation is reported but has not seen to be associated with the shunt size. In case of a low resistance malformation, the fistulous tract shunts significant amount of blood and causes a reversal of arterial flow in the segment distal to the AVM, resulting in a parasitic circulation, which causes decreased arterial pressures in the distal capillary beds and can cause tissue ischemia . About one half of the patients with AVM are asymptomatic. However, these malformations may result in severe complications and the patients may present with pulmonary hypertension because of an existing large left to right shunt, congestive heart failure, subacute bacterial endocarditis, myocardial ischemia resulting from steal phenomena, rupture or thrombosis of the CAVM or associating arterial aneurysm (4). Symptoms and risk of these lethal complications increase with age.

The diagnosis of coronary AVM is challenging due to its low prevalence, yet it should be considered in many symptomatic or asymptomatic patients presenting with cardiac murmurs. Differential diagnosis includes patent ductus arteriosus, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, prolapse of the right aortic cusp with a supracristal ventricular septal defect, internal mammary artery to pulmonary artery fistula, and systemic arteriovenous fistula. Invasive angiography remains the gold standard for diagnosis of coronary AVMs. However, the relation of coronary fistulas to other cardiac structures is at times unclear, and their origin and course can be limited due to overlap of adjacent structures. Most of the fistulas are small and found incidentally during coronary angiography. Non-invasive diagnostic modalities including transthoracic echocardiography combined with doppler and color flow imaging, transoesophageal echocardiography, magnetic resonance imaging and contrast enhanced multislice tomography can be used as an adjunct to coronary angiography .

Although the natural history of the coronary AVM is variable and spontaneous closure is reported in some cases, it is recommended by most of the authors to treat symptomatic coronary Arteriovenous Malformations. The treatment of asymptomatic coronary AVM is still controversial with some authors recommending closure of coronary AVM in these patients to prevent fistula related complications that has been seen to increase with age. Currently there has been no consensus in surgical vs. medical treatment of patients with coronary AVMs. Surgical closure by epicardial and endocardial ligations are gold standard for treatment and remains safe and effective with good reported success. Successful surgical occlusion of these malformations without cardiopulmonary bypass has been reported. Ligation of the coronary AVM may be performed on the outside of the heart without CPB bypass when there is a simple and easily accessible coronary AVM. The use of Percutaneous closure technique needs several conditions: anatomy of the fistula should be favorable for this treatment (eg. Non-tortuous vessel, the fistula should be unique with distal narrowing to avoid embolism to the drainage site, and distal portion of the fistula should be accessible with the closure device (5). Transcatheter closure devices have been successful.

CONCLUSION

Coronary AVM is a rare congenital disease and about half of the patients with the condition are asymptomatic. For patients who present with symptoms such as angina, dyspnea and arrhythmias, a coronary AVM should be suspected. Diagnosis of the condition can be easily and non-invasively made by cardiac CT angiography. Transthoracic echocardiography and magnetic resonance imaging are other diagnostic modalities which can be used for AVM evaluation and to calculate the shunt fraction. Shunt size, location, course and insertion site can be easily depicted, allowing for surgical or percutaneous procedural planning. Closure of the coronary AVM is recommended for symptomatic patients which can be done percutaneously, if suitable anatomy and especially if patient needs percutaneous coronary intervention for a co-existing coronary pathology. Surgical closure can now be performed even without cardiopulmonary bypass in a beating heart.

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