A Clinical and Angiographic Profile and Assessment of Outcomes in Coronary Artery Ectasias: Single Centre Experience

Rashmi Kerr¹, Cinosh Mathew², Ram Gopal Shahi³, Rajneesh Calton⁴

Abstract

Objective: To assess the clinical and angiographic profile of patients with coronary artery ectasias (CAE) and assess their outcomes.

Method: One year retrospective and one year prospective study was carried out in the Department of Cardiology in CMC, Ludhiana from January 2011 to December 2012 on all patients undergoing coronary angiographies and each patient was followed up for 1 year. Their outcomes were noted and statistical analysis carried out.

Results: There were a total of 327 males (66.3%) and 166 females (33.7%) in the study. Mean age of patients were 51 to 60. The incidence of CAE was 79 (16.02%). 6.9% patients had pure ectasia without CAD. The distribution of CAE according to Markis classification was, Type 1 in 21 (26.5%), Type 2 in18 (22.7%) and Type 3 in 28 (35.44%) and Type 4 comprised of 12 (15.18%). Among all 3 categories UA was the commonest presentation (p=0.004). Hypertension was the commonest risk factor in both patients with CAE (44 patients, 55.7%) and CAD (167 patients, 52.6%) but not statistically significant. Regarding outcomes among Type 1, 84% patients remained asymptomatic, while 8.8% of patients were symptomatic with UA episodes. 4% were lost to follow up. Among Type 2, 68.8% remained asymptomatic while 24.4% had recurrence of symptoms. 6.6% were lost to follow up. Among the Type 3, 70.12% of patients remained asymptomatic while 20.12 % had recurrence of symptoms, 8.17% were lost to follow up. Five patients (1.57%) expired, all of whom were from Type 3. Most patients improved with the treatment opted by them over a 1 year period however the patients that remained symptomatic on treatment were three (8.8%) in category 1, 12 (24.4%) in category II and 55 (20.12%) in category III. Four in Category II required repeated hospitalization while 16 in category III required rehospitalizations.

Conclusion: CAE is not an uncommon finding among patients presenting with acute coronary syndromes requiring invasive evaluation to confirm disease severity and decide management. However it is a benign entity requiring optimal medical management.

Introduction

Coronary artery ectasia (CAE) is defined as the dilatation of the coronary artery of 1.5 times the reference range for that vessel.¹,²

Editorial Viewpoint

• A three year retrospective study for presence of coronary artery ectasias (CAE).
• CAE was founding 16% of patients.
• Unstable angina was commonest presentation and hypertension was commonest risk factor.
• A slow flow with underlying CAD could be a potential risk factor for catastrophic coronary event.

They represent the direct result of inappropriate coronary dilatation. Angiographic signs of turbulent and stagnant flow include delayed antegrade flow, segmental backflow and local deposition of dye in the dilated coronary segment.³,⁴ It is graded according to Markis into four grades based on the extent of ectasia in decreasing order of severity. Type 1 is diffuse ectasia of two or three vessels, Type 2 is diffuse disease in one vessel and localized disease in one vessel, Type 3 is diffuse ectasia in one vessel while Type 4 is localized or segmental ectasia.³ This major group of angiographic findings in the absence of flow limiting stenosis could present...
with significant coronary events and lead to morbidity among these patients. It has been underreported and undertreated and could lead to recurrent hospitalizations.

Various studies have found the incidence of CAE between 1%-16%. The maximum reported incidence of CAE by Waly et al in an Egyptian study in 1997 was 16.4% of all patients undergoing coronary angiograms. In various Indian studies the incidence has been reported to vary between 2%-10%.  

Various modalities have been used to identify CAE however coronary angiography is the gold standard for detection of cases. The other methods utilized include intravascular ultrasound (IVUS), which is an excellent tool for assessing luminal size and characterizing arterial wall changes. CAE commonly affects isolated vessels, however all three vessels are also affected in a substantial proportion. Of interest is the diffuse fusiform type of CAE that tends to have more frequently bilateral distribution and association with abdominal aortic aneurysms. It usually coexists with underlying coronary artery stenosis and in patients where it occurs in the absence of stenotic lesions it tends to be diffuse.

The clinical sequelae and prognosis needs special attention and despite absence of flow limiting stenosis, Kruger et al convincingly has documented the presence of angina, positive exercise stress test and pacing induced myocardial ischemia in patients with ‘dilated coronaropathy’. Along with this unstable angina syndromes may occur despite the absence of stenosis.

The exact etiopathogenesis of this abnormal luminal dilatation is unknown however various hypothetical causes have been implicated. The commonest ones being coronary atherosclerosis, extensive medial degeneration and hyalinization possibly as a result of chronic vascular inflammation.  

Another probable mechanism causing this is over stimulation of the endothelium by Nitrous Oxide (NO) and NO donors. Yet another mechanism proposed is the role of matrix metalloproteinases (MMP) which are actively involved with proteolysis of extracellular matrix proteins.

This study was done to identify the incidence of CAE among all patients undergoing coronary angiographies, look at their presentations, the risk factors associated with the development of it, their detailed angiographic findings, and their outcome.

**Method**

A prospective and retrospective study of one year including all patients undergoing coronary angiographies in a tertiary level centre in North India in Punjab from 1st January 2011 to 31st December 2012.

A detailed protocol was filled for each patient which included their name, address, phone number, age sex, hospital number and date of admission. The presenting complaints were noted and risk factors with duration recorded. Their presenting blood pressures, pulse rate and ECG’s were noted. A detailed 2 dimensional echocardiography was also done.

The laboratory parameters, cardiac enzymes (CPK, CPK-MB, TROP-T) were recorded on admission and after 24 hrs, the baseline lipid profile and fasting and post prandial sugars were recorded. The treatment administered to the patient on admission was noted.

All patients underwent conventional coronary angiograms on the GE ADVANTX LC+ cath lab and SEIMENS ARTIS ZEE cath lab till 31st December 2012. All angiograms were done via the femoral approach using Judkins left and right catheters except in special situations where the Amplatz catheters, multipurpose catheters, and LIMA catheter were used. All conventional views were recorded, (RAO caudal, LAO cranial, LAO caudal, plain LAO, lateral, AP cranial, plain AP, AP caudal, RAO cranial, plain RAO, and a detailed description of the angiography was noted. They were classified into single vessel disease (SVD) double vessel disease (DVD) and triple vessel disease (TVD) based on the number of vessels affected. Further they were classified according to the vessel affected and the segments with the severity of disease noted according to the segments. Presence of calcification and thrombus was noted.

**Definition of Coronary Artery Ectasia**

All patients having coronary artery ectasia as defined by a dilatation of 1.5 times the reference range for that vessel with or without presence of delayed clearance of dye with sluggish blood flow were classified according to Markis into four types based on the extent of ectasias in decreasing order of severity as described earlier.

All patients were further sub classified into three categories

Category 1: CAE alone (Figure 1).

Category 2: CAE with angiographically significant stenotic lesion (Figure 2).

Category 3: No CAE, only angiographically significant coronary artery disease.

**Fig. 1: CAE (Category I)**
The management based on angiographic details was also noted and the treatment followed by the patient in the form of revascularization via percutaneous transluminal coronary angioplasty (PTCA) or coronary artery bypass surgery (CABG) or medical management was also recorded. In case of PTCA the stent used with its size was also noted, and similarly in case of CABG the graft details were noted.

Follow up was done on OPD basis at intervals of 1, 3, 6, 9 and 12 months and a record of symptoms, signs, lab parameters and ECG with change in left ventricular function, interventions and results of intervention was also noted. The final outcome at one year was also recorded.

**Results**

There were a total of 327 males (66.3%) and 166 females (33.7%) in the study. Most of the patients were in the 51 to 60 years age group followed by the 61 to 70 years and 41 to 50 year age group. The mean age group was 55.75 years with a minimum age of 30 years and a maximum age of 73 years. 397 patients (80.52%) had findings on their coronary angiographies while 96 patients (19.48%) had angiographically normal coronaries. The incidence of CAE was 79 patients (16.02%). Patients who had pure ectasia comprised of 21 patients (26.5%). Type 2 comprised of 18 patients (22.7%). Type 3 consisted of 28 patients (35.44%) and Type 4 comprised of 12 patients (15.18%). Majority of the patients had Markis Type 3 ectasias confined to one vessel (Figure 3). Category I comprising of the pure ectasia group, consisted of 34 patients (8.5%) of whom 21 (67.64%) were males and 13 (32.36%) were females. Category II comprising of patients with angiographically significant coronary artery disease with coronary artery ectasias consisted of a total of 45 patients (11.3%) of whom 33 (73.3%) were males and 12 (26.7%) were females. Category III comprised of patients with pure stenotic artery disease. There were a total of 318 patients (80.2%) in this category of which 216 (68.3%) were males and 102 (31.7%) were females (Table 2).

The distribution of coronary artery ectasias according to Markis classification was:

- **Type 1** comprised of 21 patients (26.5%).
- **Type 2** comprised of 18 patients (22.7%).
- **Type 3** consisted of 28 patients (35.44%) and
- **Type 4** comprised of 12 patients (15.18%).

Majority of the patients had Markis Type 3 ectasias confined to one vessel (Figure 3). Category I comprising of the pure ectasia group, consisted of 34 patients (8.5%) of whom 21 (67.64%) were males and 13 (32.36%) were females. Category II comprising of patients with angiographically significant coronary artery disease with coronary artery ectasias consisted of a total of 45 patients (11.3%) of whom 33 (73.3%) were males and 12 (26.7%) were females. Category III comprised of patients with pure stenotic artery disease. There were a total of 318 patients (80.2%) in this category of which 216 (68.3%) were males and 102 (31.7%) were females (Table 2).

Among category I, LAD was affected in 24%, LCx in 8%, RCA in 4% while in category II, RCA

**Table 1: Angiographic findings in the study group**

<table>
<thead>
<tr>
<th>Frequency (%)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal angiography</td>
<td>96 (19.5)</td>
</tr>
<tr>
<td>Isolated CAE</td>
<td>34 (6.9)</td>
</tr>
<tr>
<td>CAE + CAD</td>
<td>45 (9.1)</td>
</tr>
<tr>
<td>CAD patients</td>
<td>318 (64.5)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>493</td>
</tr>
</tbody>
</table>

**Table 2: Category-wise distribution of patients**

<table>
<thead>
<tr>
<th>Category I (n=25)</th>
<th>Category II (n=54)</th>
<th>Category III (n=309)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>Female</td>
<td>Total</td>
</tr>
<tr>
<td>-------</td>
<td>--------</td>
<td>-------</td>
</tr>
<tr>
<td>Number</td>
<td>21</td>
<td>13</td>
</tr>
<tr>
<td>% dist</td>
<td>67.64</td>
<td>32.36</td>
</tr>
<tr>
<td>Markis 1</td>
<td>8.5%</td>
<td>11.3%</td>
</tr>
</tbody>
</table>

**Fig. 2: CAE with obstructive CAD (Category II)**

**Fig. 3: Distribution of coronary artery ectasias according to Markis grading**
was affected in 29.6% followed by LCx in 18.5% and in LAD (16.7%). All three vessels were affected in 36% in category I and 16.7% in category II which was also found statistically significant with a p value of 0.01. The incidence of CAE affecting isolated RCA in category II as compared to category I was also statistically significant with a p value of <0.0001 (Table 3).

Of the 413 patients that were taken into the study, in the CAE and the non CAE group the commonest presentation was chest pain (84.8% and 74.9%), followed by breathlessness (31.6% and 22.4%) and palpitations (10.1% and 12.6%) (Figure 4).

Among category I, 30 patients (88.2%) presented with UA which was the commonest presentation in this category, followed by Non STEMI in 2 patients (5.9%), acute LVF in 2 patients (5.9%) while none presented with STEMI. Among category II, most patients presented with UA, 19 patients (42.2%), 16 patients 35.5% with STEMI’s, 9 patients (20%) with non STEMI and 1 patient (2.3%) with acute LVF. Among category III, the commonest presentation was UA in 143 patients (44.9%), STEMI in 93 patients (29.25%), Non STEMI in 63 patients (19.8%) and acute LVF in 19 patients (5.97%). In all 3 categories UA was the commonest presentation and it was found to be statistically significant (p=0.004).

Hypertension was the commonest risk factor in both patients with CAE (44 patients 55.7%) and CAD (167 patients 52.6%). Diabetes was present in 104 patients (32.8%) of patients who had CAD, and in 17 patients (21.5%) in patients with CAE + CAD. Smoking was present in 70 patients (22.1%) with CAD but in only 10 patients (13.7%) in patients with CAE. These risk factors were found not to be statistically significant.

In this study, 26 patients (32.9%) with CAE had LV dysfunction while 53 patients (67.1%) had a normal LV function. Among the non CAE group, 151 patients (47.4%) had LV dysfunction whereas 167 patients (52.6%) had normal LV function. This was found to be statistically significant (p value of 0.0001).

In our study we found that pure CAE in the absence of any flow limiting stenosis was seen in 34 patients (8.5%) while 20 patients (5.03%) had non critical CAD. The distribution of single vessel disease (SVD) was 122 (30.73%). Double vessel disease (DVD) was seen in 100 patients (25.18%) and triple vessel disease (TVD) in 121 patients (30.47%).

Among category I, all patients (n=34) were advised medical management, while among category II, 9 patients (25.9%) were advised medical treatment while 44 patients (44.4%) opted for surgical management, 31 patients (57.4%) were advised PTCA while 30 patients (56.6%) opted for it, and 16.7% were advised CABG while none opted for it. Among category III, 49 patients (15.9%) were advised medical treatment while 191 patients (61.8%) opted for it, 146 patients (47.2%) were advised PTCA while 76 patients (23.3%) opted for it, and 114 patients (36.9%) were advised CABG while only 43 patients (13.9%) opted for it.

Regarding outcomes among category I, 84% patients remained asymptomatic, while 8.8% of patients were symptomatic with UA episodes. 4% were lost to follow up. Among category II, 68.8% remained asymptomatic while 24.4% had recurrence of symptoms. 6.6% were lost to follow up. Among the category III, 70.12% of patients remained asymptomatic.
Table 4: Outcome of patients at the end of 1 year

<table>
<thead>
<tr>
<th>Category I n=34</th>
<th>Category II n=45</th>
<th>Category III n=318</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic</td>
<td>3 (8.8%)</td>
<td>12 (24.4%)</td>
</tr>
<tr>
<td>Good / asymptomatic</td>
<td>30 (88.2%)</td>
<td>30 (68.8%)</td>
</tr>
<tr>
<td>Lost to follow up</td>
<td>1 (2.9%)</td>
<td>3 (6.6%)</td>
</tr>
<tr>
<td>Expired</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

*p value 0.4821
while 20.12 % had recurrence of symptoms, 8.17% were lost to follow up, 5 patients (1.57%) expired, all of whom were from category III (Table 4).

Most patients improved with the treatment opted by them over a 1 year period however the patients that remained symptomatic on treatment were 3 (8.8%) in category I, 12 (24.4%) in category II and 55 (20.12%) in category III. 4 of them in Category II required repeated hospitalization while 16 in category III required rehospitalizations.

Patients with dilatation with delayed clearance of dye in their coronaries was seen in 45 patients (43.3%) while aneurismatic dilatation with delayed clearance of dye was seen in 34 patients (32.7%) and slow flow was seen in 25 patients (24%). The commonest angiographic finding was dilatation with delayed clearance of dye (Table 5). Chest pain was the commonest presentation seen in 22 patients (88%), breathlessness in 7 patients (28%) and palpitations in 3 patients (12%). There was a statistically significant correlation with the presence of chest pain and the presence of slow flow in the coronaries (p value of 0.001). The commonest clinical manifestations was that of unstable angina (52%) followed by STEMI (28%), followed by NSTEMI (16%) followed by LVF in 4%. A statistical significance exists with the clinical presentation of unstable angina in patients with slow flow in their coronaries with a p value of <0.001. All the patients with slow flow that presented with STEMIS were in Category II (with coexisting obstructive coronary artery disease). This indicates that presence of slow flow with underlying CAD was an additional risk factor in the development of myocardial infarctions. None of the patients with pure stenotic lesions in absence of slow flow developed myocardial infarctions. On analyzing presence of risk factors, dyslipidaemia was seen in 11 patients (44%), hypertension in 9 patients (36%), diabetes in 7 patients (28%) and smoking in 5 patients (20%). Dyslipidemia was the commonest risk factor found in patients with slow flow however was not found statistically significant. There were 4 patients (16%) who remained symptomatic, 20 patients (80%) who were asymptomatic and 1 patient (4%) who was lost to follow up.

Discussion

A total of 16.2% patients were found to have CAE in the study group which is the highest seen in all Indian studies conducted to date. Of these, 45 patients (56.96%) i.e. more than half had associated coronary artery disease indicating the presence of concomitant atherosclerotic disease which has been implicated in the etiopathogenesis of CAE. According to Markis classification Markis type 3 with isolated single vessel involvement was the commonest manifestation among patients in the study. The vessels affected by CAE was isolated LAD involvement, seen in 15 patients (18.9%), isolated LCx in 11 patients (13.9%), isolated RCA in 14 patients (17.7%), LAD +LCx in 12 patients (15.1%) LAD+RCA in 2 patients (2.5%), RCA and LCx in 4 patients (5.06%). All three vessel involvement was seen in 21 patients (26.5%). The

The commonest vessels affected by ectasia in category I patients was all three vessels together (LAD + LCx + RCA) seen in 13 patients (38.23%) while in Category II the commonest involvement was isolated RCA seen in 13 patients (28.8%). This was found statistically significant with a p value of <0.001. In most of the other studies mentioned in the review, a majority had RCA involvement but the reason for this association has not been established.

On studying angiographic details in the CAE group it was observed that 34 patients (43.03%) had aneurysmal dilatation with delayed clearance of dye while 45 patients (56.97%) had pure dilatation with delayed clearance of dye.

The commonest presenting complaint was that of chest pain in 67 patients (84.8%) followed by breathlessness in 25 patients (31.6%) and palpitations in 8 patients (10.1%). The commonest presentation in the CAE group was that of unstable angina in 49 patients (62.02%) followed by STEMI in 16 patients (20.25%), NSTEMI in 11 patients (13.92%) and acute LVF in 3 patients (3.79%). This was found statistically significant with a p value of <0.004 indicating the association of CAE presenting with unstable angina episodes. The common risk factors were studied to look for association between hypertension, diabetes, smoking and dyslipidaemia in the development of CAE, however no statistical correlation could be ascertained. Hence further analysis needs to be done to identify risk factors that might have led to its development in such a large subset in this part of the country.
All patients in category I with pure ectasia were advised optimal medical management and remained asymptomatic on follow while only 3 cases (8.8%) remained symptomatic but never required repeat hospitalizations during the study period. The ones that remained symptomatic were the ones with aneurysmal dilatation of the ectatic segments indicating that it is a more severe form of ectasia as compared to the patients with dilatation in the absence of aneurysmal formation indicating a more severe form of the disease requiring more stringent follow up and medical management. Hence it can be concluded that CAE is not an uncommon angiographic finding in this geographic area and in fact has a high prevalence of 16.2%. These patients present with significant coronary events that require hospitalizations. However the prognosis of these patients is good and only a few remain asymptomatic on follow up not necessitating readmissions. The ones that remained symptomatic were the ones with aneurysmal dilatation of the ectatic segments indicating it was a more severe form of the disease requiring more stringent follow up and medical management. Further studies need to be done to identify reasons for a high prevalence of CAE in this geographic area and identify risk factors involved in its development.

**Angiographic Alterations in Flow Characteristics**

A total of 104 patients were found to have flow alterations, slow flow was seen in 25 patients (24.0%), aneurysmal dilatation with delayed clearance of dye was seen in 34 patients (32.7%) and dilatation with delayed clearance of dye was seen in 45 patients (43.3%).

Pure slow flow in the absence of CAD was seen in 16 patients (64%) while slow flow in the presence of CAD was seen in 9 patients (36%). The commonest presentation in patients with slow flow was that of chest pain in 22 patients (88%), breathlessness in 7 patients (28%) and palpitations in 3 patients (12%). The presentation with chest pain and presence of slow flow in the coronaries was statistically significant with a p value of 0.001.

This group also presented with acute coronary syndromes like unstable angina in 13 patients (52%), NSTEMI in 4 patients (16%) STEMI in 7 cases (28%) and acute LVF in 1 patient (4%). The clinical presentation with unstable angina and presence of slow flow in the coronaries was found statistically significant with a p value of <0.001.

All the patients with slow flow that presented with STEMI were in Category II. This indicates that presence of slow flow with underlying CAD was an additional risk factor in the development of myocardial infarctions. None of the patients with pure stenotic lesions in absence of slow flow developed myocardial infarctions. Hence it can be concluded that presence of slow flow in the presence of underlying CAD is an additional risk factor in the development of catastrophic events.

On studying the risk factor profile in these patients an association of dyslipidaemia was seen among these cases 11 patients (44%) however this was not statistically significant and additional risk factors need to be looked into for the development of slow flow situations. On follow up 4 patients (16%) remained symptomatic while 20 patients (80%) remained symptom free on optimal medical management.

Hence it can be concluded that apart from looking at severity of stenosis during angiographies it is important to identify flow characteristics which also can be implicated in the development of acute coronary syndromes. Angiographic evidence of slow flow is also a significant angiographic finding and these patients also present with acute coronary syndromes even in the absence of flow limiting stenosis but have a favorable prognosis on optimal medical management. Also the presence of slow flow with underlying CAD is a risk factor in the development of catastrophic coronary events like myocardial infarctions. Hence this entity should be treated and not left alone to ensure streamlined blood flow in the coronaries.

**Limitations of the Study**

The study was conducted in a limited group (493 cases) needed a larger number of patients for further interpretation. This study was biased by the fact that it was a hospital based study and not a population based study, hence the numbers that have been generated through this study is not applicable to the general population. To identify the actual incidence of CAE even among asymptomatic patients a large population based study using noninvasive modalities eg CT coronary angiography may be required. This being a tertiary referral centre, gets complicated and referred cases hence the patient selection was not representative of the general population.

**Conclusion**

CAE is not an uncommon finding among patients presenting with ACS requiring invasive evaluation to confirm disease severity and decide management. Flow characteristics is another important finding which should be noted while performing angiographies since every effort should be made to streamline blood flow in the coronaries to avert catastrophic cardiovascular events.

**Future Directions**

To identify risk factors that were not looked at in the development of CAE eg racial, dietary, familial etc. We also need to follow up these patients with CAE and look at their long-term (>1 year, 5 years, 10 years) outcomes by conducting non-invasive studies eg. CT Coronary angiographies to
identify the changes occurring in the coronary anatomy after few years in patients with CAE.

References


