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Case Report

Internal Medicine Section

A Case Report on Spontaneous Coronary Artery Dissection in a Patient with Churg-Strauss Syndrome

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ABSTRACT

Spontaneous Coronary Artery Dissection (SCAD) is a rare but of one the important causes of sudden cardiac arrest and Acute Coronary Syndrome (ACS). It is complex and often under diagnosed. It has multifactorial aetiologies. It is predominantly seen in young women presenting with ACS, with no pre-existing history of Coronary Artery Disease (CAD) and is commonly diagnosed via coronary angiography. It may be associated with autoimmune disease, connective tissue disorder, collagen vascular disease, Marfan syndrome, intense physical exercise and during peripartum period. The various treatment modalities for SCAD are conservative medical treatment, Percutaneous Coronary Intervention (PCI), and surgery. Here, authors describe a case of 33-year-old women who presented with ACS, with no pre-existing history of CAD. Patient responded well with medical management. This condition has grave prognosis, if not detected and treated promptly. So it is very important to keep SCAD as differential diagnosis in ACS among young patients.

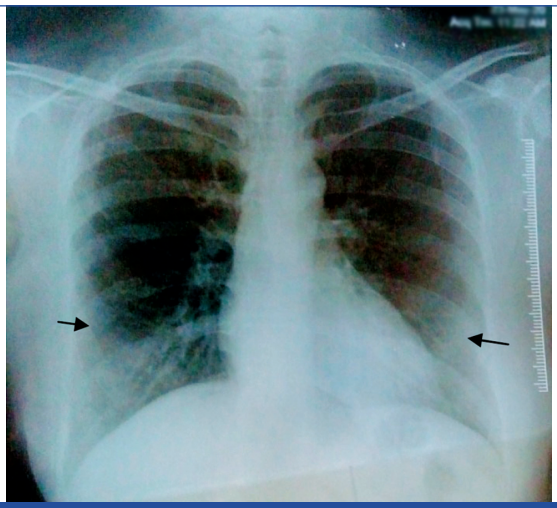
Keywords: Acute coronary syndrome, Autoimmune disease, Connective tissue disorder, Sudden cardiac arrest, Young women

CASE REPORT

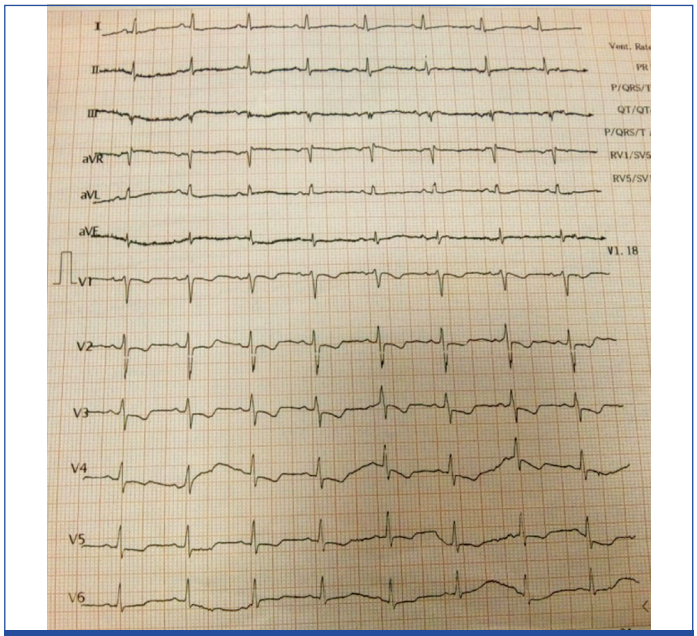
A 33-year-old female, known case of bronchial asthma, presented with polyarthralgia (small and large joints), weakness, fatigue and reduced appetite for 15 days. She also complained of retrosternal chest pain radiating to left arm since one day. There was history of abnormal behaviour of one day. Patient was known case of bronchial asthma, well controlled with Metered Dose Inhalers (MDI). She suffered from sinusitis. Her last child birth was two years ago. There was no history of diabetes, hypertension or dyslipidaemia.

On evaluation, following findings were observed, random blood sugar 92 mg/dL, Haemoglobin 12.1 gm%, Total Leukocyte Count (TLC) 7000/cumm, eosinophil 22%, hypereosinophilia Absolute Eosinophil Count (AEC) 1540, platelet count 1,90,000/µL, Vitamin D 13.8 ng/mL, Total bilirubin 0.7 mg/dL, Direct bilirubin 0.2 mg/dL, Serum Glutamic Oxaloacetic Transaminase (SGOT) 60 IU/L, Serum Glutamic Pyruvic Transaminase (SGPT) 62 IU/L, urea 23 mg/dL, creatinine 0.9 mg/dL, thyroid profile Thyroid Stimulating Hormone (TSH) 2.7, Triiodothyronine (T3) 1.38, Tetraiodothyronine (T4) 9.0 and International Normalised Ratio (INR) 1.2. Lipid profile showed total cholesterol 190 mg/dL, triglyceride 150 mg/dL, High Density Lipoprotein (HDL) 58 mg/dL, Low Density Lipoprotein (LDL) 120 mg/dL. Coronavirus Disease 2019 (COVID-19) Reverse Transcriptase Polymerase Chain Reaction (RT-PCR) test was negative. Among fever protocol, Peripheral Blood Smear (PBS) for Malarial Parasite (MP) was negative, dengue serology was negative, widal test and typhi dot was negative. Urine examination was normal but chest X-ray showed pulmonary infiltrates [Table/Fig-1].

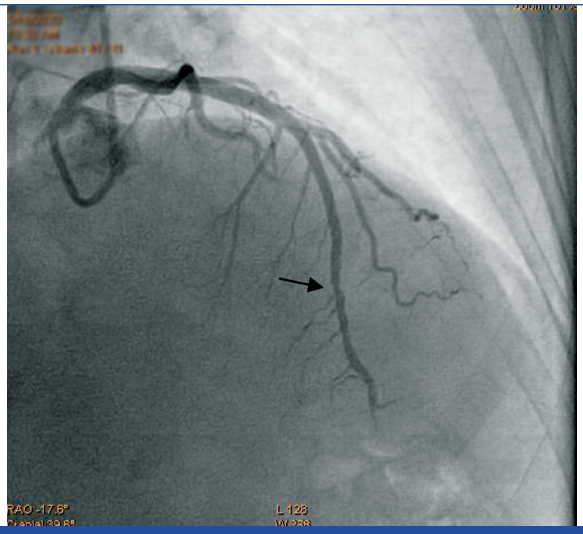
Electrocardiogram (ECG) [Table/Fig-2] showed Flat T Lead I, II, III, avL, avF and T Inversion V1-V6, Trop T was Positive, Creatine Kinase MB (CKMB) 18 IU/L. Echocardiogram demonstrated Left Ventricular Ejection Fraction (LVEF) 60%, no regional wall motion abnormality and valves were normal. Coronary Angiography (CAG) showed right dominant circulation, normal Left Main Coronary Artery (LMCA), Left Anterior Descending (LAD) Type 1 SCAD on LAD with TIMI (Thrombolysis in Myocardial Infarction) grade III flow at origin of D, with normal Left Circumflex and right Coronary Artery [Table/Fig-3,4].



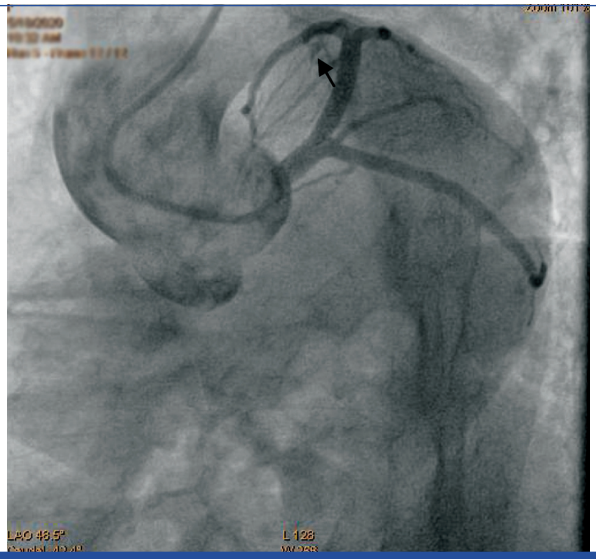
[Table/Fig-1]: Chest X-ray showed pulmonary infiltrates (Right>Left).



[Table/Fig-2]: ECG revealed Flat T Lead I, II, III, avL, avF and T Inversion V1-V6.



[Table/Fig-3]: CAG: Type 1 SCAD on LAD with TIMI-III flow at origin of D2.



[Table/Fig-4]: CAG: Right dominant circulation and Type 1 SCAD on LAD.

Ultrasound abdomen revealed left kidney hydronephrosis with thinning of cortex. Left adnexa shows anechoic cyst of size 45×35 mm with thin septa. Other organs were normal. Magnetic Resonance Imaging (MRI) Brain revealed multifocal altered signal intensity foci in bilateral frontoparieto occipital lobes, bilateral thalamoganglionic region and both cerebellum with many of the lesions showing restricted diffusion. Image findings were non specific, although possibility of encephalitis cannot be ruled out.

Considering polyarthralgia, Connective tissue work up was done and Rheumatoid Factor (RF) negative, C-reactive Protein (CRP) positive, Anti-cyclic Citrullinated Peptide antibodies (CCP) 5.9 U/mL, Antinuclear Antibodies (ANA) 0.42, Antiphospholipid Antibodies (APLA) IgM 4.9 U/mL, IgG 6.2 U/mL, Uric acid 4.1 mg/dL, Antineutrophilic cytoplasmic antibodies (ANCA) cANCA 4.12 U/mL and pANCA 3.2 U/mL. Extractable nuclear antigens (ENA) profile (Qualitative) was Negative. Serum autoimmune encephalitis panel was negative.

To diagnose Churg-Strauss syndrome, the American College of Rheumatology (ACR) proposed six criteria [1]: Asthma, peripheral blood with eosinophilis count more than 10% pulmonary infiltrates (transient), paranasal sinusitis, histopathology demonstrating vasculitis with presence of extravascular eosinophils and Mononeuritis multiplex or polyneuropathy. If four or more criteria are present, it yields a sensitivity of 85% and a specificity of 99.7%. In index case, patient met with five of above six criteria for the diagnosis of Churg-Strauss syndrome.

In view of TIMI-III flow, the patient was managed with Tab. Ecosprin 150 mg OD, Tab. Clopid 75 mg OD, Tab. Atorvastatin 40 mg HS, Tab. Metoprolol XL 25 mg and Glucocorticoids. Patient responded well with the treatment and improved. The patient was maintained on conservative medical management with close outpatient follow-up in view of COVID-19 pandemic. Unfortunately, the patient got infected with coronavirus during her monthly follow-up and later succumbed to COVID-19.

DISCUSSION

The SCAD is defined as an emergency condition that occurs when coronary artery wall is separated by haemorrhage with or without tear. It is a rare, complex and under diagnosed cause of myocardial infarction. It has been seen that in 1.7-4% cases SCAD leads to ACS [2]. Index patient developed acute myocardial infarction with no signs of atherosclerosis. SCAD is a rare underlying cause of ACS [3]. There are three types of SCAD. Type 1 has a double lumen under contrast staining of the arterial wall. Type 2 reveals diffuse (2-3 cm), smooth narrowing and can vary in severity (depending on intramural haematoma). Type 3 mimics atherosclerosis and appear as focal or tubular stenosis [4].

Motreff et al., described five specific angiographic features of SCAD: (1) no atheroma on other coronary arteries; (2) radiolucent flap(s); (3) arterial wal showing contrast dye staining; (4) starting and/or ending of the angiographic ambiguity on a side branch; and (5) smooth and linear narrowing of the lumen calibre, or stenosis of varying severity which can mimic a “stick insect” or “radish” [5]. SCAD is seen most commonly in young women that is, around 70% of the patients (female to male ratio- 2:1) and 30% of them occur in the peripartum period [6]. The most frequent site of dissection is LAD which accounts for 60% of coronary dissections. The second most common site is Right Coronary Artery (RCA) which is predominantly seen in males, followed by the left main artery [7].

The aetiology of SCAD is multifactorial and is divided into four groups [8]. First group consists of hereditary connective tissue disorders which are associated with a defective arterial wall (e.g. Marfan and Ehlers-Danlos syndromes). Second group includes underlying atherosclerotic changes which is most commonly seen in men at an age of average 55 years. The third group is related to peripartum period in females due to fluctuation in blood levels of oestrogen and relaxin, shear stress and increased blood flow [9]. The last group is idiopathic SCAD. Some other important causes of SCAD are chest trauma, severe exercise, consumption of certain medications like cocaine, cyclosporine, 5-fluorouracil, oral contraceptives, and fenfluramine [10].

The management of SCAD comprises of medical treatment, PCI, and surgery. Conservative medical management is done usually for patients with mild involvement. Surgery or PCI is indicated in patients with extensive dissections leading to persistent ischaemia. Conservative medical management of SCAD is similar to the ACS, and includes use of anticoagulants, aspirin, clopidogrel, beta blockers, nitrates, and sometimes calcium channel blockers [11].

The most important feature seen on histological examination is predominant eosinophilic infiltrate which is seen in almost 50% of dissections. The presence of eosinophil granule components in the vessel wall and their tissue damaging effects helps in explaining role of eosinophils in the dissection process. Eosinophilic granule comprise of enzymes like acid phosphatase, collagenase, major basic protein, arylsulfatase, and beta-glucoronidase. These enzymes damage the collagen fibers of the arterial wall leading to initiation of dissection process. In fact, SCAD has also been reported in cases of Churg-Strauss syndrome, and drug-induced eosinophilia [12,13].

Left main coronary artery or multivessel dissection is associated with bad outcome [14]. The main modality for diagnosing SCAD is coronary angiography. Common coronary angiographic findings detected are intimal flap, two separate communicating lumens, multiple dissecting lines, and coronary aneurysm communicating with the lumen. Association of LAD is more commonly seen in

females, whereas right coronary artery (RCA) inclusion is prevalent in males. Dissection of left main artery is rare [15].

The PCI requiring cases present a unique challenge due to difficulty in identifying the true arterial lumen and placement of a secure guidewire. Intravascular imaging modalities such as intravascular ultrasound (IVUS) or Optical Coherence Tomography (OCT) may be required for precise localisation of the site and extent of dissection and identify the true lumen. ChromaFlo, has been used for optimal therapeutic strategies during PCI. It shows blood flow by comparing sequential axial IVUS images and as blood flows, it interprets any differences in the position of echogenic blood particles [16].

A new promising technology OCT is emerging. OCT has high-resolution (10-20 μm) intraluminal images with ultra-structural details. Lim C et al., has mentioned OCT as a useful adjunct because of its small diameter as compared to IVUS probe [17]. In the present study, these modalities were not used as it could cause wire induced propagation of dissection and also because the patient was stable.

The 1994, Chapel Hill consensus conference on the classification of vasculitis did not modify the ACR criteria [1,18]. Renal involvement (35% vs 4%) and peripheral neuropathy (84% vs 65%) is more commonly seen in ANCA positive patients than those without ANCA. Furthermore, in the biopsy sample of ANCA patients vasculitis was more often observed than ANCA negative patients (79% vs 39%). Interestingly, it has been observed that patients without ANCA are more likely to develop fever (55% vs 30%) and cardiac disease (49% vs 12%) [18,19].

Management of SCAD is guided by the clinical symptoms, haemodynamic status of the patient, the extent and location of the dissection. In cases of distal dissection with preserved coronary flow conservative medical therapy is reasonable. Hence, through the present case authors would like to signify the importance of a neglected cause of acute myocardial ischaemia and sudden death i.e SCAD.

CONCLUSION(S)

The SCAD is a rare cause of coronary artery syndrome seen most commonly in young women with no previous history of cardiac disease. Clinical symptoms, electrocardiographic findings and laboratory investigations are often similar to AMI. Early diagnosis is important for the treatment. The most preferable modality of treatment of SCAD is conservative management.

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