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Case Report

WPW Syndrome type A in an Adult Male - A Unique Case Report: Fascinating three Decades Roller-Coaster Journey of WPW Syndrome type A and its Subsequent Resolution.

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Abstract:
Wolff-Parkinson-White syndrome is the most common form of ventricular preexcitation and affects 1-3 per 1,000 persons worldwide. Many patients remain asymptomatic throughout their lives, however, approximately half of the patients with Wolff-Parkinson-White syndrome experience symptoms secondary to tachyarrhythmias, such as paroxysmal supraventricular tachycardia, atrial fibrillation, atrial flutter, and rarely, ventricular fibrillation and sudden death. Patients with Wolff-Parkinson-White syndrome may present with a multitude of symptoms such as unexplained anxiety, palpitations, fatigue, light-headedness or dizziness, loss of consciousness, and shortness of breath. We report a peculiar case of three decades of fascinating journey of an adult male suffering from WPW syndrome Type A and its spontaneous resolution eventually.
Keywords: WPW Syndrome, spontaneous resolution of delta wave, three decades natural history of WPW.

Introduction

It was in 1930, when Louis Wolff, John Parkinson, and Paul D. White together described a set of eleven cases of an electrocardiographic pattern consisting of a "functional bundle branch" and a "short PR interval" in the healthy young people with paroxysms of tachycardia (Figure 1).[1]

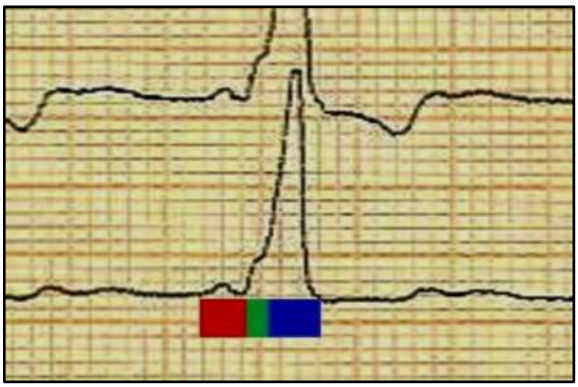


Figure 1: ECG of WPW Syndrome: 1) short PR - red, 2) delta wave - green, 3) rest of the QRS complex- blue

ECG features of WPW in sinus rhythm

- PR interval < 120ms
- Delta wave: slurring slow rise of initial portion of the QRS.
- QRS prolongation > 100ms
- Discordant ST-segment and T-wave changes (i.e. in the opposite direction to the major component of the QRS complex)
- Pseudo-infarction pattern in up to 70% of patients-due to negatively deflected delta waves in inferior/anterior leads ("Pseudo-Q waves"), or prominent R waves in V1-3 (mimicking posterior infarction)

Complementarily Surawicz et al [2] defined the ECG criteria of WPW syndrome. The pathognomonic ECG findings in WPW are the delta wave, characterized by a slurred upslope in the QRS complex and a short PR interval <120 ms (Figure 2) [2].

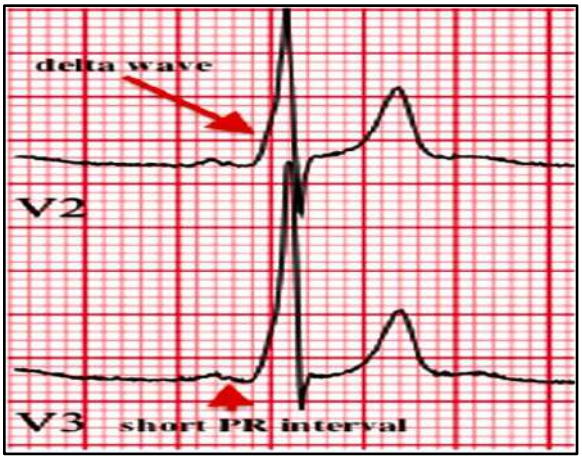


Figure 2: Characteristic ECG findings in WPW. There is presence of short PR interval (< 120 ms) and delta wave appearing as slurred upstroke of QRS complex.

The first successful surgical division of an aberrant atrioventricular (AV) connection for Wolff-Parkinson-White (WPW) syndrome was performed by Dr. Will C. Sealy on May 28, 1968. [3] As the surgical procedures for the cure of WPW syndrome were developed, the surgical anatomy of aberrant AV connections was defined. Sealy and associated led these efforts to elucidate the anatomy of WPW syndrome. On the basis of their elegant studies, the four regions where aberrant AV connections are found were designated as thee right free wall, left free wall, anterior septum and posterior septum. (Figure 3, 4)[4]

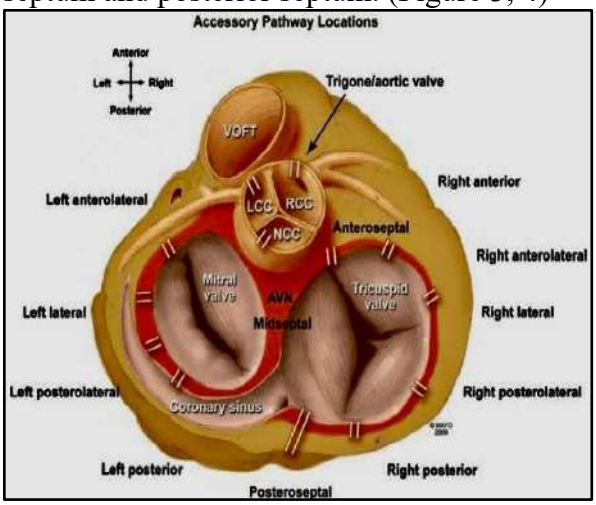


Figure 3: Location of Accessory pathways.

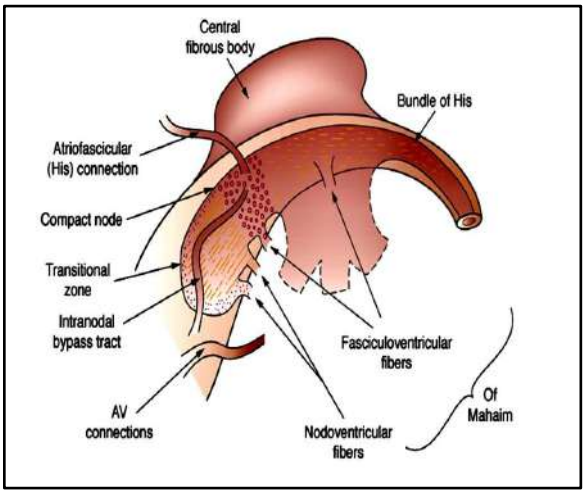


Figure 4: Schematic diagram of of three zones of the AV node and various types of perinodal and atrioventricular bypass tracts.

Wolff-Parkinson-White (WPW) syndrome is a distinctive pre-excitation disorder whose incidence varies from 0.1 to 3/1000 in the

healthy individuals and a prevalence of 0.1 to 0.3% in the general population. The occurrence is higher in males and gradually comes down as the age progresses due to loss of pre-excitation. The majority of the individuals with an extra pathway remain asymptomatic throughout their lives, and the risk associated with sudden cardiac death (SCD) is <0.6%. [5] WPW syndrome is defined as a congenital abnormality concerning the existence of abnormal conductive tissue between the atria and the ventricles in association with supraventricular tachycardia. It involves pre-excitation, which occurs due to conduction of an atrial impulse, not by way of the normal conduction system, but through an extra atrioventricular (AV) muscular connection [termed an accessory pathway (AP)] that bypasses the AV node. Patients with WPW syndrome may be asymptomatic or may present with palpitations, presyncope, syncope, or SCD. [6]

CASE PRESENTATION

Timeline

March 1996	First encounter – 38 years male presented with 2 years history of atypical chest pain with mild hypertension and multiple ECG's done during this period revealed WPW type A. There was no history of palpitations or syncope. Holter monitoring did not show any arrhythmia. TMT elucidated presence of delta wave at higher heart rates, without any evidence of inducible ischemia.
May 1996	Invasive Coronary angiography performed at a tertiary care medical institute in Lucknow revealed normal coronaries with presence of a short segment of myocardial bridge overlying the mid of left anterior descending (LAD) artery.
January 2014	64 slice CT coronary angiogram was organized at private diagnostic centre in Lucknow, which identified focal areas of mild stenosis in LAD and right coronary artery (RCA). Left circumflex artery (LCX) was normal. Additionally, a short segment of myocardial bridge was identified in the distal part of LAD.
November, 2017	Patient had an episode of syncope at 3.00 Am on 17-3-2017, while urinating in a standing position. ECG done in OPD was showing WPW Type A, similar to the previous ECG's. Holter was done and it did not detect any arrhythmia. Hence, the episode was considered to be a micturition syncope and the patient was reassured and advised suitably.
January 2022	Presented with recurrent episodes of palpitation for 2 days. Resting ECG dated 5-01-2022 disclosed the presence of supraventricular tachycardia (SVT) with a ventricular rate of 147/min. This was the first episode of SVT.
January 2022	Holter recording done over a period of 48 hours (from 05-01-2022 to 07-01-2022) unveiled multiple episodes of supraventricular

	tachycardia of prolonged duration varying from 3 hours 13 mins to 4 hours 6 minutes with a ventricular rate ~ 150 minute. Moreover, sinus beats exhibited prolonged PR interval and loss of delta wave, suggestive of spontaneous resolution of WPW syndrome.
January 6, 2022	Subsequent ECG's are showing prolonged PR interval with absence of delta wave, consistent with the diagnosis of resolution of WPW syndrome.
February, 2022	Electrophysiologic study (EPS) was done with the aim to locate and ablate the accessory pathway for permanently addressing the problem of recurrent SVT'S. However, EPS detected normal sinus node function, normal atrio-ventricular conduction with prolonged HV interval. Furthermore, no inducible supraventricular or ventricular tachycardia was found.
February, 2023	TMT was conducted on 22-02-2023 and notably there was presence of significant inducible ischemia although the patient did not complain of any ischemic chest pain. Duke Treadmill score was -2.95 suggestive of medium risk category. Currently, the patient is now 62 years of age, asymptomatic and doing well with medications for Type 2 Diabetes Mellitus, Hypertension, Hypothyroidism and Asymptomatic Coronary Artery Disease.
March 4, 2023	128 Slice CT Angiography was carried out and it revealed eccentric calcified plaque in proximal LAD causing moderate luminal narrowing. A small segment of myocardial bridge was noted over the distal LAD.

CASE REPORT

A 38 year healthy looking male presented to us in our OPD way back in March, 1996 (approximately 27 years back), with concerns of short episodes of atypical chest pain with mild nervousness and anxiety for past 15-16 months. He also described experiencing a "sharp chest pain" that did not radiate and resolved spontaneous within a couple of minutes. These symptoms caused him to stop his work and resulted in interference with his daily activities. He denied history of shortness of breath, presyncope or syncope, orthopnea, paroxysmal nocturnal dyspnea, weight gain or pedal edema. There was no family history of sudden cardiac death or cardiac arrhythmia. The patient gave history of hypertension (controlled on medication), however, he was non-diabetic and not suffering from a dyslipidemia or thyroid abnormalities. He was smoking 5-6 cigarettes per day, although he denied tobacco chewing, alcohol or substance abuse.

On clinical examination patient was haemodynamically stable. His pulse rate was 70/min. regular and BP was 100/70 mmHg in the right hand, in the sitting posture. Cardiovascular examination was not significant, there was no jugular venous distension, S1 and S2 were normal with no murmurs heard over precordium. His multiple ECG'S done elsewhere and the one in December, 1994 (Figure 5), illustrate short PR interval, delta wave on the ascending limb of positive QRS complex in the standard and precordial leads which is consistent with WPW Syndrome Type A.

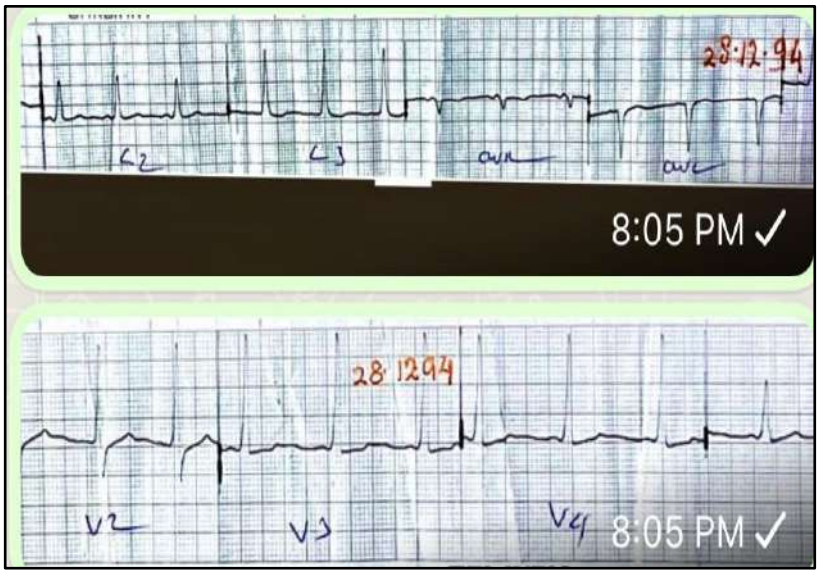


Figure 5: ECG suggestive of WPW syndrome Type A - short PR interval, slurred upstroke of QRS complex indicative of delta wave in leads 2, 3, AVF, V1- V6. Type A WPW is described as upright positive delta wave in all precordial leads V1-V6.

Our ECG done at the time of presentation in March, 1996 (Figure 6) was exhibiting similar features as that of the previous one.

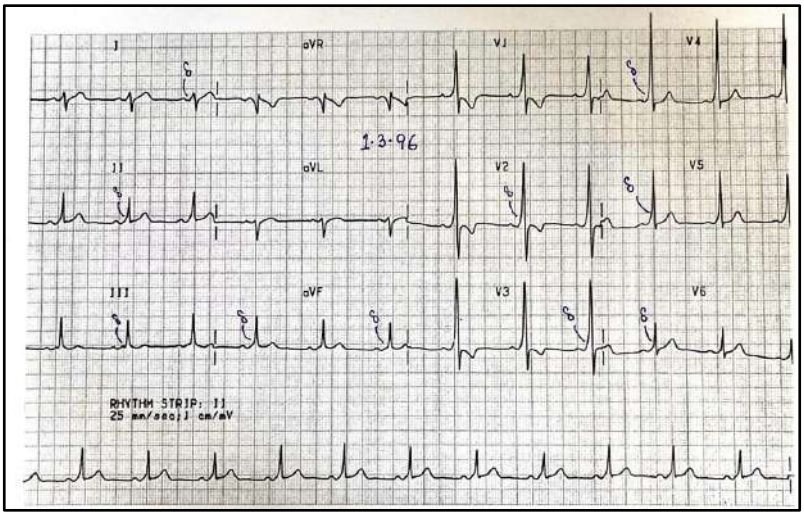


Figure 6: ECG dt 1.3...96 suggestive of WPW Type A.

Laboratory Investigation, which included complete blood count to rule out anemia, complete metabolic profile to rule out electrolyte abnormalities, lipid and thyroid profile to assess the presence or absence of dyslipidemia or thyroid abnormalities were all within normal range. Trop T test to rule out myocardial injury was normal. An exhaustive transthoracic color echocardiography was performed which revealed a left ventricular ejection fraction of 63% with normal biventricular systolic function and dimension. No structural heart disease could be identified. TMT demonstrated persistence of delta wave and WPW syndrome at higher heart rates suggesting a high risk category, even though there was no evidence of inducible ischemia (Figure 7). Moreover, x-ray chest (PA) view and 24 hr holter test were carried out and were also normal.

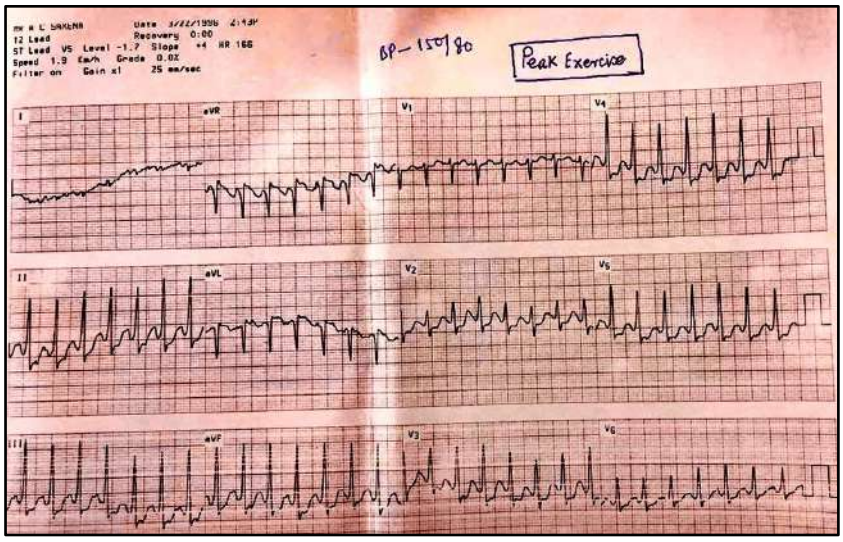
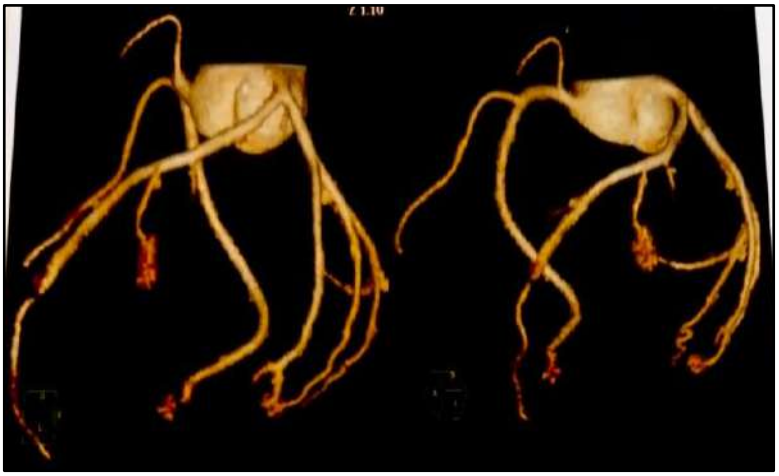


Figure 7: TMT done in 1996 identifies presence of delta wave at high heart rate of 166/ minute implying a high risk category.

Patient was advised invasive coronary angiography in view of his predominant symptoms of atypical chest pain. The angiogram displayed normal coronaries with presence of a short segment of myocardial bridge overlying the mid of LAD. For the long term management of WPW the patient was counseled to contact as soon as possible whenever, he had any symptoms of palpitations, presyncope, syncope, or anything usual. He was also recommended a 6 monthly check up with a fresh ECG, to look for any progression or regression of WPW syndrome. The patient was extremely co-operative and followed the instructions diligently. During the asymptomatic phase from 1996 to 2014, several ECG's were done routinely and all appeared similar: short PR, delta wave and a wide QRS. Nonetheless the patient again started complaining of frequent short episodes of agonizing atypical chest pain in January 2014 for which a 64 slice CT coronary angiogram was undertaken at a private diagnostic centre in Lucknow, which identified focal areas of mild stenosis in LAD and right coronary artery (RCA) (Figure 7a). Left circumflex artery (LCX) was normal. Additionally, a short segment of myocardial bridge was identified in the distal part of LAD. The patient was reassured about the mildness of the disease and advised accordingly.



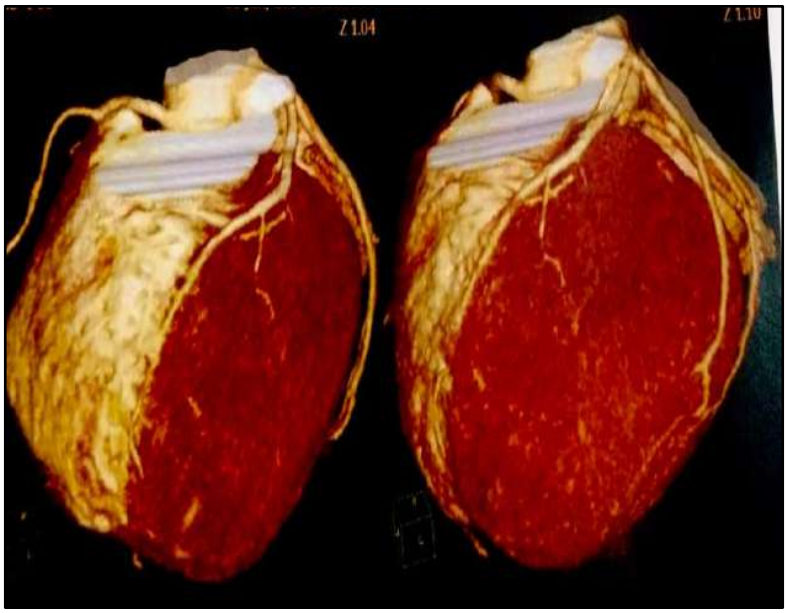


Figure 7a: Photograph of 64 slice CT Angiography (2014), depicting focal areas of mild stenosis in LAD and RCA. LCX was normal.

Again after a symptom-free interval of 8 years from 1994 to 2022 patient presented with recurrent episodes of palpitation for last 24-48 hours in January 2022. There was no history of syncope during these episodes. Resting ECG dated 5-1-2022 (Figure 8) disclosed the presence of supraventricular tachycardia (SVT) with a ventricular rate of 147/min.

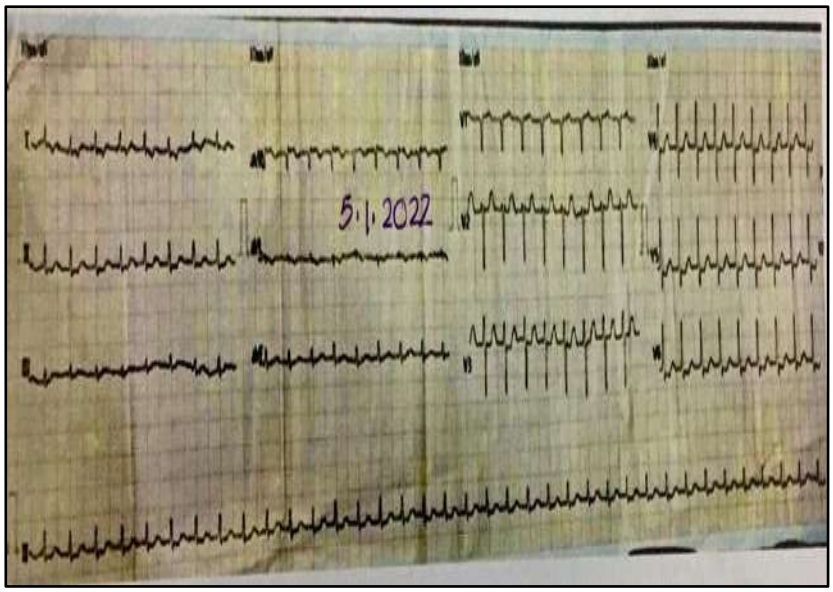


Figure 8: ECG dt 5.1.2022 is exhibiting Supraventricular Tachycardia with a Ventricular rate of ~ 147/ min.

Holter recoding done over a period of 48 hours from 5-1-2022 to 7-1-2022 unveiled multiple episodes of SVT of prolonged duration varying from 3 hour 13 minutes to 4 hours 6 minutes with a ventricular date of ~ 150/min. (Figure 9).

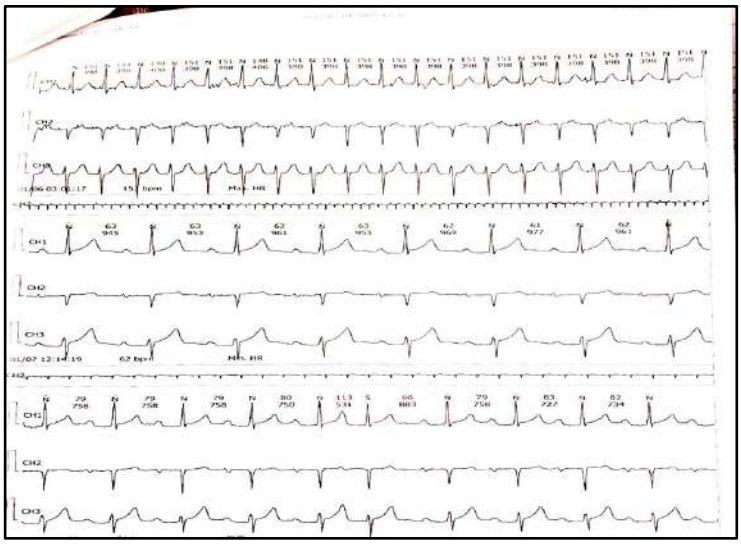


Figure 9: Holter tracing depicting the presence of Supraventricular Tachycardia with a Ventricular rate ~ 150/ min.

Subsequently the ECG done on 06-2-2023 identified a conspicuous ECG finding: absence of delta wave and the PR interval was prolonged (PR 263 m/sec) with a normal sinus rhythm (Figure 10). Absence of delta wave may be suggestive of resolution of WPW syndrome.

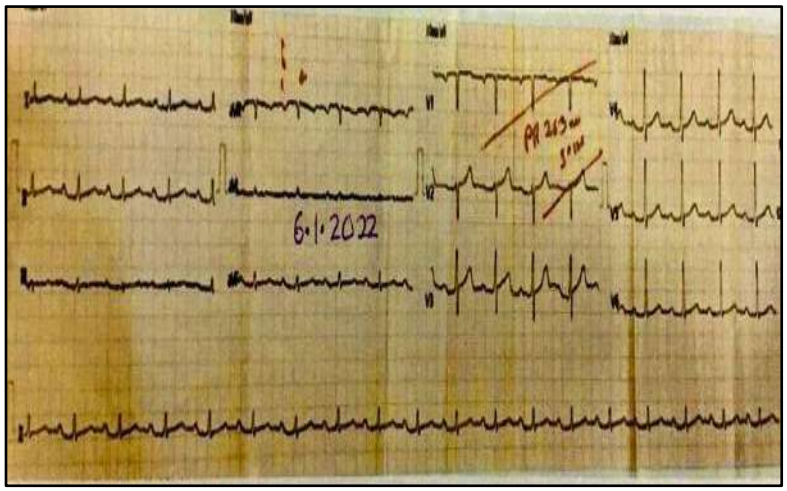


Figure 10: Resting ECG done after Subsidence of Supraventricular Tachycardia shows normal sinus rhythm with absence of delta wave and prolongation of PR interval (PR 263 msec)

Thereafter, Electrophysiologic study (EPS) was accomplished on 26-2-2022 with the aim to locate the accessory pathway, and ablate it by radio-frequency ablation (RFA). Notwithstanding, to our surprise EPS detected normal sinus node function, normal atrio-ventricular conduction with prolonged HV interval. Furthermore, no inducible supraventricular or ventricular tachycardia was found.

Henceforth, TMT was conducted on 22-02-2023 and notably there was presence of significant inducible ischemia (Figure 11) although the patient did not complain of any ischemic chest pain.

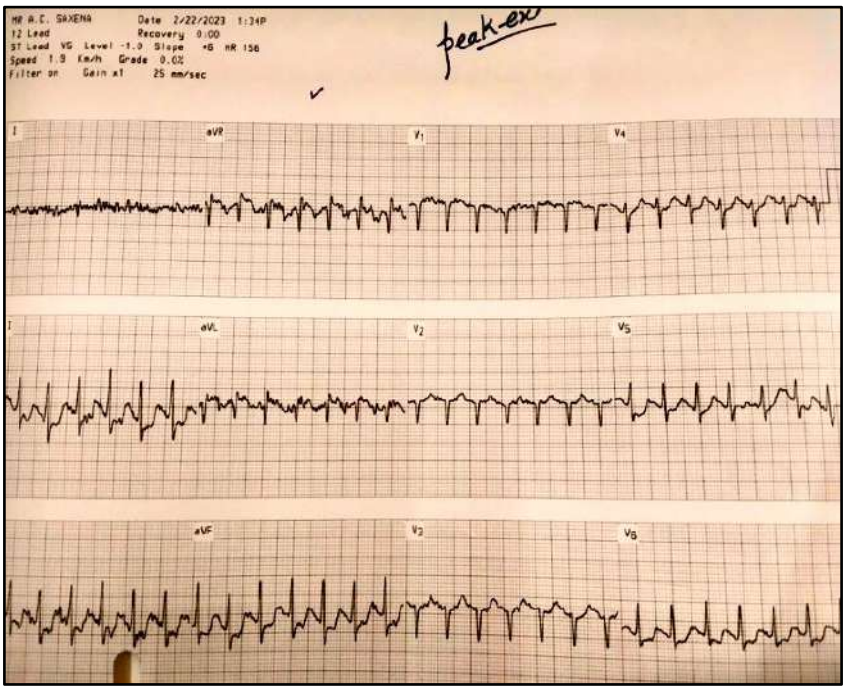


Figure 11: TMT dt 22.2.23 was remarkable and discerned presence of significant inducible ischemia with horizontal ST downsloping depression in L2, L3, AVF, V4-V6 and ST elevation in AVR.

Duke Treadmill score was -2.95 suggestive of medium risk category. Currently, the patient is now 62 years of age (Figure 12), asymptomatic and doing well with medications for Type 2 Diabetes Mellitus, Hypertension, Hypothyroidism and Asymptomatic Coronary Artery Disease.

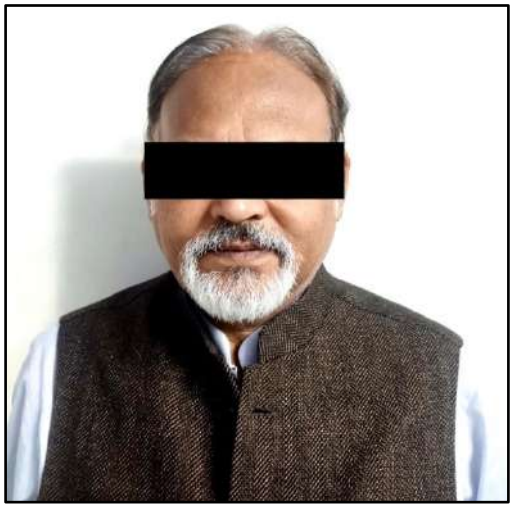


Figure 12: Current photograph of the patient

128 Slice CT Angiography carried out on March 4. 2023 (Figure 13) is reflecting eccentric calcified plaque in the proximal LAD, causing moderate luminal narrowing (approximately 50-60%). A small segment of myocardial bridge was noted over distal LAD.

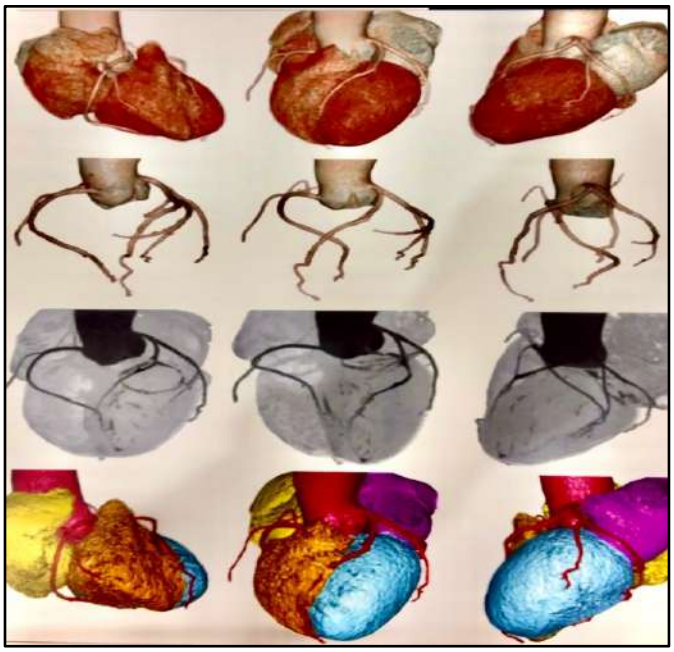


Figure 13: Current photograph (March 4,2023) of 128 slice CT Angiography illustrating eccentric calcified plaque in proximal LAD causing moderate luminal narrowing (50-60 %).

DISCUSSION

WPW syndrome is present in 0.1-0.5% of the total population. [7, 8] Its prevalence is higher in young children because the pattern of the syndrome can disappear spontaneously before 12 years of age, [9] mainly when the accessory pathway refractory period is long. [10]

When the refractory period is short, the spontaneous changes are rare and there is a stability of findings in children over 12 years of age and in adults with inducible supraventricular tachycardia. [11] The prevalence of WPW syndrome has been determined in young adults, [8] and its real prevalence in elderly subjects is unknown because these patients frequently have other heart diseases.

In WPW syndrome, the electrical impulse travels through the normal pathway via the AV node and also through an accessory pathway called the bundle of Kent. This accessory conduction pathway bypasses the AV node, allowing the electrical impulse to travel rapidly from the atria to the ventricles, causing ventricular preexcitation. [12] The conduction of impulse through both pathways, through the AV node and through the accessory pathway, causes rapid ventricular depolarization, resulting in tachycardia. [13]

The accessory pathway between the atria and ventricles develops from an aberrant embryonic evolvement of myocardial tissue connecting the fibrous tissues that divide the two chambers. [14] During early cardiogenesis, there is normally a direct connection between atrial and ventricular myocardium, which disappears with the in growth of the annulus fibrosis. Abnormalities in the annulus fibrosis allow this direct AV continuity to persist and provide anatomic structure for abnormal electrical conduction, causing ventricular preexcitation. [15]

The prognosis for infants with WPW syndrome is excellent. Spontaneous resolution was noted in 60-90 percent of

infants with WPW syndrome by one year of age without surgical treatment. [16]

While many of these patients are asymptomatic, WPW may also manifest as syncope, near-syncope, palpitations, supraventricular tachycardia (SVT), atrial fibrillation (AFib), or even sudden cardiac death (SCD). [17] Catheter ablation of the accessory pathway is considered curative, and is an excellent procedure for those with symptoms or are at risk of SCD and are of appropriate age. [18, 19] Our adult male patient has been afflicted with WPW syndrome for last three decades. Majority of this period was asymptomatic, although intermittently, there have been few instances of atypical chest pain, anxiety and palpitations due to documented SVT.

Spontaneous resolution of manifest WPW syndrome, defined as disappearance of the delta wave on electrocardiography, occurred in 14.6% of the entire study group. [20] A total of 133 patients (30%) presented at ≤ 3 months of age. Of these, 47 (35%) had spontaneous resolution of their WPW syndrome. Of the patients presenting at >3 months of age, 18 (5.7%) had spontaneous resolution, the oldest at 20 years of age. Of the 116 asymptomatic patients, 14 (12%) had spontaneous resolution of their WPW syndrome. [20] Intermittent loss of pre-excitation via ambulatory monitoring may occur in as many as 67% of asymptomatic cases. [21]

Abrupt and complete normalization of the PR interval with loss of delta wave during exercise testing or following procainamide, pro-pafenone, or disopyramide administration has been considered a noninvasive marker of low risk. [17, 22]

In our patient loss of delta wave was identified just after the subsidence of recurrent episodes of prolonged SVT'S. Conspicuously, this was the first bout of documented SVT'S and it occurred in the third decade of his suffering from WPW syndrome. Spontaneous resolution of WPW syndrome was further ascertained by the EPS study done few weeks later which discovered normal sinus node-function, normal atrio-ventricular conduction and prolonged HV interval, No inducible supraventricular or ventricular tachycardia was documented.

To the best of our knowledge we could not encounter any similar case study even after exhaustive review of the literature and hence the current case report is unique because of protracted roller-coaster course of WPW syndrome and its spontaneous resolution beyond 60 years of age.

CONCLUSION

In a country like India prevalence of WPW in still not much reported, the reason is lack of knowledge and awareness among the people. Early diagnosis, patient education, and correct treatment are of primary significance in patients with WPW syndrome. Asymptomatic cases need periodic observation and follow-up. For those with frequent episodes of symptomatic tachyarrhythmias, therapy should be initiated in the form of pharmacological therapy and catheter ablation.[23] Although the incidence of SCD in WPW is 1 in l00 symptomatic cases, early diagnosis and treatment are definitely associated with an excellent prognosis.[24]

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