24 years old asymptomatic, nonsmoker naval aviator reported for routine medical evaluation. There was no history of syncope, palpitations or shortness of breath. The general and systemic examination was normal and so were all the hematological and the biochemical parameters. A routine ECG was done as a part of evaluation protocol.

Questions

a) What does the ECG shows?
b) What is your differential diagnosis?
c) What is the significance of the findings?
d) What are the aeromedical considerations?
e) What is the final diagnosis?
Answers

a) The ECG shows

i) rSr pattern in the right precordial lead (lead V1) with normal QRS duration.

ii) Elevation of J point, concave upward ST segment elevation seen in leads V2 & V3 and relatively tall, symmetrical precordial T waves.

b) The differential diagnosis considered in this individual includes

i) The rSr pattern in lead V1 represents partial right bundle branch block (RBBB)

ii) The ST elevation seen in the leads V2 and V3 may represent either Early repolarization syndrome or Brugada Syndrome (ST-segment elevation in association with RBBB).

c) The significance of the finding depends on the final diagnosis.

i) Diagnosis of RBBB does not merit any investigation apart from an echocardiography for the structural integrity and a stress test for assessment of reversible ischemia. RBBB has been reported to occur in 0.2-0.4% of military aviators and 0.2-2% of civil population. The present available evidence does not suggest any increase in the risk of progressive conduction abnormalities or increased susceptibility to cardiac disorders in this subset of individuals with RBBB.

ii) Early repolarization syndrome is a common, normal ECG variant seen in 2-5% of normal population. It is characterized by minimally concave upwards ST segment elevation, relatively tall and frequently symmetrical T waves, prominent J waves, mid-precordial U waves and a rapid precordial transition. Although presumed to be benign there are a subset of individuals with early repolarization who are at increased risk of arrhythmia. There are at present no diagnostic method to distinguish between the benign early repolarization and early repolarization at-risk.

In 1992, Brugada and Brugada described an autosomal dominant disease characterized by ST elevation in the right precordial leads (V1 to V3), right bundle branch block and susceptibility to ventricular tachyarrhythmia in presence of a structurally normal heart disease. This entity is now called “Brugada syndrome”. In 1998, the gene responsible for some cases (25%) of Brugada syndrome was identified as the cardiac sodium channel gene (SCN5A), the same gene responsible for LQT3 variant of long QT syndrome. It’s common in young Asian males and there is a striking male preponderance (M: F- 8:1). It may manifest as syncope or cardiac arrest, usually in the 3rd or 4th decade of life. Cardiac events typically occur during sleep or at rest. The initial data from Brugada published in 1992 suggested the risk of sudden death with in 3 years from diagnosis to be 30% in both asymptomatic and symptomatic individuals. Subsequent data estimated the risk to be much lower.

Because the diagnosis is easily made by means of the ECG, an increasing numbers of patients with this syndrome are being identified. In some individuals the concealed and intermittent form may complicate the diagnosis. Concealed forms may be unmasked only with pharmacological provocative test (ajmaline, flecainide or procainamide) which also accentuate the ST segment elevation. The ST segment changes are different from the ones observed in acute septal ischemia, pericarditis, ventricular aneurysm and in some normal variants like early repolarization.

No drug therapy is effective in preventing arrhythmic events. The only prophylactic treatment to prevent cardiac arrest is ICD.

d) In cases of partial RBBB with early repolarization, if the echocardiography and the stress test are normal there is no aeromedical implication and the individual can exercise unrestricted flying privilege after a brief period of observation.

No guidelines exist at present for the disposal of Brugada syndrome. Considering the risk of sudden ventricular arrhythmia and cardiac death (which can not be objectively measured) it seems prudent ground the aircrew.

e) Partial right bundle branch block with early repolarization.