Granulomatous Atrial Cardiomyopathy presenting with Atrial arrhythmias and mediastinal lymphadenopathy – A new syndrome.

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Introduction: We present a series of patients presenting with granulomatous inflammation and thoracic lymphadenopathy presenting as atrial arrhythmias. Such an association has not been recognized before.

Methods: Twelve patients presenting with atrial arrhythmias including Atrial fibrillation (AF), Atrial flutter (AFL), Atrial tachycardia (AT) and Atrioventricular nodal reentrant tachycardia (AVNRT) were included in this study. Computed chest tomograms and 18-fluorodeoxyglucose positron emission tomographic scans (18FDG PET-CT) were performed in all patients. A biopsy of the lymph nodes or myocardium was performed in 10 patients to diagnose granulomatous inflammation characteristic of Sarcoidosis or Tuberculosis. We tested for evidence of tuberculosis by tuberculin test, culture and PCR in all patients.

Result: The mean age of patients was 47±14.03 years and they were predominantly male (66.6%). The Left ventricular ejection fraction (EF) at presentation was 49.25±14.93% and left atrial size was 4 cm. All patients had AF as the presenting rhythm while AFL was present in 4 patients and one patient had AVNRT. There was evidence of abnormal PET uptake in atrial myocardium without significant ventricular uptake in all patients. Abnormal PET uptake was also present in the mediastinal lymph nodes (75%) and cervical lymph nodes (25%). Cardiac sarcoidosis was the diagnosis in 75% of the patients while the remaining had evidence of tuberculosis in addition to sarcoidosis. Four of these patients presented as stroke with atrial fibrillation. All patients were treated with immunosuppressive therapy (Corticosteroids and Methotrexate) and 4 patients received anti-Tuberculosis therapy in addition to immunosuppression. Over a mean follow up of 27 months, there was a significant improvement in clinical status commensurate with a decline in atrial PET uptake in 75% of the patients and the LVEF also significantly improved to 56.58±13.36% (p=001).

Conclusion: Atrial arrhythmias with evidence of granulomatous lymphadenopathy and atrial inflammation may be a presenting feature of Cardiac sarcoidosis or Tuberculosis. This syndrome should be suspected in young individuals presenting with multiple atrial arrhythmias and stroke in the absence of conventional risk factors.