Pheochromocytoma masquerading long QT with T wave alternans and leading to subsequent syncope due to Torsades des pointes

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Introduction: Long QT syndrome is an important cause of malignant arrhythmias leading to syncope and sudden cardiac death, especially in younger population. Being a genetic disease, it remains incurable and requires lifelong palliative treatment with beta blockers and implantable cardioverter defibrillators. T wave alternans in the presence of long QT syndrome is an ominous marker and warrants aggressive treatment. Pheochromocytoma mimicking long QT syndrome and leading to TDP has been rarely reported. However, to the best of our knowledge, this is the first case demonstrating long QT syndrome with T wave alternans secondary to pheochromocytoma.

Methods: An 11-year-old boy, born out of consanguineous marriage, presented with recurrent episodes of palpitations, sweating and syncope related to exertion over a period of 1 month. A diagnosis of long QT syndrome with T wave alternans was made on baseline ECG. The child was immediately started on propranolol and cardiac sympathetic denervation was contemplated. There was no family history of sudden cardiac death. However, echocardiography surprisingly revealed gross left ventricular hypertrophy (LVH) with normal systolic function and without outflow tract obstruction. Cardiac MRI done to investigate LVH demonstrated concentric hypertrophy without any scar suggestive of secondary LVH. During the hospital stay, the child was noted to have episodic brief torsades des pointes (TDP) along with significant fluctuation of heart rate and blood pressure. A clinical suspicion of pheochromocytoma in view of LVH and fluctuant vitals, triggered further investigations. A raised urinary vanillylmandelic acid (VMA) and a 3.3 x 3.0 x 3.4 cm contrast enhancing mass arising from left adrenal gland clinched the diagnosis. Blood pressure control and QT interval improved after initiation of prazosin.

Result: The child underwent laparoscopic excision of tumour after ruling out tumour metastasis on 68Ga-DOTATATE PET-CT. Histopathological analysis of the specimen revealed tumour cells expressing synaptophysin, chromogranin and S-100 protein confirming the diagnosis of pheochromocytoma Post-surgery QT interval returned to normal and the child remained asymptomatic without any medications.

Conclusion: The case illustrates possibility of pheochromocytoma being an important treatable secondary cause of long QT syndrome with T wave alternans and malignant ventricular arrhythmias. High index of suspicion, as in this case, holds the key for making the diagnosis and appropriate management.