CAROTID ARTERY DISSECTIONS, FIBROMUSCULAR DYSPLASIA AND FAMILIAL LONG QT SYNDROME-A UNIQUE ASSOCIATION-THERAPEUTIC CHALLENGES

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A 38-year-old woman with history of familial Long QT Syndrome (LQTS) and migraines without aura presented with one month history of bilateral carotidynia. Her 14-year-old son had LQTS genotype 2. Neurologic examination was normal except for tender cervical carotid arteries. Carotid ultrasound showed a bulbous cervical dilatation of both proximal internal carotid arteries (ICAs). Computed Tomography Angiography showed bilateral ICAs dissections and underlying fibromuscular dysplasia (FMD). Magnetic Resonance Angiography of the neck demonstrated aneurysmal dilatation of ICAs with small residual lumen and subacute thrombus on axial T1-weighted fat-suppressed images. Additional investigations showed right renal artery FMD. She received therapeutic doses of enoxaparin as a bridge to warfarin; target International Normalized Ratio of 2.0 to 3.0.

Cervicocephalic arterial dissections (CCADs) have not been previous reported in association with LQTS. Therapeutic interventions in CCADs have included immediate anticoagulation with heparin followed by 3-to 6-month course of warfarin, and platelet antiaggregants instead of, or following, warfarin. Angioplasty and stenting or surgical correction is reserved for selected individuals with pseudoaneurysms or those who have multiple recurrent events following adequate medical therapy. Although anticoagulants are often empirically recommended, their value in patients with extracranial CCADs has not been firmly established. Anticoagulation is withheld in patients with intracranial dissections because of the risk for subarachnoid hemorrhage. The Cervical Artery Dissection Study (CADISS) is an ongoing randomized multi-center open treatment trial comparing therapy with anticoagulants or antiplatelets for acute symptomatic extracranial carotid or vertebral artery dissection. We suggested subgroup analysis of patients with intraluminal arterial thrombus.