

A TRICKY CASE OF ABNORMAL ECG FINDINGS LEADING TO A DIAGNOSIS OF TUBEROUS SCLEROSIS

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ABSTRACT

A 16-months old infant, previously healthy, was admitted to our department for a focal onset seizure. During the hospital stay, a pre-sedation electrocardiogram (ECG) was conducted in view of a brain Magnetic Resonance (MRI) in deep sedation scheduled in order to study origin of his focal seizure.

A peculiar ECG pattern was depicted with dome-shaped ST segment elevation in V5, V6, D I, aVL leads and T waves inversion in V1 and V2.

In addition, because of numerous hypochromic café au lait patches detected on trunk and four limbs, diagnostic process of tuberous sclerosis was started. No lesions were found in abdomen echography. An echocardiography was conducted and a diagnosis of pericarditis with hyperechogenicity of antero-lateral pericardial sheets with mild pericardial effusion deserving anti-inflammatory therapy.

Both segmental and global function were preserved and no coronary anomalies were depicted. Daily electrocardiograms were performed, always comparable with the first one.

A brain MRI highlighted millimetric subependymal nodular lesions both in the right frontal horn, medium cell and lateral ventricle and in the left frontal horn (maximum 6 mm with a calcified aspect) and medium cell. Taking this into account, besides hypochromic café au lait lesions, a diagnosis of tuberous sclerosis could have been asserted.

Electrocardiogram data have been remained stable, with an ST elevation in lateral leads whereas echocardiography showed a mild pericardial hyper-refraction without typical lesions of tuberous sclerosis. In light of optimal clinical conditions of the patient, the non-unique interpretation of ECG findings and a substantial negative echocardiography, an acute condition such as pericarditis was excluded and the anti-inflammatory therapy was stopped.

Sequential ECG and echocardiography was performed highlighting a persistent antero-lateral ST elevation with mild pericardial effusion (maximum 4 mm) and hyperechogenicity of antero-lateral pericardial sheets (13 mm x 4 mm) fitting with the diagnosis of tuberous sclerosis rhabdomyomas.

An elevation of ST segment could be tricky to be interpreted. In our case ECG findings of antero-lateral dome-shaped ST and T waves inversion and the consensual ovular lesion found in the pericardial context, have been interpreted at first as a myo-pericarditis; subsequently, as the diagnosis of tuberous sclerosis has come, we have interpreted them as infiltration of the rhabdomyoma into the normal conduction system. To the best of our knowledge, it is extremely rare to assess a diagnosis of tuberous sclerosis starting from an abnormal ECG.

