Hedinger syndrome - rare disease, easy diagnosis, bad prognosis

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Introduction: Hedinger syndrome or carcinoid heart disease is a term that represents all cardiac manifestations of carcinoid syndrome. It is a rare disease that requires a high level of clinical suspicion. The presence of cardiac symptoms in the context of carcinoid syndrome means an advanced stage of the disease.[1,2]

Case report: We present the case of a 65-year-old male with clinical manifestation of progressive dyspnea, leg edema and profuse diarrhea. Before cardiology referral, the patient underwent multiple diagnostic tests for diarrhea including an infectious disease and gastroenterology workup. Upon presentation, after taking the patient's history and performing the physical examination, which showed signs of cardiac decompensation, an electrocardiogram was performed showing sinus rhythm with tachycardia and right bundle branch block. Laboratory tests showed polycythemia with slightly elevated cardiac troponin T and N terminal pro brain natriuretic peptide levels. Due to his symptoms combined with 12-lead ECG abnormalities, CT pulmonary angiography was performed, and pulmonary embolism was excluded. Upon admission transthoracic echocardiography was performed and showed preserved ejection fraction, however the tricuspid valve insertion was higher than the mitral valve with signs of severe tricuspid regurgitation. Transesophageal echocardiography excluded the presence of shunts and confirmed severe tricuspid regurgitation with a failure of tricuspid leaflets coaptation. Tumor markers showed an elevated neuron-specific enolase and chromogranin A levels. Hedinger syndrome was suspected. Further tests showed an increase in 5-hydroxyindoleacetic acid. Thoracic and abdominal CT showed liver metastases of unknown origin. A biopsy of the liver lesions was suggestive of a neuroendocrine tumor. Later, a somatostatin receptor scintigraphy showed multiple positive liver lesions, and a magnetic resonance cholangiopancreatography confirmed a neuroendocrine tumor of the tail of the pancreas. Despite all treatment modalities, the patient's condition got progressively worse, until he eventually passed away.

Conclusion: Due to bad outcomes, it is important to rise clinical suspicion of Hedinger syndrome in patients with right-sided heart failure and vasomotor changes, mainly by imaging methods and specific laboratory tests.[2,3]

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